judged particularly unfavorable with annual mortality rates reported up to 6%. However, prior studies were confined to pts referred to tertiary centers, selectively focused on high-risk assessment. To analyze natural history independent of such selection bias, we studied a cohort of 25 children identified with HCM from a community-based practice since 1972. Initially ages were 0.5–18 yrs (mean 6); 15 (60%) were male. Eight pts (32%) had basal outflow obstruction with gradients 35–125 mm Hg; LV wall thicknesses were substantial, 10–40 mm (mean 22), with a variety of patterns of left ventricular hypertrophy (LVH) evident. During follow-up (1–21 yrs; mean 8), 4 pts died suddenly, aged 7–19 yrs including 1 each after myotomy-myectomy or defibrillator implant. Each of the 4 pts with sudden death had massive LVH (wall thicknesses 40, 38, 31, 25 mm at ages 7, 7, 19, 10 years). Annual mortality rate was 2.0%; survival was 90% at 5 yrs. Eight surviving pts developed limiting symptoms, including 4 who underwent myotomy-myectomy. In conclusion: 1) HCM in childhood may be associated with substantial morbidity/mortality; 2) risk for youthful sudden death was substantially lower in this unselected population representative of the true disease state, than reported from referral centers; 3) massive LVH may be an independent risk factor for sudden death in young children with HCM, suggesting the possibility of aggressive management for this patient subgroup.

4:15

767-2 Timing of Intervention for Restrictive Cardiomyopathy in Children

Rachel J. Weller, Linda J. Addonizio, Maryanne R. Kichuk, Welton M. Gersony, Daphne T. Hsu. *Columbia University, New York, NY*

Restrictive cardiomyopathy (RCM) is a rare entity characterized primarily by diastolic dysfunction leading to heart failure (HF). The timing of cardiac transplantation (CT) in children with end-2tage RCM has not been well defined. The mode of presentation, clinical course and outcome of 13 children (9 girls, 4 boys) with RCM referred for CT were reviewed. Mean age at diagnosis was 4.6 yrs. (range 0.1–16 yrs.). The presenting symptom was HF in 4 pts, irregular heart rate in 3 pts, heart murmur in 3 pts, dyspnea in 2 pts, and syncope in 1 pt. All pts had evidence of diastolic dysfunction at presentation; 11/13 children had preserved systolic function and 2 pts had poor systolic function. Significant atrial tachyarrhythmias occurred in 4 pts (31%); 2 of these pts also developed complete heart block requiring pacemakers. Major embolic events occurred in 2 pts (15%): stroke in 1 pt, pulmonary embolus in 1 pt. One pt developed a protein-losing enteropathy. Two pts had unremitting HF from presentation; severe HF developed in the remaining 11 pts 3.3 yrs (range 0.3–8.5 yrs) following presentation. Cardiac catheterization performed at CT referral (2.8 ± 2.4 yrs after presentation) demonstrated high filling pressures: LVEDP: 30 ± 9 mmHg; RVEDP: 16 ± 6 mmHg. The mean cardiac index was 2.2 ± 0.38 L/min/m2. The mean PA pressure was 50 ± 20 mmHg with a mean pulmonary vascular resistance index (PVRI) of 10.8 (range 1.8–30). In 9 pts PVRI was >6 U/m2. The mean time to transplant or death in 12/13 pts was 7.3 ± 5 months after onset of HF. Four pts died of HF; 3 of these were not candidates for CT due to extremely elevated PVRI. CT was performed in 8 pts with 2 operative deaths from pulmonary hypertension and donor right HF. In summary, HF developed late in 11/13 (85%) children with RCM, but progressed rapidly. Severe pulmonary hypertension was present in 9/13 pts (69%) at the time of cardiac transplant referral and precluded successful transplant in 5. Cardiac catheterization should be performed early in children with RCM to evaluate pulmonary vascular resistance and optimize the timing of transplantation.

4:30

767-3 Pediatric Dilated Cardiomyopathy (DCM): Prognosis in a Developing Nation Is Comparable to Developed Nations

K. Kumar, D. Thatai, A. Saxena, V. Srinivas, R. Juneja, S.S. Kothari, S. Shrivastava, S.E. Lipshultz. *Children's Hospital, Boston, MA; All India Institute of Medical Sciences, New Delhi, India*

