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Echocardiographic and electrocardiographic parameters as predictors of a worse outcome in children with hypertrophic cardiomyopathy

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Background: The clinical presentation and natural history in children with HCM is heterogeneous, ranging from asymptomatic forms to malignant expressions that may result in sudden cardiac (SCD) or heart failure-related death. The aim of study was to assess the prognostic value of echocardiographic parameters in addition to clinical and electrocardiographic characteristics in children with HCM.
Methods: Retrospective analysis of 88 pts, mean age 10,6yrs with HCM diagnosed from 1991 to 2010. Mean follow-up was 6,9yrs. All 88 pts were divided into two groups: gI- 31(35%) children with an unfavorable course of HCM [death n=6, CA n=4, 5pts were qualified for HTx (performed in 1pt, 1pt is waiting for HTx, 3pts died on waiting list), 9pts required surgical myectomy, in 12pts ICD was implanted (4 pts as a secondary, 8 pts as a primary prevention)] and gII- 57pts(65%) who were stable, treated only pharmacologically. Patients demographics, clinical symptoms, family history of SCD as well as the results of echocardiography, 12-leads ECG, 24-h Holter ECG were analyzed and compared between the groups.
Results: Children in both groups do not differ regarding age at diagnosis, incidence of syncopes, chest pain, family history of SCD, however pts in gI more often had symptoms such as fatigability, exertional dyspnea (39%vs11%;p=0.017). Patients in both groups differed significantly regarding NYHA functional class (p=0.01). In gI septal thickness (333%vs220% of mean normal range relative to BSA;p<0.0001), thickness of the posterior wall of LV (179%vs138%;p=0.0037) were significantly higher and more frequently LVOTO (48%vs16% pts;p=0.001) was observed. The Sokolow-Lyon index (66mm vs 44mm;p=0.0004), QTc interval (435ms vs 409ms;p=0.002), QTc dispersion (52 ms vs 44 ms;p=0.047) were significantly higher and nsVT was more frequent (24%vs5% pts;p=0.009) in gI. During follow up, 9pts in gI died, 6(6,8%) from progressive heart failure and 3(3,4%) from SCD, mean annual mortality rate was 1,48%.
Conclusions: 1. There was a significant correlation between LV hypertrophy, LVOTO and adverse clinical course of HCM and the need for more aggressive therapy. 2. The value of the Sokolow-Lyon index, QTc interval, QTc dispersion and the presence of nsVT have been associated with a worse prognosis in children with HCM.