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PROLONGED USE OF PROSTAGLANDIN E_1 TO MAINTAIN PATENCY OF THE DUCTUS ARTERIOSUS IN CONGENITAL HEART DISEASE.

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Prostaglandin E_1 (PGE_1) has been used to maintain the patency of the ductus arteriosus when a shunt is required for adequate pulmonary blood flow or for oxygenation of the systemic blood. Previously an infusion of 0.1 μ g/kg/min has been used to stabilize infants temporarily prior to surgery. Maximum reported infusion time has been 4 days. Side effects such as apnea and hyperthermia may occur. In the past 5 months, we have used PGE_1 in 5 infants with right or left sided obstructive lesions. In one infant with transposition of the great vessels and high pulmonary pressure after a balloon septostomy, we infused PGE_1 intermittently for 18 days in an effort to increase pulmonary blood flow. Multiple attempts to wean the infant from PGE_1 resulted in marked hypoxemia. Selected results from the 18 day infusion are shown below:

Age (days)	1	4	11	9	2	5	3	7	1	8	8	15	15	16	18	20	20	20	25
μ g/kg/min	0	0	1	0	0	1	0	1	0	1	1	0.07	0.03	0.02	0	0.007	0	0	0
PaO_2 (mmHg)	14	44	17	32	43	19	41	48	44	37	46	28	41	34	29				

On days 16-20, the infant maintained an excellent pO_2 on far less than the recommended dose and gradually could be weaned from the drug without developing acidosis. Side effects noted at 0.1 μ g/kg/min did not occur at low doses. This experience suggests that 1) long-term infusion of PGE_1 may be a feasible way to manage small infants with certain complex cardiac conditions who are not deemed suitable for corrective surgery early in life and 2) an adequate response with fewer side effects may be realized at lower doses.

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ASSESSMENT OF ELECTROCARDIOGRAPHY AND VECTORCARDIOGRAPHY IN THE DETECTION OF RIGHT VENTRICULAR HYPERTENSION IN PATIENTS WITH POST OPERATIVE RIGHT BUNDLE BRANCH BLOCK.

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The electrocardiogram (ECG) and vectorcardiogram (VCG) have been used as a predictor of right ventricular pressure (RVP) elevation. However, in cases of postoperative (PO) tetralogy of Fallot (TF), the correlation between the ECG and VCG and increased RVP remains controversial due to the ECG pattern of right bundle branch block (RBBB). 21 patients (pts) with TF with PO RBBB by ECG, 3 to 17 yrs, who underwent cardiac catheterization at least 1 year PO, were evaluated. All 21 had ECG's while 11 had VCG's. Group A included 9 pts (4 with VCG) whose RVP > 46 mmHg; Group B included 12 pts (7 with VCG) whose RVP ≤ 45 mmHg. Multiple ECG and VCG parameters were compared to RVP. Among Group A pts with VCG, the right maximal spatial voltage (RMSV) and the terminal anterior forces (TAF) were both greater than 1 mV (mean 1.2 mV and 1.52 mV, respectively) in all pts. In Group B, the RMSV was less than 1 mV (mean 0.61 mV) in 11 of 11 pts, while the TAF was less than 1 mV (mean 0.67 mV) in 10 of 11. On the ECG of Group A, 55% of patients had a QR pattern in V1 (0% in Group B), while 45% had a right superior axis (RSA) (10% in Group B). Numerous other ECG and VCG parameters measured were not useful. Thus, the RMSV and the TAF of the VCG appear helpful in predicting residual RVP elevation, while the ECG, except for the presence of a QR in V1 or RSA, is not helpful.

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DYNAMIC EXERCISE TESTING WITH ECHOCARDIOGRAPHIC CORRELATES IN THALASSEMIA MAJOR (Thal).

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27 patients aged 5-28 yrs. with Thal were studied by exercise testing (ET) (Bruce protocol) and by M mode echocardiography (Echo). Hemoglobin (Hgb) was determined 1 hr. prior to ET. 19/27 had no digitalis therapy (DIG). Endurance time: 16/27 had low endurance time (END) ($< 60\%$ of predicted mean [PM] for age); all patients with Hgb < 11 gms.%, 5/6 boys and 4/7 girls > 15 yrs., and 4/8 boys and 2/6 girls < 15 yrs. had low END. END was not predictable by ejection fraction (EF) on Echo nor related to DIG. Heart rate response (HR): 8/27 (2 boys, 6 girls) had decreased HR (> 2 standard deviations below the PM) at maximal exercise (ME), not related to age or Hgb. 4/27 (2 boys, 2 girls) had increased HR at ME, not related to age or Hgb. EF and left ventricular internal diameter shortening on Echo were not predictors of HR. Dysrhythmias: 1/27 developed ventricular premature contractions (runs of 2 or 3) not seen on 24 hr. taping or standard ECG. 1/27 showed atrial premature beats. 1/27 on DIG showed sinus arrests during recovery. ST segments-T waves: No changes of ischemia were noted. Conclusions: (a) 60% of patients with Thal showed decreased END that correlated with decreased Hgb (< 11 gms.%); (b) HR was not related to Hgb, age or DIG and was unexpectedly low in 1/3 of the patients; (c) myocardial function by Echo was not a predictor of exercise response; (d) an ischemic response was not identified; (e) dysrhythmias were uncommon; (f) Echo and ET showed less myocardial dysfunction than expected by the natural history of Thal with acquired hemochromatosis.

