199 CARDIAC STATUS OF CHILDREN WITH SICKLE CELL ANEMIA (SS) Peter C. Sodt, Geeta N. Dalal, Lucille A. Lester, John W. Moohr, Rene A. Arcilla; Pritzker School of Medicine, The University of Chicago Hospitals and Clinics, Department of Pediatrics, Chicago, Illinois

Echocardiograms (E) were obtained from 44 children with SS and from 54 normal Blacks (Nl). Equations for predicting the normal values (PN) for 12 echo parameters were derived ($PN = A \cdot BSA^b \cdot HR^c \pm K$), and the observed values from SS then expressed as standard deviation from normal (σ PN). Increased cardiac output, LVEDD, LVESD, wall thickness, LA_{max} and Ao diameter were observed in SS ($p < 0.001$); however, the functional parameters %MAS, VCF and PEP/ET did not differ from Nl. Density function curves (DFC), derived by discriminant regression analysis using 7 echo parameters were obtained with equation: $Y_{dv} = K_1 \sigma PN_1 + K_2 \sigma PN_2 + Kn \sigma PN_n \pm X$, where Y_{dv} = discriminating value and K, X = constants. There was clear separation of the DFC of SS and Nl. Eleven children with SS underwent hypertransfusion therapy (HT) for 3/12-3 yrs (packed RBC q 2-3 weeks) to achieve Hgb $\geq$ 10 gm %, % S Hgb < 20%. E were obtained periodically before/after, and during HT. Mean values during HT/off HT were: Hgb 9.7/7.2 ($p < .001$), % S Hgb 12.1/88.2 ($p < .001$). The DFC and Y_{dv} before and after HT did not differ from those of 44 untreated SS ($P = NS$); during HT, there was significant shift ($p < 0.001$) of DFC and Y_{dv} towards Nl. We conclude that cardiac size and function are abnormal in SS children, improve during HT, but revert back to abnormal state after discontinuing the HT.

200 THE OPTIMAL AGE FOR ELECTIVE REPAIR OF COARCTATION OF THE AORTA. Henry M. Sondheimer, Mark S. Rosenbloom, Fredric A. Hoffer, Marie S. Blackman, Frederick B. Parker (Spon. by Frank A. Oski), Department of Pediatrics, SUNY-Upstate Medical Center, Syracuse.

We reviewed all 108 children undergoing emergency and elective repair of coarctation of the aorta over a 13 year period in our institution. No deaths occurred in 60 children having surgery over one year of age although there was a 25% mortality rate (12/48) for those under one. Blood pressures were checked regularly and survivors were judged to be in one of three categories: 1. Persistent Coarctation (diminished femoral pulses and/or a 20 mm. Hg. arm to leg pressure difference present from the first post-op visit onward); 2. Recoarctation (same as persistent coarctation but appearing sometime after the first post-op visit); or 3. Good Result (less than a 20 mm. Hg. pressure difference with good femoral pulses).

Patients were then grouped by the age at surgery (under 1, 1 to 6, 7 to 10, 11 plus). Patients who had surgery between 7 and 10 years of age had the best long term results. In this group 2/28 patients had persistent coarctation but 26/28 had good results as defined above. In addition, final arm systolic blood pressure was below the 90th percentile for age and sex in all 26 of these patients, so there was no persistent paradoxical hypertension. Persistent or recoarctation was seen in 40% of children operated on between 1 and 6 and 18% of those having surgery past ten years of age. Our data suggests that 7 to 10 years is the optimal age for elective repair of coarctation of the aorta.

● 201 ALTERATIONS IN CARDIAC SARCOPLASMIC RETICULUM FUNCTION IN A CONGESTIVE CARDIOMYOPATHY MODEL Nancy A. Staley, George R. Noren, and Stanley Einzig University of Minnesota Medical School, Veterans Administration Medical Center, Department of Pathology, Minneapolis, Minnesota

In a naturally occurring model of congestive cardiomyopathy (CCM) of turkeys, alterations of Ca++ transport in isolated cardiac sarcoplasmic reticulum (SR) were documented in the young CCM turkey at 1, 10, and 28 days of age; prior to the development of cardiac dilatation. Ca++ binding in CCM birds was reduced to 55-75% of values measured in age-matched commercial control turkeys ($p<0.05$ to <0.01) and Ca++ uptake was reduced to 52-87% of control values ($p<0.05$ to <0.01). Ca++ stimulated ATPase values at 1, 10, and 28 days of age were similar in CCM and control turkeys. However, at 56 days of age when all CCM birds showed marked to moderate left ventricular dilatation, Ca++ stimulated ATPase was reduced to 75% of control values ($p<0.05$). Cyclophosphamide immunosuppression of newly hatched CCM turkeys significantly elevated Ca++ binding and Ca++ uptake in isolated cardiac SR from 10-day-old birds by 73% and 58% respectively ($p<0.05$ and <0.01) over values measured in untreated CCM birds. These results were not significantly different from values obtained in 10-day-old control turkeys. In summary this unique depression of SR Ca++ uptake and Ca++ binding, prior to the onset of cardiac dilatation, and which is reversed by immunosuppression, provides an experimental link between this animal model and a proposed mechanism for the pathogenesis of congestive cardiomyopathy in man.

