

Family history of sudden death is not an independent risk factor for sudden death in childhood hypertrophic cardiomyopathy - risk is determined by ECG- and echocardiography predictors

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BACKGROUND Reports from tertiary centres in adult cardiology specializing in hypertrophic cardiomyopathy (HCM) have indicated family history of sudden death (SD) as a significant risk factor, and it is classed as one of three strong risk factors meriting implantation of an internal cardiac defibrillator (ICD) in the latest American Heart Association guidelines. However, several studies from paediatric cardiology centres with few events have failed to confirm this association in paediatric HCM patients. We have studied this question in a national cohort.

PATIENTS AND METHODS Patients with a diagnosis of HCM before age 19 years attending all five regional centres of paediatric cardiology in Sweden have been studied. There were 27 patients with sudden death or re-suscitated cardiac arrest, and 103 patients with at least two years follow-up and without heart-failure death (mean follow-up 9.2 years). These were studied with Cox proportional hazard regression in respect to previously suggested possible risk factors for sudden death.

RESULTS On univariate Cox-hazard analysis first and latest ECG risk score (both $p < 0.001$) (see Eur Heart J 2010;31:439-449), both first and latest septal thickness in percent of 95th centile (SEPPER) (both $p < 0.001$), VT on Holter ($P = 0.003$), left ventricular outflow tract obstruction at rest ($p = 0.001$), and latest left atrium:aortic ratio ($p = 0.002$) were significant risk factors whereas family history of SD ($p = 0.89$), and maximal wall thickness in mm ($p = 0.80$) were not. Beta-blocker therapy ($p = 0.002$) was the only therapy to significantly reduce risk, also evident as dose-related protection (propranolol-equivalent/kg $p = 0.012$). On multivariate Cox-hazard last ECG risk score ($p < 0.001$), last SEPPER ($p = 0.001$), gender ($p = 0.054$) and beta-blocker therapy ($p < 0.001$, protective) were significant. Out of 34 patients with family history of SD 7 died suddenly, and 27 have survived (median 8 years). Those who died were identifiable as high-risk phenotypes with initial SEPPER median 289% [IQR 194-435], and initial and last ECG risk scores 7 [4-9] and 8 [7-11], versus survivors: first SEPPER 134% [105-192] $p = 0.0022$, first risk score 2 [1-4] $p = 0.017$, and last ECG risk score 1 [0-5] $p = 0.0026$.

CONCLUSION Family history of SD is not an independent risk factor in childhood HCM; the phenotype determines risk of sudden death.