175 EFFECT OF AMRINONE ON CARDIAC FUNCTION IN CHILDREN WITH SEVERE CONGESTIVE HEART FAILURE (CHF). William A. Neal, West Virginia University, Morgantown, WV, and Mary E. Pierpont, University of Minnesota, Minneapolis, MN, Departments of Pediatrics.

Amrinone (A), a newly synthesized bipyridine derivative, has been shown to exert a positive ionotropic action when administered to adults with severe CHF. There are no published data regarding its affect on children.

Two children, 12 years old, fulfilled rigid criteria for inclusion in A protocol: failure to respond to conventional therapy, cardiac index (CI)$\leq$2.2 L/Min/M^2, and left ventricular filling pressure (PCW)$\geq$18 mHg. A was given by IV bolus 1 mg/kg and a sustaining infusion of 10 mg/kg/min, followed by oral administration of 50 and 75 mg TID respectively. Non-invasive and Swan-Ganz monitoring were utilized to establish efficacy of the drug. Maximum benefit was as follows:

	Control		Amrinone		
	CI L/Min/M^2	PCW mmHg	CI	PCW	ΔCI%
pt#1	1.40	23	3.40	12	143
pt#2	2.02	30	3.85	14	91

Both children had sustained improvement of cardiac function, though the first died of cardiomyopathy 4 days after A was begun. The second patient continues to show symptomatic and objective improvement with mild thrombocytopenia as the only adverse reaction. Limited experience with this new ionotropic agent in children with intractible CHF is encouraging.

176 COST-EFFECTIVENESS OF PREOPERATIVE CATHETERIZATION IN INFANTS WITH D-TRANSPOSITION OF THE GREAT ARTERIES, INTACT VENTRICULAR SEPTUM. Jane W. Newburger and Olli S. Miettinen (Sponsored by Alexander S. Nadas). Harvard School of Public Health and Harvard Medical School, Children's Hospital Medical Center, Department of Cardiology, Boston

We analyzed the need for routine cardiac catheterization (CC) prior to corrective surgery in infants age 6-12 mo. with d-transposition of the great arteries, intact ventricular septum (dTGA, IVS), s/p balloon atrial septostomy in the neonatal period, who have at most a grade 2/6 murmur, small left ventricular forces on electrocardiogram, and no evidence of fixed subpulmonic stenosis (subPS) by echocardiogram or previous angiography. We compared mortality and costs among those undergoing preop CC with those proceeding directly to a baffle procedure. We assumed 1) that the frequency of the above clinical constellation among infants with severe subPS is 1%, among infants with pulmonary artery hypertension (PAH) is 25%, and among those with neither complication is 90%; 2) that the frequency of severe fixed subPS is 5% and of PAH is 2%; 3) that the mortality from CC is 0.1%, from the baffle procedure in uncomplicated dTGA,IVS, 5%, from baffle procedure with subpulmonic resection, 15%, and from baffle procedure without subpulmonic resection in infants with severe subPS, 100%. On these premises, the policy of routine CC carries an excess mortality (0.4/1000) when compared with the policy of operating without CC. Even if the frequency of severe subPS were as high as 10%, the cost of averting one death by the policy of routine preop CC would be $5-10 million. Thus routine preop CC may not be warranted in this clinical setting.

● **177** EFFECT OF ISCHEMIA ON MECHANICAL FUNCTION IN THE NEONATAL RABBIT HEART. Kenya Nishioka, Jay M. Jarmakani, UCLA Medical Center, Department of Pediatrics, Los Angeles, California.

This study was designed to evaluate the effect of ischemia and reperfusion on myocardial mechanical function in the neonate. Newborn (NB,n=14) and adult (A,n=14) rabbit hearts were studied utilizing the isolated arterially perfused septal preparation that was maintained at 27°C or 33°C and paced at 90 beats/minute. The muscle was perfused initially with an oxygenated Krebs-Henseleit solution. After stabilization of mechanical function, 60 minutes of global ischemia was induced and then the muscle was reperfused with oxygenated solution. The muscle was kept in a humidified, warm, oxygen-poor atmosphere. Developed tension (DT), maximal rate of tension development (+dT/dt) and resting tension (RT) were recorded continuously. The changes in DT and +dT/dt were similar. During ischemia, +dT/dt declined hyperbolically at the same rate in all groups, and the values after 60 minutes of ischemia were not significantly different from zero. During ischemia, the RT increased significantly in both groups and the increase in the A was significantly ($P < 0.001$) greater than in the NB. After 60 minutes of reperfusion, the recovery of +dT/dt (97 $\pm$ 5% of control at 27°C; 48 $\pm$ 12% at 33°C) in the NB was significantly ($P < 0.001$) greater than in the A (58 $\pm$ 6% at 27°C; 16 $\pm$ 6% at 33°C). After reperfusion, the RT returned to control in the NB, but not in A. These data indicate that the NB myocardium is more resistant to ischemia than the A myocardium.

