175 AMRINONE FOR PULMONARY HYPERTENSION IN INFANTS AND CHILDREN. Thomas J. Kulik, Dana E. Johnson, Thomas P. Green, James E. Lock. University of Minnesota Medical School, Univ. of Minn. Hospitals, Dept of Pediatrics, Minneapolis

Amrinone (AM) is an orally active inotropic agent that is both a systemic and a pulmonary vasodilator. We used intravenous AM to treat life-threatening pulmonary hypertension (PH) in 4 neonates (NEO), 12 to 36 hrs old, in whom right to left ductal shunting indicated severe PH; and in 5 children (CH), 6 mo. to 12 yrs old, whose mean pulmonary artery pressure (PAP)≥ mean aortic pressure (AoP). These patients were unresponsive to hyperventilation and conventional pharmacologic therapy of their PH.

All NEO had persistant fetal circulation (PFC), either idiopathic or 2° to a diaphragmatic hernia, and had severe cyanosis without heart failure. AM (1-3 mg/kg bolus) decreased AoP by 22% (+2% to -37%), and decreased arterial pO_2 (-6 torr; range: +2 to -22 torr), possibly indicating a greater fall in AoP than in PAP.

Of the 5 CH, 4 had idiopathic PH, 1 had PH 2° to a previously closed VSD; all were acyanotic. A single dose of AM (1-3 mg/kg) had little effect on AoP (-3% to +3%) or PA wedge pressure (measured in 2 patients, ±1 mmHg), but it markedly decreased PAP in all CH (22% to 44%; mean = 27%, p<.01). However, in 3 of the 5 CH, little change in PAP occurred after a 2nd or 3rd dose of AM.

To summarize: In NEO with PFC, AM was not beneficial and may have somewhat increased cyanosis. In CH with PH, AM initially selectively decreased PAP but early tachyphylaxsis was seen in most children. This tachyphylaxsis may severely limit AM's role in the treatment of PH .

† 176 LONG-TERM RESULTS OF BALLOON VALVULOPLASTY OF THE PULMONARY VALVE. D. Kveselis, A.P. Rocchini, A. Rosenthal. Univ. of Michigan

The long-term efficacy of transluminal balloon valvuloplasty (val) was evaluated in 7 children (age 1.5-9.9 yrs) with pulmonary stenosis 1 yr post-val. Prior to, immediately (Imm) post-val and at 1 yr post-val, the following parameters were evaluated at cath: pulmonary artery (PA), right ventricular (RV) and aortic pressures, heart rate (HR), pulmonary blood flow (PBF) and pulmonary valve area (PVA); ECG, vectorcardiogram (VCG) and doppler echocardiogram were performed.

	HR bpm	PBF L/min/m²	RVp mmHg	RV-PA Gradient mmHg	PVA cm²/m²
Preval	100±15	3.30±0.36	108±30/10±2	89±32	0.34±0.07
Postval	107±22	3.33±0.45	60±5**/8±2	39±5**	0.50±0.18**
Followup	93±9	3.86±1.2	56±12**/7±2	38±10**	0.76±0.29**,+

**p<0.001 prevalv vs. postvalv, prevalv vs. followup; +p<0.05 postvalv vs. follow up X±SD

One yr post-val, RV-PA gradient and RV press decreased further from Imm post-val values in 6/7 pts. In addition, there was also an increase in both PBF and PVA over Imm post-val values. Although the ECG only mildly improved 1 yr post-val (R in V_1 decreased from 13±2 to 11±4 mm), the VCG significantly improved with a decrease in right maximal spatial voltage (2.0±.9 to 1.1±.4 mV, p<.05) and a loss of the clockwise horizontal loop in all pts. Pulmonary regurgitation was present at 1 yr post-val (auscultation 2/7; doppler 6/7). Inferior vena cava thrombosis occurred in 1 pt. Balloon val appears to be an effective, safe method for producing long term relief of valvar pulmonary stenosis.

177 SIMPLIFIED METHOD FOR DETERMINING LEFT-TO-RIGHT SHUNTS USING PULSED DOPPLER ECHOCARDIOGRAPHY Alan M. Langsner, Eva B. Griepp, Brian Kiely and Monika M. Rutkowski (sponsored by Eugenie F. Doyle) New York University School of Medicine, Division of Pediatric Cardiology, N.Y., N.Y.

The principle of using Doppler techniques to measure left-to-right shunts noninvasively has been established. In practice, however, problems have been encountered using previously described methods, which involve determining the cross-sectional area of the aorta (Ao) and pulmonary artery (PA), or equivalent sites: these measurements are difficult and/or impossible to obtain accurately in the pediatric population, and small errors in the measured values may be magnified during calculation.

To simplify the method, only the areas of the Doppler velocity curves were used to calculate the magnitude of left-to-right shunting, expressing the ratio of these flow integrals as Doppler Q_P/Q_S. The short axis parasternal view was used for the main PA and the suprasternal view for descending Ao since these views allow sampling essentially parallel to blood flow, eliminating the need to correct for sampling angle.

Doppler flow ratios were compared to Q_P/Q_S obtained by oximetry at cardiac ctheterization in 18 children with uncomplicated shunts, ages 1 week to 14 years: 5 with atrial septal defect (ASD), 11 with ventricular septal defect (VSD), and 2 with both ASD and VSD. The correlation coefficient was 0.83 (regression analysis y = 1/2 + 3x/5) demonstrating that this simplified method gives accurate results while eliminating the need for difficult and problematic echocardiographic and Doppler flow measurements.

