●115 SEPARATION OF CARDIAC SARCOPLASMIC RETICULUM FROM FETAL AND MATERNAL SHEEP INTO FREE AND JUNCTIONAL SUBPOPULATIONS. Lynn Mahony, Michael Moulton (spon. by J. Warshaw), UTHSCD, Dept. Peds., Dallas.

We have previously shown decreased Ca accumulation in crude cardiac sarcoplasmic reticulum (SR) vesicles isolated from fetal sheep. However, recent data indicate that the cardiac SR is subspecialized and that these crude preparations are quite heterogeneous. To further define developmental changes in SR Ca transport, SR vesicles from fetal and maternal sheep hearts were seperated by sucrose density gradient centrifugation into free (F) and junctional (J) fractions.

	Fetal		Maternal	
	FSR	JSR	FSR	JSR
Ca ATPase activity (μmol Pi/mg protein/hr)	94±36	20.6±7.0	129±29	52±24
Ca Uptake(umol/mg protein)				
(-)Ryanodine	2.8±2.6	1.1±0.2	4.0±2.6	0.8±0.6
(+)Ryanodine		2.5±0.6		3.8±2.1

The FSR vesicles displayed a high density of Ca pumps and high Ca uptake that was insensitive to ryanodine. In contrast, although Ca uptake was decreased in JSR vesicles, uptake increased markedly in the presence of ryanodine. Gel electrophoresis showed selective enrichment of JSR vesicles with the Ca binding protein, calsequestrin. CaATPase activity and maximal Ca uptake were decreased 30-50% in fetal FSR and JSR ($p<0.05$). These data indicate that previously described differences in Ca transport in cardiac SR vesicles isolated from fetal and maternal sheep persist in our highly purified preparations of FSR and JSR. Further characterization of these SR subpopulations should enable us to better define developmental changes in SR function. *mean±SD,n=3.

116 CARDIAC PERFORMANCE DURING UPRIGHT EXERCISE BEFORE AND AFTER REPAIR OF COARCTATION. Anne M. Murphy, Mary Blades, Frederick W. James, and Stephen Daniels, Children's Hospital Medical Center, Cardiology Division, Cincinnati, Ohio

Coarctation patients are reported to have enhanced cardiac performance in the remote post-operative period. We examined data from coarctation patients (N=22) studied both before (mean 1.7 mo.) and after (mean 32 mo.) repair to evaluate cardiac performance. Controls were 10 longitudinally studied normal children matched for mean age, wt., ht., and BSA at time of first (COA-1 vs CTR-1) and second study (COA-2 vs CTR-2). Heart rate, blood pressure, and carotid pulse pre-ejection pre-ejection period to LV ejection time ratio (P/L) were measured. Cardiac output (CO) in L/min was determined by acetylene rebreathing in 8 COA and 6 CTR pts, with calculation of stroke volume (SV) in cc. Data was obtained at maximal exercise on an upright bicycle ergometer:

	HR	SBP	DBP	P/L	CO	SV	MWL+
COA-1	166	171	89	.279	7.5	52	373
CTR-1	183	144	74	.226	8.9	53	470
P	<.05	<.025	<.025	<.01	NS	NS	NS
COA-2	179	176	67	.252	11.9	74	600
CTR-2	173	159	71	.247	11.6	67	680
P	NS	NS	NS	NS	NS	NS	NS

(+ maximal workload Kg-M). At rest COA-1 also had significantly higher SBP, DBP and P/L. HR and CO did not differ at rest in COA-1 vs CTR-1, and no parameter differed at rest in COA-2 vs CTR-2. We conclude that patients studied both before and after repair of coarctation demonstrated a normalization of blood pressure and ventricular function (P/L). Cardiac performance was not elevated in either pre- or post-operative state.

117 THE SPECTRUM OF PULMONARY VASCULAR DISEASE IN INFANTS AND CHILDREN WITH CARDIOMYOPATHY. Anne M. Murphy, David Witte, Kevin Bove, Samuel Kaplan, Cardiology and Pathology, Children's Hospital Medical Center, Cincinnati, Ohio

The presence of pulmonary vascular disease (PVD) in patients undergoing cardiac transplantation may result in death from acute right ventricular failure. This study was undertaken to explore the frequency and severity of PVD in pediatric patients with cardiomyopathy. We reviewed autopsy records to identify patients with pathologic changes of chronic cardiomyopathy. The pulmonary slides were reviewed by two pathologists and graded using Heath-Edwards criteria for arterial changes; and normal, mild, moderate and severe for changes in the pulmonary veins. A total of 6/16 patients had Heath-Edwards grade 2 or 3 and/or moderate to severe changes in the pulmonary veins (Group I); the remaining 10 had mild or no evidence of PVD (Group II). Group I patients generally had a longer duration of illness (42±53 mo.), but it did not differ significantly from that in Group II (12±33 mo.). The age at diagnosis was significantly greater ($P<0.05$) in Group I vs. Group II (103±61 vs. 31±49 mo.). Mitral regurgitation was evident at diagnosis in 3/6 patients in Group I vs. 3/10 in Group II. We conclude that moderate to severe pulmonary vascular disease is common in pediatric patients with cardiomyopathy, and its severity cannot be predicted by the clinical course.