178 ASSOCIATION OF REYE'S SYNDROME AND CARDIOMYOPATHY George R. Noren, John D. Tobin, Jr., Nancy A. Staley, Richard W. Asinger, Stanley Einzig. Hennepin County Medical Center, University of Minnesota Medical School, Department of Pediatrics, Minneapolis, Minnesota

Seven children (aged 3½-15 years) consecutively admitted, with the diagnosis of Reye's were evaluated for cardiac dysfunction by echocardiography, EKG and serum enzymes. The preceding viral disease was varicella in 6 and influenza A in 1. Two died and had microscopic myocarditis. All had normal LD_1 and CPK-MB isoenzymes.

			Echocardiographic Studies			
Case	EKG	BSA	%ΔLVD	PEP/LVET	EF(%)	VCF
1(V)	nl	1.26	34	.42	67	1.83
2(V)	nl	0.78	37	.33	71	1.96
*3(V)	abn	1.19	15	.43	35	0.70
*4(V)	abn	1.15	17	.39	37	0.88
5(V)	nl	0.76	27	.38	54	1.08
+6	abn	1.66	--	.46	--	--
7(V)	abn	--	Echo study unsatisfactory in this patient.			

*=deaths; +=pericardial effusion; V=varicella

In addition, a retrospective study of 17 cases of fatal varicella demonstrated microscopic myocarditis in 11. In none of the 11 children was myocarditis recognized prior to death. This study suggests that the heart is a significant target organ in Reye's syndrome, particularly when preceded by varicella, and its function must be considered in the care of these children.

179 POST-OPERATIVE ASSESSMENT OF FONTAN OPERATION Soraya Nouri, Mark Sivakoff, D. Glenn Pennington (Sponsored by Thomas Aceto), St. Louis University, Depts. of Pediatrics & Surgery, St. Louis, Missouri

From 1975 to 1980, 10 patients (pts.) age 2.5-24 years (y)(mean 13.6$\pm$6y) had modified Fontan operation for tricuspid atresia (7), univentricular heart (2), and transposed great vessels (1). Six had right atrial (RA) to pulmonary artery (PA) anastomosis (ANS), 4 with valved conduit and 2 direct ANS. Four had RA to right ventricular (RV) ANS, 1 with a non-valved conduit and 3 direct ANS. Eight pts. also had Glenn shunts. All 9 survivors are in normal sinus rhythm. Seven pts. had cardiac catheterization 2-60 months (m) post-operatively (post-op)(median 8m). RA mean pressures (p) were 3-19mmHg with RA 'a' waves of 10-25mmHg. Pts. with RA-PA ANS had narrow PA pulse p of 2-3mmHg, while pts. with RA-RV ANS had wider, biphasic PA pulse p due to RV contractions. No RA-PA p gradients were present. Aortic saturation was 79%$\pm$9 pre-operatively (pre-op) and 95%$\pm$3 post-op; left ventricular (LV) end diastolic p was 7.0$\pm$5.8mmHg pre-op and 6.7$\pm$1.9 post-op. RA regurgitation into hepatic veins and coronary sinus was more marked in pts. with RA-PA ANS. Post-op LV contractility was decreased in all (average LV ejection fraction 53.2%$\pm$12) except the youngest pt. While all pts. had improved exercise tolerance post-op, treadmill testing in 5 was impaired with average duration of 6.9 minutes, and maximal endurance index of 238$\pm$49 (normal 344$\pm$65). These data suggest that RA-RV ANS results in better forward flow and less regurgitation than RA-PA ANS. Exercise tolerance and oxygenation improved post-op but most pts. have clinical and angiographic evidence of decreased cardiac function.

180 HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY (HOCM) IN NEWBORN AND EARLY INFANCY PERIOD. Dov B. Nudel, Sandra C Brunson, Norman Gootman, Murray G. Baron, Milton J. Reitman, Helen Cahn, Nancy A. Chase. SUNY at Stony Brook, Long Island Jewish-Hillside Med Ctr., New Hyde Park, N.Y., North Shore Univ. Hosp., Manhasset, N.Y., Univ. of Tennessee, Memphis, Depts. of Pediatrics, Pathology and Radiology.

Clinical course and diagnostic studies in 6 infants under 8 weeks with HOCM are presented. One presented with severe CHF, one with severe arrhythmias; both died. In 4, findings were subtle; VSD-like murmurs, mild tachypnea and cardiomegaly. ECGs showed a superior axis in 5 and WPW in 1. Initial M-mode echos were inconclusive. Sector Scans done on 3 were diagnostic in 1, but became more specific at a later age. Cardiac catheterization (CC) was performed at age 1-49 (mean 18) days. Initial resting LV-AO gradient was 50$\pm$11 (n=5). Three showed the Brockenbrough response and in 2 no adequate post ectopic beat was recorded. Angiography showed severe septal hypertrophy, hypercontractility, distortion of LV cavity and compression of RV. In 4, the mitral valve (MV) was visualized and showed SAM. In 1 an additional fixed subaortic obstruction was suspected. A second CC in the survivors 10$\pm$4.25 mos. later showed a larger gradient, 80$\pm$8.17 in 3, and an unchanged gradient in 1. Followup period was 9-54 (mean 23) mos. and all survivors are well on beta blockers. Diagnosis was confirmed at autopsy in 2 and surgery in 1. Conclusion: 1) Prognosis was not related to pressure gradients. 2) Those presented with severe symptoms died. 3) Since physical findings, ECGs and echos in this age are atypical, a high index of suspicion and possibly CC is needed for accurate assessment.