

YIA-4

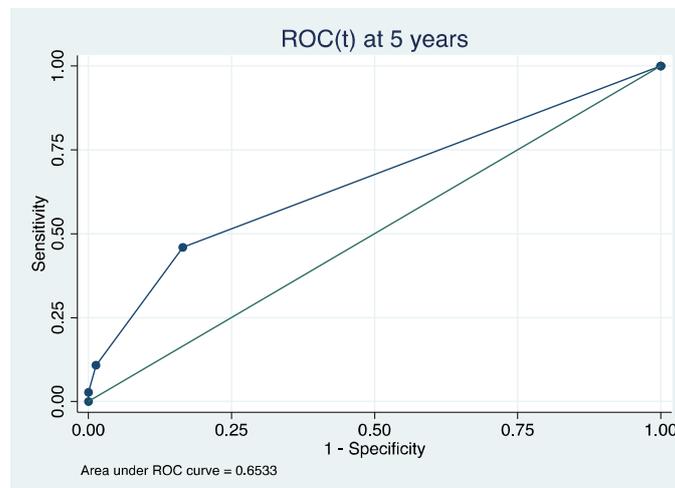
A validation study of the paediatric guidelines of the 2014 European Society of Cardiology risk stratification algorithm for sudden cardiac death in hypertrophic cardiomyopathy

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Introduction (or Basis or Objectives):

Sudden cardiac death (SCD) is the most common cause of death in children with hypertrophic cardiomyopathy (HCM). However, identifying which patients are at high risk and may benefit from an implantable Cardioverter Defibrillator (ICD) is challenging. A personalised risk score for adults with HCM has recently been incorporated into the European Society of Cardiology (ESC) guidelines, but cannot be used in children. Instead, current ESC recommendations suggest consideration of an ICD in children with HCM if two or more clinical risk factors are present. However, this approach to risk stratification in childhood HCM has not been formally validated.



Methods:

687 paediatric HCM patients derived from a retrospective, national cohort study in United Kingdom were assessed for 4 clinical risk factors (RF) for SCD: severe left ventricular hypertrophy (maximal wall thickness >30mm or Z score >6); unexplained syncope; non-sustained ventricular tachycardia; and family history of SCD. The primary end point was a composite of SCD, appropriate ICD therapy or sustained ventricular tachycardia.

Results:

Over a follow up period of 4596 patient years (median follow up 5.2 years), SCD or equivalent event occurred in 33 of 531 (6.2%) patients with no RF, 20 of 138 (14.5%) patients with 1 RF, 4 of 16 (25%) patients with 2 RF and 1 of 2 patients (50%) of patients with 3 RF. Annual rate of SCD was 0.01%, 0.02%, 0.04% and 0.08% respectively. The risk of SCD increased with additional clinical risk factors (1 RF hazard ratio 1.84, $p=0.032$; 2 RF hazard ratio 3.58 $p = 0.016$; 3 RF hazard ratio 9.00 $p= 0.08$). The positive predictive value for SCD if 2 or more risk factors were present is 27.8%. The concordance statistic for the ability to correctly identify a patient at high risk based on the number of risk factors was 0.62 at 1 year and 0.65 at 5 years.

Conclusions:

The risk of SCD is higher for patients with increasing numbers of clinical risk factors. However, the current ESC guidelines have a low ability to discriminate between high and low risk individuals. Future studies are required to develop and evaluate risk stratification strategies in childhood HCM.

Legend:

Receiver operating characteristic (ROC(t)) Curves for the European Society of Cardiology (ESC 2014) guideline at 5 years.