Risk factors for sudden death and for heart failure-related death in childhood hypertrophic cardiomyopathy.

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Background: To date limited data is available to predict the progression to end-stage heart failure with subsequent death (non-SCD), need for heart transplantation or sudden cardiac death (SCD) in children with hypertrophic cardiomyopathy (HCM). Current treatment strategies in children are predominantly based on the known risk factors for SCD in adults with HCM.

Objectives: The aim of study was to determine predictors of long-term outcome in children with HCM including known adult risk factors for SCD and previously proposed pediatric risk factors for death. Moreover, we distinguish both arrhythmic and heart failure end points to determine specific risk factors these two modes of unfavorable outcome.

Material and Methods: A total of 112 children (median age 14.1, IQR 7.8-16.6 years, 60% male) were followed up for the median of 6.5 years for the development of morbidity and mortality, including the pre-specified primary end points: the cardiovascular death, resuscitated cardiac arrest, appropriate implantable ICD discharge, a heart transplant and arrhythmic or heart failure-related secondary end points. Heart failure end point included heart failure-related death or heart transplant and arrhythmic end point included resuscitated cardiac arrest, appropriate ICD discharge or sudden cardiac death.

Results: Overall 23 (21%) patients reached the pre-defined composite primary end point. At 10 year follow-up the event-free survival rate was 76%. Thirteen patients (12%) reached the secondary arrhythmic end point and 10 patients (9%) reached the secondary heart failure end point. In multivariate model prior cardiac arrest (r=0.658), QTc dispersion (r=0.262) and NSVT (r=0.217) were independent predictors of the arrhythmic secondary end point while heart failure (r=0.440), left ventricular posterior wall thickness (r=0.258), left atrial size (r=0.389) and decreased early transmitral flow velocity E (r=0.202) were all independent predictors of the secondary heart failure end point.

Conclusion: There are differences in the risk factors for sudden death and for heart failure-related death in childhood hypertrophic cardiomyopathy. Only prior cardiac arrest, QTc dispersion and non-sustained ventricular tachycardia predicted arrhythmic outcome in patients aged <18 years. Left atrial size, left ventricular posterior wall thickness and decreased early transmitral flow velocity were strong independent predictors of heart-failure related events.