133 DIGITIZED ECHOCARDIOGRAPHIC ASSESSMENT OF LEFT VENTRICULAR DYSFUNCTION IN CHILDREN AND ADOLESCENTS WITH SICKLE CELL ANEMIA. Frederick W. Arensman, Welsey Covitz, Carol L. Boe, Carol A. Jones, Lynn K. Reyes, Saadeh B. Jureidini. (Spon by Albert Pruitt) Medical College of Georgia, Department of Pediatrics, Augusta, GA

To examine systolic and diastolic left ventricular function in sickle cell anemia (SS), 70 SS patients (pts) and 45 age-matched controls underwent routine M-mode and computer assisted echocardiographic analysis. Shortening fraction (SF) and LV systolic time intervals (LVSTI's) were calculated in the routine manner, and the following variables were derived by computer: R-R interval, max (Max) and minimum (Min) LV cavity size, Max and Min septal and freewall thickness, Max normalized rates of LV filling and emptying and Max normalized rates of septal and freewall thickening and thinning. Timing of each Max event was normalized for RR interval.

Compared to controls, SS patients had no significant differences in SF or LVSTI's. SS LV cavity size was significantly increased in systole ($p<0.001$) and diastole ($p<0.001$) and R-R intervals were shorter ($p<0.001$). Systolic abnormalities included thicker septal thickness ($p<0.001$) and freewall thickness ($p=0.002$) and delays in achieving Max septal thickness ($p=0.001$) resulting in delayed Min cavity size ($p<0.001$). In diastole Max rate of septal thinning was delayed ($p<0.001$) and accompanied delay in Max rate of filling ($p<0.001$).

Digitized echocardiographic assessment of LV function in sickle cell anemia reveals both systolic and diastolic dysfunction previously undetected by routine echo analysis.

Sponsoring member's name and signature

134 HEMODYNAMIC EFFECTS OF HYDRALAZINE (Hz) IN INFANTS WITH IDIOPATHIC DILATED CARDIOMYOPATHY (CM) Michael Artman, Mark Parrish, Scott Appleton, Robert Boucek, Thomas P. Graham, Vanderbilt Univ., Nashville, TN.

Experience with vasodilator therapy for CM has been limited primarily to adults. Thus, we evaluated the effects of Hz (0.5-1.0 mg/kg IV) during cardiac cath in 13 infants with CM. Ages were 2 to 13 (mean=6.7)months. Each infant had congestive heart failure (CHF) and angiographic evidence of markedly depressed LV ejection fraction (0.24±0.11; mean±1S.D.) with LV dilation (LV end-diastolic volume = 349±125% of normal). Hemodynamic measurements obtained immediately prior to Hz were compared to those taken 30 min after Hz. Results for mean arterial pressure (MAP;mmHg), systemic arteriolar resistance(Rs;units/M^2),stroke volume index (SVI;ml/beat/M^2)and cardiac index(CI;l/min/M^2;by thermodilution)are tabulated below:

	MAP	Rs	SVI	CI
Pre-Hz	70±8	21.1±3.3	26.9±4.9	3.24±0.53
Post-Hz	60±11*	12.0±2.7*	34.5±7.5*	4.71±0.99*

*=post-Hz different from pre-Hz value ($p<0.001$)

The 41±14% decrease in Rs was accompanied by a 45±16% increase in CI. Hz increased heart rate(122±19 to 138±18;$p<0.001$) but this change did not account entirely for the rise in CI, as evidenced by the increase in SVI. Hz also reduced pulmonary resistance and wedge pressure. Oral Hz has been included in the treatment regimen of 10 infants followed for 1 to 38(mean=11) months. Of these, 9 have sustained clinical improvement. We conclude that Hz can be a beneficial adjunct to the management of CHF in young infants with idiopathic dilated cardiomyopathy.

135 QUANTITATIVE TWO-DIMENSIONAL (2D) ECHO STUDY OF LEFT VENTRICULAR (LV) SHAPE IN NORMAL NEWBORNS (NB) - EARLY (EC) VS LATE (LC) UMBILICAL CORD CLAMPING Annabelle Azancot, Thomas Caudell, J. Crequat, G. Toscani, J.H. Ravina, Hugh D. Allen, Hôpital Bretonneau-Bichat, INSERM 120, Paris, France, & Univ. of Arizona Health Sciences Ctr, Tucson

Using a computerized method which quantitatively analyzes overall LV shape and distortion, we recently reported a study in which fetuses and NB had flattening of the septal portion of the LV which tended toward roundness during infancy. Our present study evaluates effects of EC (≤5 sec; n=6), and late (LC)(3 min; n=7) cord clamping on LV shape in NB. Vaginal delivery was accomplished with all infants kept at the same level. NB were studied serially at <.5 hr and at 10-48 hrs. Mean gestational age (EC=39.5 ±.37 wks; LC=39.6±.55) (mean±SD), weight (EC=3.22 ±.3 kg; LC=3.21±.40), and Apgar scores (>8 EC and LC)(all p=ns) were recorded. Mean venous hematocrit for EC was 47±.03%, and for LC was 59±5% ($p<.02$). Diastolic (d) and systolic (s) 2D standard short axis frames were evaluated. Shape factor (SF) from this method for a circle = 1; for an indented ellipse = 7. Serial LC diastolic septal LV shape distortion was significantly greater than for EC (SFd LC at .5 hour = 5.08±.86 vs SFd EC at .5H=2.91 ±.68, $p<.001$; SFd LC 10-48 hour = 4.25±.65 vs SFd EC 2.45±.48, $p<.01$). In systole, LV shapes were all rounder, but SFs in LC was significantly higher than in EC at <.5 hour (3.15 ±.45 vs 2.15±.43, $p<.05$). Systolic SF data were similar at 10-48 hours (EC=2.15 ±.40; LC=2.3±.48). Placental transfusion significantly alters and distorts LV shape.

