163 CHRONIC DIGOXIN ADMINISTRATION INHIBITS THE MYOCARDIAL NaK-PUMP IN LAMBS. Thomas J. Hougen, M. Andre Vasu, Carl Chipman, Edward J. Woodford, Gerald R. Marx, Harvard Medical School, Department of Cardiology, Children's Hospital, Boston, MA

The physiological and myocardial effects of digoxin in infants and children are debated and uncertain. In order to examine the response of the myocardial NaK-pump to chronic digoxin administration active transport of the K^+ analog Rb^+ was measured in vitro in serial right atrial biopsies in young lambs. Nine lambs (mean age 7d) were lightly sedated, instrumented, and ECG, aortic pressure, LV pressure and LVdP/dt were recorded. General anesthesia was then induced and a right atrial biopsy was obtained for Rb^+ active transport determination. This procedure was repeated 34d (mean) later after either no treatment (controls, n=4) or digoxin (n=5). Digoxin was given I.M. to maintain a plasma concentration of 1.3 ng/ml (mean) with no toxicity. The changes in hemodynamics between initial and final study were similar in both groups. No significant difference was observed in controls between initial and final Rb^+ active transport values (0.381 $\pm$ 0.015 ($\pm$ SEM) nmol Rb^+/mg wet wt/30 min vs 0.364 $\pm$ 0.023, paired t). In contrast, digoxin inhibited Rb^+ active transport when compared to initial values (0.349 $\pm$ 0.014 vs 0.246 $\pm$ 0.033, $P < 0.05$). Under these conditions the NaK-pump was inhibited indicating that chronic digoxin administration in therapeutic concentrations does have a myocardial effect in the developing mammalian heart.

164 LEFT VENTRICULAR SYSTOLIC TIME INTERVALS BY DOPPLER-COMPARISON WITH CAROTID PULSE METHOD Kai Hsieh, Steve Colan, Cynthia Holland, Debbie MacPherson, Stephen Sanders; Dept. of Cardiology, Harvard Medical School, Children's Hospital, Boston, MA 02115 (Spon. by Thomas J. Hougen)

We compared left ventricular systolic time intervals (L STI's) derived from pulsed Doppler(D) and from conventional(C) carotid pulse tracing(CPT) and phonocardiography(PCG) in 24 subjects, aged 1.7 to 18.5 yrs, median=12.1 yr. Using the apical 2-chamber view, D frequency-time curves from the proximal ascending aorta were recorded simultaneously with CPT,PCG and ECG. L STI's from the D tracings were measured as follows: pre-ejection period (PEP)-beginning of QRS to onset of D systolic upstroke; ventricular ejection time(VET)-onset of D systolic upstroke to maximal end-systolic flow reversal. The difference in pulse transmission time between onset and end of systole resulted in a systematically shorter PEP and longer VET by C. C PEP was best predicted by the regression equation C PEP=1.03(D PEP)-14, and C VET by C VET=0.99(D VET)+8. The cumulative distribution of the error of the predicted values and correlation coefficients(r) using these formulae were:

	C$\pm$5msec	C$\pm$10msec	r
Predicted C PEP	58%	100%	0.949
Predicted C VET	79%	100%	0.995

We conclude that D is a more direct and potentially more accurate non-invasive method for measuring L STI's. In addition, it is easily obtained in pediatric patients.

165 THE RENAL SPECTRUM OF PATIENTS WITH WILLIAMS SYNDROME Julie R. Ingelfinger, Jane Newburger, Dept. of Cardiology and Div. of Nephrology, Harvard Med. School and The Children's Hospital, Boston, MA.

We assessed the prevalence of renal abnormalities in children with Williams syndrome and heart disease. Of 32 patients (pts) followed by the Cardiology Service, 17 were male. Median age was 19 years (range 1.5-48 years). In 9 pts, limited information was available from chart review; in the remaining 23, renal structure and function were evaluated by urinalysis, BUN, creatinine and ultrasound or intravenous pyelography (IVP). Systemic arterial hypertension was present in 19 (59%) of the 32 pts. Among the 13 pts whose hypertension was severe enough to warrant renal arteriography, 8 (25% of all pts)were demonstrated to have renal artery stenosis. Albuminuria was present in 33% (8/24) of the pts in whom urinalysis was performed. Among the 23 pts who were studied with either IVP or renal ultrasound, 8 had structural abnormalities; 3 had a single kidney, 2 unusually small kidneys, 2 calcium-related nephrocalcinosis and end-stage renal disease, and 1 scarred kidneys secondary to reflux nephropathy. Pts with Williams syndrome had a relative risk of single kidney, which was at least 47 times that of the general population ($p<.0001$).