Mortality from pediatric DCM is 33–67% 1–5 yrs after presentation in studies from the US and UK. A Turkish study found 10% mortality over 2 yrs of follow-up of 110 children with DCM. We determined prognosis of pediatric DCM in northern India where, compared to the US, 1) pediatric intensive care was more limited (e.g., 0.02 vs 1.84 PICU beds/100,000 population in northern India vs eastern MA), 2) PICU care was not as sophisticated, and 3) the average cost of admission for this diagnosis was less (usually < $200 US). 128 consecutive children with DCM (77 males, 51 females, median age, 24 mos, mean age, 46 mos, range, 2–180 mos) were studied. Multivariate and survival analyses were performed on the 97 children with follow-up (mean, 23 mos, range, 1–96 mos) and included sex, age at onset, delay after onset of symptoms, viral illness, CHF, ST-T changes, LVH, cardiothoracic ratio (CTR), LVEF, LA/AO, LV thickness/dimension. At presentation, CHF was noted in 82% of pts and the mean LVEF was 0.41 ± 0.15. 52 pts (54%) improved on therapy and of these, 23 (24%) became symptom-free without medication. 34 pts worsened despite therapy, and 18 (19%) of these died. Deaths occurred within 3 yrs of follow-up. Multivariate analysis showed female sex (RR: 2.15; 95% CI: 1.07, 2.87), cardiomegaly on presenting CXR (RR for CTR > 0.68: 2.24, 95% CI: 1.04, 4.85), and LA enlargement at presentation (RR for LA/AO > 1.7: 2.02, 95% CI: 1.00, 4.22) as the variables associated with poor outcome (death/worsening). Female sex was the only significant univariate and multivariate predictor of death. *Conclusions:* The prognosis of pediatric DCM in northern India is at least comparable to those reported for pediatric DCM in developed nations. The prognosis of pediatric DCM may not improve without further understanding of its etiologies and the development of etiology-specific therapies. The developing female myocardium may be more susceptible to damage than the male myocardium.

4:45

767-4 Survival and Risk Factors for Death After Cardiac Transplantation in Infants: A Multi-Institutional Study

Charles Canter, David Naftel, Randall Caldwell, Richard Chinnock, Elfrieda Pahl, Elizabeth Frazier, James Kirklin, Mark Boucek, Robert Morrow and the Pediatric Heart Transplant Study Group. *Univ. of Alabama at Birmingham, B'ham, AL 35294*

Despite the increasing application of cardiac transplantation (C Tx) in infants, reported survival rates vary and risk factors for death are poorly understood. In order to examine early survival and risk factors for death in infants (<1 yr of age) undergoing C Tx, 142 infants (36 < 1 mo) underwent primary C Tx from 1/1/93 to 1/1/95 at 21 centers in the Pediatric Heart Transplant Study (PHTS). Diagnoses were hypoplastic left heart (66%), other congenital heart disease (17%), cardiomyopathy (14%) and other (3%).

Results: Actuarial survival after C Tx was 84% at 1 mo, 70% at 1 yr, and 69% at 2 yrs, with the greatest hazard for death in the first 3 mos. The principle cause of death was early graft failure in 20 pts (52% of deaths); infection in 10 (26% of deaths), and rejection in 4 (10%). The likelihood of death from early graft failure (or isolated right ventricular failure) was 13% at 30 days. By multivariable analysis, risk factors for early mortality were history of previous sternotomy (p = 0.0003), recipient non-blood group A (p = 0.02), non-identical blood donor (p = 0.01), and donor cause of death other than closed head trauma (p = 0.04). Survival was favorable in identical blood groups with no previous sternotomy (80% at 1 yr). Diagnosis at listing, waiting time (mean waiting time 1.3 mo), graft ischemic time (range 68 to 479 minutes, mean 228), and recipient inotropic support at listing were not predictive for mortality post C Tx.

Inferences:

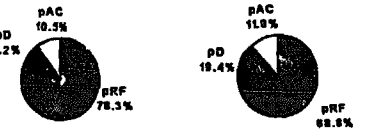
- In the current era, strategies to improve donor heart function at implantation would have the greatest impact on survival after infant C Tx.
- If early mortality from acute graft failure could be reduced by 75%, anticipated 1 yr survival would approach 80%.

5:00

767-5 Left Ventricular Filling Patterns Predict Acute Rejection in Pediatric Heart Transplant Patients

Douglas C. Semler, Thomas R. Kimball, Sandra A. Witt. *Children's Hospital Medical Center, Cincinnati, Ohio*

Transplant rejection may effect LV diastolic function. Automatic border detection is a new echo technique which continuously tracks the endocardium, allowing global assessment of LV filling. The aim of this study was to determine if changes in LV filling predict rejection in pediatric transplant patients. Twelve patients (2–18 y.o.) had echoes at 2 study periods: 1) while healthy without rejection and 2) at biopsy-proven rejection. The percent of total LV filling due to rapid filling (pRF), diastasis (pD) and atrial contraction (pAC) were measured by automatic border detection from a parasternal transverse view. *Results:* The percent of total LV filling contributed by each diastolic phase is presented in the pie charts below:



Pre-rejection ***Rejection***

pRF decreased (p < 0.10) and pD increased significantly (p < 0.05) during rejection. pAC and heart rate did not change.

We speculate that during rejection, inflammation increases LV stiffness