●127 NORMALIZATION OF PLASMA NOREPINEPHRINE FOLLOWING REPAIR OF INTRACARDIAC LEFT TO RIGHT SHUNTS. Robert D. Ross, Stephen R. Daniels, David C. Schwartz, David Hannon, Samuel Kaplan, Children's Hospital Medical Center and University of Cincinnati, Cincinnati, Ohio.

Plasma norepinephrine (PNE) is elevated in patients with congestive heart failure (CHF) and the degree of elevation corresponds to the severity of the CHF. This study was performed to determine whether PNE returns to normal in children with CHF secondary to a left to right intracardiac shunt after repair of the defect. We measured PNE in 32 children (aged 0.1 to 13.3 years) undergoing cardiac catheterization and divided them into four groups for analysis: I) Pre-operative (op) patients with severe CHF from an intracardiac left to right shunt (N=10), II) Intracardiac left to right shunts repaired 0.3 to 4.0 years previously (N=7) who had the same degree of left to right shunting (Qp/Qs) and CHF pre-op as Group I, but post-op were asymptomatic on no medications, III) Pre-op patients with cyanotic heart disease and no CHF (N=7), and IV) Cyanotic heart disease (N=8), repaired 0.7 to 7.0 years previously.

Results:	Group I	Group II	Group III	Group IV
PNE(pg/ml): Mean ± 1SD	906.0±294.8	318.7±161.0	286.0±64.6	205.±95.3
		p < 0.001	p = ns	

For group II, there was an inverse relationship between the LN PNE and time interval since repair (R=-0.71). We conclude that PNE is markedly elevated in patients with severe CHF but returns to normal in a log linear fashion after repair of intracardiac left to right shunts and resolution of CHF. In contrast, PNE in cyanotic heart disease is normal pre- and post-operatively.

128 SYSTOLIC AND DIASTOLIC LEFT VENTRICULAR FUNCTION AFTER ANATOMIC CORRECTION OF TRANSPOSITION OF THE GREAT ARTERIES.* George Sandor, Robert Freedom, Michael Patterson, Jacques Leblanc, William Williams, George Trusler, Phillip Ashmore(Spon. by Donald E. Hill)University of British Columbia, Department of Pediatrics, Vancouver, B.C.

Anatomical repair(AR)of transposition of the great arteries(TGA) has been done because of concerns of right ventricular function post-atrial repair. There is little information about LV function post-AR. This study assessed LV function in 3 patients post-AR.All had pulmonary artery banding, 2 VSD's and 1 coarctation. Mean age of repair was 2.75 yr.,mean age at catheterization was 4.9 yr. LV end-diastolic volume index(EDV_I),end-systolic volume index(ESV_I), stroke volume index(SV_I),ejection fraction(EF),mass index(M_I),end-diastolic pressure(EDP),peak systolic pressure(PSP),end-systolic pressure(ESP),peak systolic stress(PSS),end-systolic stress(ESS)& end-diastolic stress(EDS)were all calculated. From a monoexponential formula LV diastolic myocardial stiffness(K_E) was calculated. The rate corrected VCF($MVCF_c$)-end systolic stress relationship was obtained. These are compared with 10 controls and significant differences are shown in the Table.

	AGE	EDV_I	ESV_I	$MASS_I$	$LVSV_I$	EF	K_E
TGA	4.9	110.9	37.3	101.0	73.6	66.7	29.5
±	.85	4.74	3.69	16.9	3.52	.27	1.84
CONTROL	8.1	79.1	22.7	68.2	56.5	71.6	10.8
±	3.96	14.55	4.42	12.34	12.13	5.10	2.2
p	.036	<.001	<.001	.038	.0027	NS	<.001

The 3 TGA patients had normal $MCVCF_c$-ESS relationship although at low normal levels.Thus systolic function is normal but LV mass and diastolic function is abnormal post-AR.*Supp. by the B.C.Heart. Found.

129 FETAL TWO-DIMENSIONAL ECHOCARDIOGRAPHY (F2DE): IMPACT ON PRENATAL MANAGEMENT AND POSTNATAL OUTCOME. Thomas V. Santulli, Jr.; Roberta G. Williams. UCLA School of Medicine, CHS, Department of Pediatrics, Los Angeles

In a recent 16 month period, 400 women were referred for F2DE. Indications for study were similar to those previously published. Using 8 2DE views and Doppler, when warranted, to better describe anatomic, rhythm or function abnormalities, we obtained complete studies in 95% of those studied.

In this highly selected group, we found: 7 with potentially life-threatening structural heart disease (SHD); 4 suspected of having SHD based on previously published descriptions; 4 with persistent well-characterized rhythm alterations without evidence of congestive failure and without SHD; and 1 with SHD and uncharacterized rhythm abnormality. With consent of families and referring obstetricians, fetuses suspected of having significant heart disease were followed; discussions of possible pre-and postnatal therapies were held regularly. In all cases perinatal management was altered because of the prenatal evaluation. Five have required surgery (3 as neonates; 2<6 mos); 2 have not required intervention; 1 died unexpectedly on day 2. Four fetuses with suspected left-sided obstructive lesions on the basis of associated findings continued postnatally to have non-specific 2DE changes; in none, however, could coarctation be found.

F2DE is a powerful tool in prenatal assessment. When significant heart disease is suspected, appropriate interventions can be planned and the neonatal outcome for babies with life-threatening heart disease improved. Of concern is the risk of false positive F2DE diagnosis and its impact on all resources; thus, ongoing reassessment of diagnostic criteria by pediatric cardiology is critical.

