**IMPACT OF CONTEMPORARY TREATMENT STRATEGIES ON PATIENTS WITH EXTREME HYPERTROPHIC CARDIOMYOPATHY PHENOTYPE**

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**Objectives:**  To determine effect of treatment on outcomes and prognosis for patients with massive left ventricular hypertrophy (LVH; ≥30mm), a high-risk subgroup of hypertrophic cardiomyopathy (HCM).

**Background:**  Massive LVH has long been thought to be a high-risk subgroup in HCM, but the impact of treatment on improving outcomes of these patients have been unresolved.

**Methods:**  Of 1700 consecutive patients who presented to the Tufts HCM Institute from 2004 to 2016, 92 were identified with massive LVH and followed for 5 ± 3 years from initial visit; 12 ± 9 years from diagnosis.

**Results:**  Of the 92 study patients, 71% were male, age at diagnosis was 26 ± 13 years, maximal wall thickness was 32 ± 3 mm. Within the study period, 88 of 92 patients (96%) have survived and 4 (4%) died. All-cause mortality rate was 0.8%/year. Death was attributable to HCM in 2 of these patients (2%, 0.4%/year). Of the 88 surviving patients, 42 (48%) had a stable course, remaining in NYHA classes I/II. The remaining 46 patients required major treatment interventions including 35 with myectomy for progressive heart failure (NYHA class III/IV) secondary to outflow obstruction, 1 without obstruction considered for transplant, and 16 with arrhythmic sudden death events terminated by primary prevention appropriate ICD interventions (n=11) or out-of-hospital external defibrillation (n=5). HCM events aborted by major treatment interventions (including appropriate ICD shocks, resuscitated cardiac arrests, and transplant; 3.3%/year), exceeded HCM mortality (0.4%/year) by 8 fold (p<0.001). Survival at 10 years was 92%, and no different than that expected for age and sex-matched general US population (p=0.62).

**Conclusions:**  Patients with massive LVH represent a small but important subset of HCM patients at considerable risk for arrhythmic sudden death, preventable with ICDs, and progressive heart failure predominantly due to obstruction and treated with myectomy. Despite expressing the most extreme HCM phenotype, these patients can nevertheless achieve extended survival and good quality of life.