136 PREOPERATIVE AND POSTOPERATIVE EXERCISE RESPONSE IN PULMONARY ATRESIA. Gerald Barber, Charles T. Heise, Dwight C. McGoon, Gordon K. Danielson, Francisco J. Puga, David J. Driscoll, Mayo Clinic, Dept. of Ped., Rochester, MN

Patients (PTS) with pulmonary atresia (PA) subjectively may have increased exercise (EX) tolerance after 1st stage (RV to Pul. Art. conduit placement without VSD closure) or 2nd stage (RV to Pul. Art. conduit and VSD closure) repair. To measure precisely the degree of EX intolerance in PTS with PA and to assess improvement of EX tolerance after operation 10 PTS preop, 4 PTS after 1st stage and 7 PTS after 2nd stage repair performed graded cycle ergometry. The groups were similar in sex, age, ht. and BSA. As percent predicted, total work performed ($P< 0.005$), maximal power ($P<0.01$) and EX time ($P<0.001$) increased following either 1st or 2nd stage repair. EX performance was similar for PTS after 1st and 2nd stage repair. Resting and exercise blood pressures were similar among the three groups. As percent predicted, resting heart rate (HR) was lower ($P<0.02$) after 1st or 2nd stage repair than preop. Maximum EX HR was highest after 2nd stage repair ($P<0.05$). Both resting and EX oxygen saturation increased with degree of repair ($P<0.05$). Resting respiratory rate, minute ventilation ($\dot{V}e$), oxygen uptake ($\dot{V}O_2$) and $\dot{V}e/\dot{V}O_2$ were similar among the groups. EX $\dot{V}e$ and $\dot{V}O_2$ were greater ($P<0.001$ and $\dot{V}e/\dot{V}O_2$ less ($P<0.001$) for the two postop groups than the preop group.

Graded cycle ergometry is a useful technique to evaluate the impact of surgery on congenital heart disease. By it we demonstrated that both 1st and 2nd stage repair of PA improves cardiorespiratory response to exercise and exercise tolerance.

137 EFFECT OF HYPERGLYCEMIA ON MYOCARDIAL FUNCTION AS WELL AS RED BLOOD CELL PARAMETERS IN THE FETAL LAMB. Harry Bard, Jean-Claude Fouron and Xavier De Muylder, Un.of Montreal, St. Justine's Hosp., Dept. Ped., Montreal,Canada.

In order to evaluate the effect of a moderate fetal hyperglycemia on myocardial as well as red blood cell function, six fetal lamb preparations were made hyperglycemic after appropriate catheter insertions between the 120th and 130th day of gestation. A micro-tip transducer catheter was placed above the fetal aortic valve allowing recording of an acurate pressure curve on which systolic time intervals were measured. Fetal hyperglycemia was obtained by a continuous glucose infusion which was maintained between 2-7 days. Hemoglobin, hematocrit, reticulocyte count, P_{50} and 2,3-DPG were determined. The mean values showed the following significant changes ($p<0.05$): glucose increased from 15 to 51 mg%, O_2 content dropped 25%; myocardial dysfunction was demonstrated by a prolongation of the pre-ejection period (PEP) (40.7 to 51.6 msec.) and changes in the PEP to ejection time ratio (0.25 to 0.33). The cardiac rythm, arterial pressure and pH remained unchanged. There was a significant correlation between the increase in PEP and the level of glycemia. This increase in PEP was not related to O_2 content. Fetal hyperglycemia also caused reticulocytosis (0.4 to 3.5%) an increase in hemoglobin level (10.3 to 11.3 mg%) and a significant drop in 2-3 DPG level (5.05 to 3.05 µmol/gHb). This study demonstrates that fetal hyperglycemia effects myocardial function and causes an erythropoietic expansion. It also results in a drop of 2,3-DPG without a demonstrable effect on hemoglobin oxygen affinity.

138 SURGICAL REPAIR OF UNIVENTRICULAR HEART WITH OUTLET FORAMEN OBSTRUCTION. Gerald Barber, Dwight C. McGoon, Gordon K. Danielson, Francisco J. Puga, David J. Driscoll, Mayo Clinic, Dept. of Pediatrics, Rochester, MN

Patients (PTS) with univentricular heart (UH) and outlet foramen (OF) obstruction have a greater surgical mortality than PTS with UH without OF obstruction. To determine the exact mortality and to attempt to delineate predictors of outcome we examined the records of all PTS with UH and OF obstruction operated upon at the Mayo Clinic from 1973 to 1983.

Of the 17 PTS identified, 12 had a modified Fontan procedure, 3 ventricular septation, 1 resection of the OF with a side to side aorta (AO) to main pulmonary artery (PA) anastomosis, and 1 transection of the main PA with a proximal PA to AO anastomosis and an AO to distal PA Gortex graft. Ten of the 17 PTS died within 72 hrs of operation. Mean age at operation was significantly greater ($P<0.001$) among survivors (14.3±2.0 yrs) than nonsurvivors (6.5±4.2 yrs). There was no difference in postoperative gradient across the OF, aortic crossclamp time or bypass time between the two groups. Survivors had a significantly lower gradient ($P<0.01$) across the OF measured at preop cath (39.6±19.9) than survivors (83.9±35.7). No PT with a gradient greater than 66 mm Hg survived. Survivors had significantly smaller ($P<0.005$) preoperative CT rations (0151±0.07) than nonvurvivors (0.61±0.05). No PT with more than 3 mm of J-point depression in any ECG lead survived.

UH with OF obstruction is a rare condition with a high surgical mortality. Mortality is related to age at operation and degree of OF obstruction, cardiomegaly and J-point depression.