In toto, 16 pts had known renal abnormalities, constituting 70% of those who underwent renal study and 50% of all 32 pts (presumed lower limit of prevalence). Our data suggest that the prevalence of significant renal abnormalities in pts with Williams syndrome is high; screening and prospective follow-up for renal abnormalities should be performed in all pts with Williams syndrome.

166 EVALUATION OF PALLIATIVE SYSTEMIC-PULMONARY ARTERIAL SHUNTS USING NMR IMAGING. Mark D. Jacobstein, Barry D. Fletcher, Thomas A. Riemenschneider (Spon. by Jerome Liebman) Case Western Reserve University, Rainbow Babies and Childrens Hospital, Department of Pediatrics, Cleveland.

Palliative systemic-pulmonary artery (SP) shunts are frequently performed on children with cyanotic congenital heart defects which result in decreased pulmonary blood flow. Postoperative assessment of the adequacy of the shunt depends on physical examination, arterial blood gas analysis, chest x-ray and doppler-echocardiography. Direct visualization of the shunt, however, has required angiographic techniques at cardiac catheterization or, more recently, digital subtraction angiography. Nuclear magnetic resonance imaging (NMRI) is a new technique capable of providing high quality resolution of vascular structures without the use of contrast agents or exposure to ionizing radiation. We have used ECG-Gated NMRI at 0.3T to evaluate 6 patients with a total of 8 SP shunts including 4 Glenn shunts and 4 Blalock-Taussig (BT) shunts. 3 of 4 Glenn shunts and 3 of 4 BT shunts could be imaged. The size and course of the shunt could be seen in its entirety. 1 Glenn and 1 BT shunt, both patent, were not imaged. We conclude that NMRI is a safe, effective, non-invasive method for visualizing and evaluating patent palliative SP shunts. In our hands, NMRI has proved superior to echocardiography in visualizing these shunts.

167 EVALUATION OF AV CANAL DEFECTS USING ECG-GATED NMR IMAGING. Mark D. Jacobstein, Barry D. Fletcher, Thomas A. Riemenschneider (Spon. by Jerome Liebman) Case Western Reserve University, Rainbow Babies and Childrens Hospital, Department of Pediatrics, Cleveland.

Preoperative assessment of AV canal defects requires precise delineation of ventricular, septal and valvular morphology. In particular, recognition of ventricular hypoplasia is critical since it may preclude successful surgical correction. This evaluation is difficult even with cineangiography and 2-dimensional echocardiography. Nuclear magnetic resonance imaging (NMRI) is a new technique capable of providing excellent spatial and contrast resolution of cardiovascular structures without the need for contrast agents. We have used ECG-gated NMRI at 0.3T to evaluate 6 children with AV canal defects, including 3 with balanced chamber sizes, 2 with hypoplasia of the left ventricle and 1 hypoplastic right ventricle. All 6 defects were readily imaged and the 3 hypoplastic ventricles identified. AV valve morphology could be determined in 5 patients. Other frequently seen structures included papillary muscles, chordae tendinae and moderator bands. We conclude that gated NMRI is a valuable adjunct to angiography and echocardiography in the preoperative evaluation of children with AV canal defects.

168 EFFECT OF OUABAIN ON MYOCARDIAL MECHANICAL FUNCTION AND SODIUM PUMP IN THE PREMATURE NEWBORN RABBIT Jay M. Jarmakani, Toshio Nakanishi, Tatsuo Shimizu, and Shigeru Uemura, UCLA Medical Center, Dept. Peds, Los Angeles, CA.

Effects of ouabain (0.75, 1, 1.5, 2.5, or 5 x 10^{-6}M) on mechanical function and sodium pump were studied in the isolated arterially perfused fetal and newborn rabbit heart. Measurements of myocardial $^{86}Rb^+$ active uptake was used as a marker of the sodium pump activity. The inotropic effect of ouabain in the fetus was not significantly different from that in the newborn at all ouabain concentrations. 2.5 x 10^{-6}M ouabain caused mechanical toxicity in the fetus but not in the newborn. In the fetal muscle, perfusion with low calcium (0.5 mM) solution reduced mechanical toxicity of ouabain. High extracellular calcium (30 mM) per se caused mechanical toxicity in the fetus, but not in the newborn. Ouabain infusion decreased tissue potassium content, and this decrease in the fetus was not significantly different from the newborn. The ouabain inhibition of Rb^+ uptake was similar in the two age groups. These data indicate that mechanical toxicity in the fetus is observed at lower ouabain concentrations than in the newborn. This difference in mechanical toxicity may not be explained by the age-related difference in sodium pump activity. The greater calcium toxicity in the fetus may be the reason for the increased ouabain toxicity in this age group.