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A novel risk prediction model for sudden cardiac death in childhood hypertrophic cardiomyopathy (HCM Risk-Kids)

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Topic(s):
Hypertrophic Cardiomyopathy

Citation:

Funding Acknowledgements:
British Heart Foundation

Background:

Sudden cardiac death (SCD) is the most common mode of death in childhood hypertrophic cardiomyopathy (HCM) but there is no validated algorithm to identify those at highest risk. This study sought to develop and validate a SCD risk prediction model that provides individualized risk estimates.

Methods:

A prognostic model was derived from an international, retrospective, multi-center longitudinal cohort study of 1024 consecutively evaluated patients aged =16 years. The model was developed using pre-selected predictor variables [unexplained syncope, maximal left ventricular (LV) wall thickness (MWT), left atrial diameter (LAD), LV outflow tract (LVOT) gradient and non-sustained ventricular tachycardia (NSVT)] identified from the literature and internally validated using bootstrapping.

Results:

Over a median follow up of 5.3 years (IQR 2.6, 8.2, total patient years 5984), 89 (8.7%) patients died suddenly or had an equivalent event [annual event rate 1.49 (95% CI 1.15-1.92)]. The pediatric model was developed using pre-selected variables to predict the risk of SCD. The model’s ability to predict risk at 5 years was validated; C-statistic was 0.69 (95% CI 0.66-0.72) and the calibration slope was 0.98 (95% CI 0.58-1.38). For every 10 ICDs implanted in patients with =6% 5-year SCD risk, potentially 1 patient will be saved from SCD at 5 years.

Conclusions: This new validated risk stratification model for SCD in childhood HCM provides accurate individualized estimates of risk at 5 years using readily obtained clinical risk factors.