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Clinical profile of hypertrophic cardiomyopathy in children – own experience

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Introduction: The clinical presentation 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.
Aim of study: Retrospective analysis of clinical profile in children with HCM hospitalized in the years 1991-2011. Mean follow-up was 7.4yrs (ranged from 1month to 20.5yrs).
Material and methods: We analyzed 92 patients, mean age 10.4±5.1yrs. Patients demographics, clinical symptoms, family history of HCM and SCD as well as the results of echocardiography, 12-leads ECG, 24-h Holter ECG were analyzed.
Results: The mean age at diagnosis of HCM was 5.7yrs. In 32(35%) children HCM was diagnosed in infancy. A family history of HCM was observed in 42(46%)pts. Family SCD due to HCM occurred in 25(27%)children. Patients most frequently reported fatigue, impaired exercise tolerance (66/72%), chest pain (29/32%), recurrent syncope (12/13%). In 60 (65%)pts asymmetric hypertrophy, in 30(33%) concentric hypertrophy, whereas in 2(2.2%) apical hypertrophy of the LV were found. The averaged septal thickness was 16.5mm, which in relation to BSA was 266% of the mean standard. The massive LV hypertrophy ≥ 30mm was observed in 9(9.8%)pts. Abnormal resting ECG recording was found in 95%pts. In 24-h Holter ECG in 13(14%)children nsVT, whereas in none VT were observed. Cardiac arrest with successful resuscitation occurred in 4(4.3%)pts. In 14(15%) children ICD was implanted, in 4(4.3%)pts for secondary prevention and in 10(11%) as primary prevention of SCD. For HTx were enrolled 5 children (in 1pt HTx was performed, 3pts died while waiting for HTx, 1 child is still waiting). In follow-up died 10(11%)pts, average annual mortality was 1.47. In 4(4.3%) children SCD occurred (mean age 11.2yrs), 6(6.5%)pts died due to progressive heart failure (mean age 8.9yrs).
Conclusion: (1) In our group of children with HCM a high incidence of familial form of HCM and sudden cardiac death in the family history were observed. (2) The most common clinical symptom was fatigue, a progressive impaired in exercise tolerance, recurrent syncope and chest pain. (3) The frequency of implantable cardioverter-defibrillator for primary prevention of SCD and annual mortality rate was comparable with the data of the literature.