

Predictors of Appropriate Interventions in Children with Hypertrophic Cardiomyopathy and a Primary Prevention Defibrillator: An international multi-center study

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Introduction: Primary prevention (PP) ICD are increasingly placed in children with Hypertrophic Cardiomyopathy (HCM) to prevent SCD using adult criteria. The reliability of PPICD to prevent SCD is unclear.

Methods: We collected data on HCM children ≤ 20 years with PPICD. Risk factors (RF) for SCD were 1) family history (FH) of SCD; 2) syncope; 3) maximal Left ventricular (LV) wall thickness (LVWT) Z value > 5 ; 4) non-sustained VT (NSVT) on ambulatory ECG; and 5) abnormal BP response to exercise (ABPR).

Results: Of 347 patients (age 14.61 ± 4.18), with PP-ICD, appropriate interventions (AI) occurred in 55 (16%). RF presence pre-ICD implant was FH-SCD in 114 (33%); LVWT in 230 (66%); NSVT in 25 (7%); ABPR in 105 (30%) and syncope in 84 (20%). Follow up after ICD implant was 0.07 to 32.95 (mean 8.82 ± 6.03) years and incidence of AI was 18.40 cases per 1000 person-years. AI incidence for each RF was: FH-SCD: 20/114 (17.5%); LVWT 36/230 (15.7%); NSVT 4/25 (16.0%); ABPR 18/105 (17.1%) and syncope 22/84 (26.2%). Risk of AI based on # of RF were: 5/31 (16%) for 0 RF; 16/126 (13%) for 1 RF; 20/146 (14%) for 2 RF; 12/36 (33%) for 3 RF and 2/8 (25%) for 4 RF.

Conclusions: The incidence of AI in HCM children with PP-ICD based on adult criteria was high. Syncope was the commonest RF associated with AI. All other RF's had similar incidence in patients with AI. Presence of ≥ 3 RF was associated with AI.