202 THE CHANGING SPECTRUM OF PEDIATRIC INFECTIVE ENDOCARDITIS (IE): AN ANALYSIS OF 26 CASES AT YALE-NEW HAVEN HOSPITAL, 1970-1979. B. F. Stanton, R. S. Baltimore and J. D. Clemens. Yale Univ. Sch. of Med., Dept. Ped. and Med., New Haven, CT.

The last decade has been marked by an increased survival of children with complex cyanotic heart lesions, increasing employment of open heart surgery and surgery requiring the insertion of prosthetic material, and an increased utilization of indwelling central catheters in the critically ill patient. To investigate whether these and other factors have contributed to a change in the natural history of IE in children we analyzed all 26 cases of pediatric IE seen at Yale 1970 through 1979.

Four important differences from previous series were noted. 1) A larger proportion of cases arose post-operatively, 14/26 (54%) of which 11 were $\leq$ 6 mos. post-op. (2) Staphylococci were not the predominant pathogens among post-surgical or catheter-related cases (3/13). Other pathogens were alpha Strep ·4, gr D Strep., Enterobacter, Klebsiella and Serratia. (3) IE associated with prosthetic material carried an unexpectedly favorable survival, 86% (6/7) compared with an 81% survival for the entire group. Moreover, all survivors of IE with prosthetic material were cured by medical regimens alone. (4) All episodes of IE occurring in previously normal hearts occurred in patients with indwelling central catheters and also carried the worst prognosis in this series (mortality 2/3).

We conclude that pediatric IE continues to be an evolving disease and that this is a reflection of new approaches to children with complex congenital heart disease and of acute care of critically ill children.

203 NORMOTENSIVE RESPONSE TO MULTISTAGE TREADMILL EXERCISE IN POSTOP COARCTATION PATIENTS John P. Thomas, William J. Gallen (sponsor Frederick M. Blodgett) Medical College of Wisconsin, Milwaukee Children's Hospital, Department of Pediatrics, Milwaukee Wis.

Persistent systolic hypertension and differential gradient after successful coarctectomy have been noted during stress exercise. We evaluated 9 patients 8 to 17 years of age with a modified multistage Balke treadmill protocol. Each patient was normatensive at rest without evidence of persistent gradient, and was clinically asymptomatic 6 months to 9 years post resection or patch angioplasty. The patients were stressed to physical exhaustion at a speed of 3-3.4 mph with a grade increase of 2.5° every two minutes. The patients were observed ten minutes during recovery. Heart rate, blood pressure and 12 lead ECG were recorded at rest and every two minutes during exercise and recovery. Simultaneous blood pressure recordings of left leg and right arm were obtained at rest and at two to four minutes into recovery phase. Duration of exercise was 12-20 minutes. Maximal heart rate was 80-95% of predicted. Systolic pressure increased 35-65 mm in the right arm and when simultaneous pressures were recorded during recovery at two to four minutes, the systolic leg pressure was 10-20 mm higher. No arrhythmias or significant ST or T wave changes were noted during exercise or recovery. In summary, a group of post coarctation patients who are normatensive, fail to demonstrate a blood pressure gradient and have insignificant associated defects, had a normal response to stress exercise.

● 204 SYSTEMIC CARNITINE DEFICIENCY PRESENTING AS FAMILIAL ENDOCARDIAL FIBROELASTOSIS (EFE) Marjorie E. Tripp, Murray L. Katcher, Henry A. Peters, and Austin L. Shug, (Spon. by Gerard Odell), Depts. of Pediatrics, Neurology, Nutritional Sciences, University of Wisconsin, Madison.

Familial occurrence of EFE suggests an inborn error of myocardial metabolism. We studied a family in which 4 of 5 sibs had EFE, apparently due to systemic carnitine deficiency (SCD). 3 of 5 died suddenly before age 3 with EFE at autopsy. Abnormal mitochondria were found in skeletal and cardiac muscle of the 3rd. The surviving 4th sib with EFE, without muscle weakness had a plasma carnitine level (PCL) of 4.8mcM/L (Nl, 50±10), skeletal muscle carnitine level (MCL) of 30nM/gm (Nl, 5000). On oral L-carnitine, (L-carnitine supplied by Sigma-Tau, Rome, Italy) PCL rose to 43.7mcM/L, exercise tolerance improved, mitral insufficiency resolved, ST-T changes partially reversed. Retrospective study of the 3rd sib's tissue found a serum CL of 4.5mcM/L, MCL of 30.9nM/gm, cardiac CL of 56.8nM/gm (Nl, 5000), liver CL of 120.7 nM/gm (Nl, 400). A 5th sib and the parents have low Nl PCL.

Cardiac metabolism is almost entirely aerobic, with mitochondrial oxidation of fatty acids providing over 65% of energy supply. Carnitine is required for fatty acid transport into mitochondria. Deficiency causes fatty deposits in cytoplasm and depressed mitochondrial function. EFE may result from lowered cardiac mitochondrial activity with inadequate oxidative phosphorylation causing poor myocardial contractility, compensatory dilatation and hypertrophy, subendocardial ischemia and fibrosis. PCL and muscle biopsy are indicated in EFE to exclude SCD, a treatable cause of cardiomyopathy.