Outcome Predictors in Pediatric Hypertrophic Cardiomyopathy

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Background: In pediatric hypertrophic cardiomyopathy (HCM), there is limited understanding of patient factors measured at the time of diagnosis that affect the subsequent risk of death or heart transplant.
Methods: The NHLBI Pediatric CM Registry collected longitudinal data on 882 children with HCM from 1990-2003 (781 HCM, 57 HCM-dilated CM [DCM], & 44 HCM-restrictive CM [RCM] or other CM). The combined outcome of death or heart transplant occurred for 16.4% of HCM pts, 40.4% of HCM-DCM pts, & 40.9% of HCM-RCM or other CM pts.
Results: Of the HCM pts, 2-year event-free survival was significantly higher for idiopathic HCM presenting at >1 yr of age [mean age 10.5 yrs] (96%), than for idiopathic HCM presenting at ≤1 yr of age [mean age 2.6 mo] (77%), HCM with a malformation syndrome (75%), HCM with RCM or other CM (61%), HCM with DCM (55%), or HCM with an inborn error of metabolism (39%). Univariate Cox regression analyses showed no significant differences in outcomes by race/ethnicity, gender, geographic region, insurance status, height, or LVED dimension. Significant associations with adverse outcomes were noted when HCM was diagnosed with CHF (3-fold higher risk); lower weight (17% increased risk/1 SD decrease); lower body-surface area; lower LV fractional shortening (7% increased risk/1 SD decrease); higher LV mass (12% increased risk/1 SD increase); higher LVED posterior wall thickness (16% increased risk/1 SD increase); higher LVED septal wall thickness (11% increased risk/1 SD increase); and larger LVES dimension (5% increased risk/1 SD increase).
Conclusions: Prognosis is worse when pediatric HCM presents with an inborn error of metabolism or in combination with other types of CM compared with idiopathic HCM presenting at ≥1 yr of age. CHF, lower weight, BSA, or LV fractional shortening, or greater LV mass, posterior or septal wall thickness, or dimension at presentation were associated with a greater risk of subsequent death or heart transplant.