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HEMODYNAMICS AND MYOCARDIAL BLOOD FLOW IN THE FETUS WITH CHRONIC PULMONIC STENOSIS IN UTERO

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We produced pulmonic stenosis (PS) in nine fetal sheep by banding the main pulmonary trunk at ± 0.45 gestation. Systemic and regional myocardial blood flow was subsequently studied at 0.85 gestation (133 ± 1.4 d) using radionuclide-labeled microspheres. Right ventricular (RV) pressures were elevated to 110 ± 11 mm Hg systolic, and 16.8 ± 6.9 mm Hg end-diastolic, compared with left ventricular (LV) pressures of 72 ± 7 mm Hg systolic and 9.8 ± 4 mm Hg end-diastolic. No superior vena caval (SVC) blood crossed the foramen ovale (FO) in a group of control animals whereas LV output was increased in the PS animals since 22% of SVC blood crossed the FO to the left atrium. At autopsy LV volume was similar in PS and control animals but RV volume was greater in the PS group (4.8 vs. 1.2 ml, $p < 0.001$). The ratios of subendocardial to subepicardial myocardial blood flow (I/O) of the control LV (1.44) and RV (1.37) were similar to those in normal conscious adult dogs. In the PS animals, LV I/O was normal (1.31) but RV I/O was reduced (1.04, $p < 0.02$). The reduced RV subendocardial flow in fetuses with PS is explained by the elevated RV pressures and volume and may have important influences on myocardial function and development.

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ECHOCARDIOGRAPHIC ASSESSMENT OF CARDIAC FUNCTION IN THE IMMEDIATE POSTOPERATIVE PERIOD.

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Echocardiographic (E) assessment of cardiac function (CF) was carried out preoperatively and at 1, 4, and 24 hours (h) post-operatively (PO) in 20 pediatric patients (pts) with varied congenital heart defects. Simultaneous measurements of cardiac output (CO) and clinical and hemodynamic data were obtained. The pts were divided into 3 groups: (I) sustained $CO \geq 2.2$ L/min/m² without pressors (P) (N=9); (II) low $CO < 2.2$ L/min/m² and/or use of P with subsequent improvement (N=8); (III) low CO and use of P with subsequent death (N=3). Left ventricular systolic time interval ratio (PEP/ET) and aortic root rock (degrees of systolic anterior motion of the aortic root from the horizontal plane) (AoR) correlated best with overall assessment of CF.

	I	II	III
PEP/ET: Preop	.31(.25-.36)	.36(.27-.49)	.34(.26-.46)
1 h	.36(.19-.48)	.57(.41-.75)	.62(.45-.97)
24 h	.35(.27-.50)	.53(.40-.69)	.98
AoR($^\circ$): Preop	32 (20-43)	31 (23-39)	34 (21-40)
1 h	16 (8-26)	13 (9-19)	8 (0-15)
24 h	20 (9-27)	16 (9-30)	0

CO, estimated using a formula based on the opening slope of the mitral valve, correlated well with CO determined by dye dilution. We conclude: (1) E is useful for monitoring CF and CO in the immediate PO period; (2) PEP/ET and AoR allow rapid noninvasive assessment of CF, early identification of pts who require intensive therapy, and monitoring of response to treatment.

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TRANSCUTANEOUS OXYGEN TENSION ($P_{tc}O_2$) IN NEONATES WITH CYANOTIC CONGENITAL HEART DEFECTS (CCHD) AND PULMONARY DISEASE (PD).

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Previous studies in sick infants utilizing transcutaneous O_2 sensors indicated significant fluctuations in resting $P_{tc}O_2$ and marked decreases in response to crying (Ped. Res. 10:422, 1976). The purpose of this study was to determine the pattern of $P_{tc}O_2$ in babies with CCHD as compared to a matched group of neonates with PD. Continuous $P_{tc}O_2$ recordings were obtained in 18 newborns: 8 with CCHD and 9 with PD. Observations were made during the resting state and 38 episodes of crying.

Resting $P_{tc}O_2$ was 26 ± 12 torr in the CCHD group and $66 \pm$ torr in the babies with PD. Fluctuations were virtually absent in the patients with CCHD (average variability: CCHD = ± 0.5 ; PD = ± 4.0 torr). $P_{tc}O_2$ fall in response to crying was found to be significantly smaller in the CCHD group (6 ± 4 torr) than in the PD patients (22 ± 16 torr). An exception was an infant with pulmonary atresia after a Waterston shunt who had predominantly pulmonary manifestations, and displayed a significant fall in $P_{tc}O_2$ (12.5 torr) during crying.

The data indicate that infants with intracardiac R $\rightarrow$ L shunts have a non-varying pattern of arterial O_2 desaturation as compared with the more labile behavior of the PD group. The stability of $P_{tc}O_2$ in neonates with CCHD may have diagnostic usefulness, and supports the reliability of isolated arterial pO_2 determinations in babies with heart disease.