118 HEMODYNAMIC VALIDATION OF DOPPLER DERIVED PULMONARY ARTERY PRESSURE IN PATENT DUCTUS ARTERIOSUS Norman N Musewe, Jeffrey F Smallhorn, Lee N Benson, Patricia M Burrows, Robert M Freedom, Richard D Rowe. University of Toronto, Hospital for Sick Children, Div. Cardiology, Toronto. Canada.

Measurement of pulmonary artery pressure(PAP) in preterm infants would be valuable in monitoring their clinical course. The presence of a patent ductus arteriosus (PDA) in this setting provides access to the pulmonary vascular bed by non-invasive means. This study addresses the reliability of Doppler derived (DD) mean, systolic and diastolic pressure measurements in 2 groups of children (Group 1: n=15, age 3.5±4.0yrs, mean PAP 33±16 ; Group 2 n=5, age 0.6±0.1yrs, mean PAP 77±15) undergoing simultaneous cardiac catheterization. The influence of ductal size on pressure measurement was also assessed.

Results	Group 1			Group 2		
	systolic	mean	diastolic	systolic	mean	diastolic
Gradient Ao to PA	64±37	51±32	43±28	9±5	7±5	5±8
DD (mmHg)	67±31	47±26	36±24	7±5	4±3	7±4
R value	0.93	0.92	0.97	0.92	-	-
p value	<.001	<.001	<.001	<.03	NS	NS
S.E.E. (mmHg)	12	10	6	2	-	-

Ductal size (1-9 mm) did not affect results. Pure left to right shunting was noted in group 1, while bidirectional shunting occurred in group 2. Three patients with near systemic PAP showed no significant DD change with 100% oxygen or vasodilator infusion, consistent with measured PAP. PAP can be predicted under varying hemodynamic states by Doppler measurement of PDA shunt velocities.

119 TRIFASCICULAR BLOCK IN NEONATE OF MOTHER WITH LUPUS ERYTHEMATOSUS. Joon M. Park, Indu Agarwal, Edwing Contreras, Bruce Bartholomew, (Spon. by Surendra K. Varma). Texas Tech University Health Sciences Center, Department of Pediatrics and Department of Internal Medicine, Lubbock, Texas.

Trifascicular block (TFB) is rare in newborn infants. Unlike congenital complete heart block, TFB has not been previously reported in infants of mothers with lupus erythematosus. We report a newborn male (birth weight 3.2kg) born by emergency caesarean section for fetal bradycardia. Maternal past history was unremarkable. Only during the last month of her pregnancy did she take Hydralazine 10mg QID daily for hypertension. At birth, the baby had a heart rate of 60 per minute and was given Atropine without response, but improved with Isoproterenol. With the exception of bradycardia of 68 per minute, physical findings were normal. Initial electrocardiogram disclosed trifascicular block and 2:1 second degree atrioventricular block. The next day his rhythm changed to sinus rhythm and bifascicular block which have remained throughout his life. He is now 17 months old and is completely asymptomatic. Viral cultures were negative. First antinuclear antibody titers (ANA) were positive ≥ 1:640 in both mother and baby. SSA and SSB titers were negative in both. At 6 months of age ANA titer became negative in the baby. However, a persistent maternal ANA titer of ≥ 1:640 at 16 months postpartum with mild arthralgias suggests continuing evidence of systemic lupus erythematosus.

120 PREEXCITATION SYNDROMES AND PECTUS EXCAVATUM. Joon M. Park, Alfred R. Farmer, (Spon. by Surendra K. Varma). Texas Tech University Health Sciences Center, Department of Pediatrics, Lubbock, Texas.

Both pectus excavatum (PE) and the preexcitation syndromes have been shown to be associated with mitral valve prolapse. This study was conducted prospectively to determine the association of preexcitation syndromes in patients with PE. Seventy-six patients under eighteen years of age, mean of five years, with PE, underwent cardiac examination, electrocardiography and echocardiography. Fifty-eight were males and eighteen were females. Thirty-five had a family history of PE. Nineteen had mitral valve prolapse and nine had scoliosis. Eight (11%) had electrocardiographic evidence of preexcitation: Wolff-Parkinson-White type in five, Lown-Ganong-Levine type in three, whereas five had paroxysmal supraventricular tachycardia. In addition, we had preexcitation in 22% of females, 7% of males, 11%/10% of patients with/without a family history of PE, 11%/10% of patients with/without scoliosis, 11%/11% of patients with/without mitral valve prolapse, and 10%/11% of patients younger/older than eight years. This study indicates a high association of preexcitation syndrome in patients with PE, especially when female had PE. Its incidence was not particularly higher even in the presence of family history, scoliosis, mitral valve prolapse or in younger pediatric patients.