178 THE HEMODYNAMIC EFFECTS OF BUCINDOLOL IN CHILDREN WITH CONGESTIVE HEART FAILURE (CHF). Linda Leatherbury, Roger N. Ruckman, David Deitchman Lester F. Soyka, Children's Hospital National Medical Center, Department of Cardiology, Washington, DC

The investigational drug Bucindolol has intrinsic sympathomimetic activity and direct vasodilating actions in animal models and adult humans with CHF and/or hypertension. Five children (2 1/2 - 21 mos; 4.0 - 10.4 kg) with ventricular septal defect or endocardial cushion defect and CHF had baseline pulmonary to systemic flow ratios (Q_P/Q_S) ≥ 3.0 at cardiac catheterization. After informed consent, each received a .05mg/kg, 5 min. infusion of Bucindolol. Hemodynamic measurements showed significant increases in pulmonary resistance/systemic resistance (Rp/Rs), and Rp (Wood units); significant decreases, in indexed pulmonary flow (Qp), left to right shunt (L→R), and (Q_P/Q_S) in 100% of the children; but no significant change in pulmonary mean pressure ($\overline{PA}$), aortic mean pressure ($\overline{AO}$), Qs, Rs or indices of ventricular function. CHF improved transiently in all children and most dramatically in the ones with more severe CHF. The pre and post drug means ($\overline{X}$) and mean % changes (%Δ) from pre drug values were:

$\overline{X}$	Qp	Qs	L→R	Q_P/Q_S	$\overline{PA}$	$\overline{AO}$	Rp	Rs	Rp/Rs
pre	13.0	3.8	9.3	3.7	43	66	3.3	19	.18
post	7.8	3.2	4.9	2.5	50	61	6.1	20	.32
%Δ	-40	-12	-48	-28	+16	-8	+97	+9	+85

We conclude that Bucindolol is of potential value in managing CHF in infants with congenital heart lesions having high pulmonary flow.

179 Monomorphic Ventricular Tachycardia (VT) in Childhood: Electrophysiologic and Pharmacologic Characteristics.

Thomas C. Martin, Antonio Hernandez, and Michael E. Cain, Washington Univ. Sch. of Med., St. Louis. (Spon. by David Goldring)

Three children with accelerated ventricular rhythms (AVR), ages 12, 14 and 15 years, were evaluated. Symptoms included chest pain (2 patients) and dizziness (1 patient). Chest x-rays were normal; ECG revealed normal axis, voltage and QT interval in sinus rhythm. Echocardiogram revealed only mitral valve prolapse (in 2 patients with chest pain). Exercise testing resulted in suppression of AVR in all patients. Holter monitor revealed multiple runs of nonsustained AVR in all patients (rate 70, 80, and 130 bpm). The QRS demonstrated a LBBB pattern in all patients (2 with RAD). Isoproterenol infusion resulted in suppression of fast AVR in one patient, while it resulted in acceleration in both patients with slow AVR (rate 110 and 130 bpm after infusion). Thus, all 3 patients demonstrated VT (rate >100 bpm). Electrophysiologic testing including programmed ventricular stimulation, revealed normal conduction systems and no inducible VT. Overdrive suppression of AVR was possible. Earliest endocardial activation was in the RVOT in 2 patients. Propranolol resulted in AVR suppression in 2 patients, exacerbation in 1. Verapamil and procainamide resulted in AVR suppression in 2 patients to whom the drugs were given. Monomorphic VT in children with normal myocardium is a subset of VT with a unique mechanism and variable pharmacologic response. Prognosis is probably good.

180 CARDIOVASCULAR ABNORMALITIES IN PATIENTS WITH ANOREXIA NERVOSA.

Douglas S. Moodie, Laurie Sandler, Donald Kirkendall, Ernesto E. Salcedo, Fetnat M. Fouad. Cleveland Clinic Foundation, Department of Cardiology, Cleveland, Ohio.

Because the 15% mortality rate in anorexia nervosa may be associated with cardiovascular abnormalities, we studied 8 anorectic patients (pts) ages 16-24 yrs (mean age 18 yrs) with chest x-rays, two dimensional echocardiograms, detailed body composition measurements, bicycle ergometer stress exercise testing, and nuclear angiographic studies of resting and exercise cardiac function. Compared to controls, our pts total weight was reduced 47%, percent fat was 67% below control, and free fat mass was reduced 33%. Chest x-rays revealed a cardiothoracic ratio <0.40 in all pts (mean 0.34). Heart weight in grams as measured by echocardiography was well below the third percentile in all pts: LV mass was <100 grams in all pts. Resting cardiac index was reduced in all pts (mean 2.2 liters/min/m²) with reductions in stroke volumes (mean 56.6 ml/min/m²) and an increase in the cardiac mean transit time. Cardiopulmonary volume - total blood volume ratios were normal, but there was a marked reduction in total blood volume. There was a blunted heart rate and blood pressure response to exercise with a marked reduction in oxygen consumption and total work performance. These changes demonstrate a compensated cardiovascular state in anorectics and suggest continued cardiac monitoring during follow-up to assess the short and long-term effects of decreased cardiac performance.