

Risk stratification and prevention of sudden cardiac death in children with hypertrophic cardiomyopathy.

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Background: Hypertrophic cardiomyopathy is the commonest cause of sudden cardiac death (SCD) in children. The aim of study was to evaluate the risk factors for SCD and the possibility of primary and secondary prevention of SCD in children with HCM .

Methods: Retrospective analysis of 92pts, mean age 10.4±5.1yrs with HCM diagnosed from 1991 to 2011. Mean follow-up was 7.4yrs. Patients demographics, clinical symptoms, family history of HCM and SCD as well as the results of echocardiography, ECG, 24-hour ECG, exercise test were examined. Data from all patients have been analyzed regarding the presence of major risk factors for SCD according to ACC/AHA/ESC 2006 recommendations.

Results: The mean age at diagnosis of HCM was 5.7±5.6 yrs. Screening for familial HCM revealed 42(46%) positive cases, of whom 79% had a first degree affected family member. The major risk factors for SCD were present in 51(55%) pts: cardiac arrest in 4(4.3%)pts, sustained VT in none of pts, family history of SCD (FSCD) in 25(27%), syncope in 12(13%), LV thickness ≥30mm in 9(9.8%), abnormal exercise BP (ABPR) in 20(21%) and nsVT in 13(14%)pts. ICD was implanted in 14(15%)pts, in all 4 pts after cardiac arrest as secondary prevention, in 10(11%) children as primary prevention.

Among the 10 patients with ICD implanted as primary prevention, 4 pts had one (3pts recurrent syncope, 1pt nsVT), 5 children had two (FSCD+nsVT; syncope+IVS≥30 mm; nsVT+IVS≥30 mm; FSCD+syncope; FSCD+ IVS≥30 mm) and 1pt had three major risk factors (FSCD+IVS≥30mm+ABPR). The appropriate shock was diagnosed in 2pts, inappropriate interventions were observed in 1pt with ICD implanted as secondary prevention. In children in whom the ICD was implanted as primary prevention, there was no adequate or inadequate discharge.

Conclusions: (1) The major risk factors for SCD was found in up to 51% of children, of whom 22 (24%) had ≥ two risk factors. (2) Most common indication for ICD implantation as primary prevention of SCD were the major risk factors such as family history of SCD, recurrent syncope and massive LV hypertrophy. (3) High-risk patients with HCM ought to be prospectively identified and ICD implantation should be considered.