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A comparison of different risk stratification strategies for sudden cardiac death in pediatric hypertrophic cardiomyopathy

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INTRODUCTION Studies on pediatric patients with hypertrophic cardiomyopathy (HCM) have failed to confirm that risk-factors for sudden death considered important in American Heart Association (AHA) 2011 risk-stratification guidelines, the only ones recognized for patients <16 yrs of age, are significant predictors in childhood-HCM, with the exception of ventricular tachycardia (VT).

METHODS A national Swedish cohort of patients with a diagnosis of hypertrophic cardiomyopathy (HCM) presenting <19 years of age contains 140 patients, follow-up of 10.9 (SD±9.0) yrs, with both ECG- and wall thickness measurements, data on syncope, and family history of sudden death (FHSCD). 129 have had 24h Holter-ECG or equivalent monitoring. There were 27/140 sudden deaths/cardiac arrests (SD/CA) in the total group and 21/129 in the latter group. The ability to predict SD/CA for proposed risk predictors was studied with ROC-curve analysis.

RESULTS Analyzed as continuous function in the ECG-monitored group the number of “major” AHA risk-factors (FHSCD, max wall thickness ≥3cm, unexplained syncope) had area under the curve (AUC) 0.53 [0.39-0.67,p=0.66]. Using AHA2011 criteria and incorporating presence of VT and/or abnormal BP-response to exercise if other major risk factor present, improves AUC only to 0.56 [p=0.40], but expressing max wall thickness as Detroit Z-score with a cut-off at ≥6 instead of ≥3cm improves AUC to 0.64 [0.51-0.77, p=0.042). Examining individual AHA-risk factors, neither FHSCD (p=0.64), or pathological BP-response on exercise (p=0.20) were significant, whereas VT on Holter had AUC=0.79 [0.63-0.86, p<0.001]. A pediatric modification of scoring using only max wall thickness Detroit score ≥6, VT on Holter and unexplained syncope as risk factors improved AUC further to 0.80 [0.69-0.91,p<0.001]. In the same population, both ECG-risk score >5 points AUC=0.87 [0.80-0.93,p<0.001], last Detroit Z-score ≥4.5 AUC=0.80 [0.70-0.89,p<0.001], and as continuous functions: ECG risk score AUC=0.90 [0.85-0.95,p<0.001] and last Z-score AUC=0.86 [0.78-0.93 p<0.001], performs better as risk predictors. ROC-curves in total group gave virtually identical values.

CCONCLUSIONS “Major” risk-factors stated in AHA 2011-guidelines have too low sensitivity and specificity in pediatric HCM-patients, and risk-stratification based on ECG-phenotype is more effective. There is a need for the development of a pediatric risk calculator based on childhood risk-factors and including ECG-phenotype.