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Risk factors for Sudden Cardiac Death in Childhood Hypertrophic Cardiomyopathy: A systematic review and meta-analysis.

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Objective: To perform a systematic literature review and meta-analysis of clinical risk factors for sudden cardiac death in childhood hypertrophic cardiomyopathy.

Methods: Medline and Pubmed databases were searched for original articles published in English from 1963 through to December 2015 which included patients under 18 years with a primary or secondary end-point of either sudden cardiac death (SCD) or equivalent (aborted cardiac arrest or appropriate ICD discharge) or cardiovascular death (CVD).

Results: Twenty eight studies (3544 patients) met inclusion criteria. We identified four conventional major risk factors which were evaluated in at least 4 studies and found to be statistically associated with an increased risk of death in at least 2 studies: previous adverse cardiac event (pooled hazard ratio 5.4 (95% CI 3.67-7.95), p <0.001, I2=0%); non-sustained ventricular tachycardia (pooled hazard ratio 2.13 (95% CI 1.21-3.74), p=0.009, I2=19%), unexplained syncope (pooled hazard ratio 1.89 (95% CI .69-5.16), p=0.22, I2=46%) and extreme left ventricular hypertrophy (pooled hazard ratio 1.80 (95% CI 0.75-4.32), p=0.19, I2=21%).
Additional ‘minor’ risk factors included a family history of sudden cardiac death, gender, age, symptoms, ECG changes, abnormal blood pressure response to exercise, left atrial diameter and left ventricular outflow tract obstruction.

Conclusions: A lack of well-designed, large population based studies in childhood hypertrophic cardiomyopathy means the evidence-base for individual risk factors is not robust. We have identified four clinical parameters which are likely to be associated with increased risk of SCD, SCD-equivalent event or CVD. Multi-centre prospective studies are needed to further determine their relevance in predicting SCD in childhood HCM and to identify novel risk markers.