●130 MECHANISM OF 2:1 ATRIOVENTRICULAR BLOCK IN INFANTS WITH CONGENITAL LONG QT SYNDROME. William A. Scott, M.D., Burt I. Bromberg, M.D., Macdonald Dick II, M.D. C.S. Mott Children's Hospital, University of Michigan, Division of Pediatric Cardiology, Ann Arbor, MI.

The mechanism of 2:1 atrioventricular block in infants with the congenital long QT syndrome has been postulated to result from a long ventricular effective refractory period interrupting conduction of successive sinus impulses. Three infants age 1-2 days exhibited bradycardia demonstrated to be 2:1 atrioventricular block by the surface electrocardiogram. Mean sinus cycle length (SCL) was 0.56 sec where as mean QT interval was 0.65 sec (QTc 0.63); this .07 sec difference between the SCL and mean QTc was sufficient to block successful capture of the ventricles by successive sinus impulses. Programmed ventricular extrastimulation in one patient demonstrated a markedly prolonged ventricular effective period (480 sec) at ventricular basic cycle length (BCL) 1000 msec shortening to 280 msec with a ventricular BCL of 400 msec. Permanent ventricular pacing shortened the ventricular effective refractory period in each infant and effectively suppressed polymorphic ventricular arrhythmias in the affected two. We conclude that the mechanism of 2:1 atrioventricular block in infants with the congenital long QT syndrome is a function of the age related SCL relative to the QT interval as well as the markedly prolonged but rate dependent ventricular effective refractory period. Ventricular pacemaker implant is effective in increasing ventricular rate, shortening ventricular effective refractory period, and suppressing complex ventricular arrhythmias.

131 ATRIAL FLUTTER IN INFANCY: NO NEED FOR CHRONIC PROPHYLAXIS. William A. Scott, Burt I. Bromberg, Macdonald Dick II, Catherine A. Webb. C.S. Mott Children's Hospital, University of Michigan, Division of Pediatric Cardiology, Ann Arbor, MI.

Many authorities recommend digoxin for one year for atrial flutter (AT.FL) in infants with normal structural hearts. To re-examine this recommendation the electrocardiogram, response to programmed extrastimulation, (PES) (n=3), and clinical course in six infants with AT.FL. alone were reviewed. AT.FL exhibited a regular sawtooth pattern with a cycle length of 148 msec (range: 130-160 msec). Age at diagnosis was 34 days (range: birth (n=4) to 180 days). There were two males and four females; all were initiallly treated with digoxin but only two converted to sinus rhythm. AT.FL resolved spontaneously in two, two were converted with atrial overdrive pacing. Post-conversion electrocardiogram demonstrated normal P wave axis, PR interval .14 sec (range: .12-.16 sec), P wave duration .08 sec, and P wave amplitude .23 mV (3 > .25 mV) at heart rate 142 bpm. Post-conversion atrial PES with burst pacing in three failed to initiate AT.FL and demonstrated normal atrial effective refractory periods (158 msec; range: 150-173 msec) and normal atrioventricular conduction system effective refractory periods (199 msec; range: 180-216 msec). Only two patients were maintained on digoxin, six months and one year after conversion. Follow-up was 5.5 years (1.5 - 18 years) without recurrence. These data suggest that AT.FL in infants without structural heart disease, following conversion, is self-limited and not easily inducible by extrastimulation. We conclude that infants with atrial flutter alone, following cardioversion, do not require chronic prophylaxis.

132 NORMAL VALUES OF MEAN AORTIC FLOW VELOCITY IN INFANTS AND CHILDREN MEASURED BY PULSED DOPPLER ECHOCARDIOGRAPHY.* Michael Seear, Luigi D'Orsogna, George Sandor, Ruby Popov, Elizabeth Patterson, Eustace DeSouza, Star Morris(Spon. by Donald E. Hill) University of British Columbia,Department of Pediatrics, Vancouver, B.C.

Traditionally, the estimation of cardiac output by pulsed Doppler echocardiography (PDE) involves the computation of mean aortic flow velocity(MAFV) and aortic cross-sectional area(CSA); as CSA varies with surface area(BSA), we postulated that MAFV should be relatively constant. However, data on normal values of MAFV in infants and children is lacking. Accordingly, 51 normal children aged one month to 15.1 years(m=5.6 years)were prospectively studied by PDE via the suprasternal view using a 3MHz transducer with fast Fourier transform spectral analysis of the doppler shift frequency in the ascending aorta. MAFV was calculated by averaging the planimetered area under the spectral display of three consecutive beats using the outer edge of the display as the defining border. Results showed that MAFV=27.8 + 5.2 cm/sec. This was relatively constant decreasing slightly with age and BSA;MAFV at one year=30.3 cm/sec. and 23.4 cm/sec. at 15 years. MAFV=30.381-0.039 age(R^2=0.123,p=0.05)and MAFV=32.051-5.368 BSA(R^2=0.097,p=0.05). Aortic root diameter(Ao.D.)progressively increased with age and BSA with Ao.D.=1.327+0.06 age(R^2=0.806,p<0.001,Ao.D.=0.968+0.956 BSA(R^2=0.844, p<0.001)respectively. We conclude that MAFV in infants and children remains relatively constant decreasing slightly with age and BSA. However Ao.D. increases more markedly with age and BSA,and,therefore accounts for the increase in cardiac output when measured by PDE. Whether MAFV alone may be used for monitoring cardiac output remains to be validated. *Supported by the B.C. Heart Foundation.