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ECG risk score: a significant advance in the risk stratification of paediatric patients with hypertrophic cardiomyopathy

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INTRODUCTION: In hypertrophic cardiomyopathy (HCM) the risk of sudden unexpected arrhythmia death in an asymptomatic individual is at its highest in between 8-16 years of age. In adult HCM patients we have shown that abnormally large ECG-voltages, and repolarisation abnormalities indicate increased risk, and have published an ECG risk score which improves risk stratification. The objective of this study is to evaluate this risk score for paediatric HCM-patients.

ECG risk score		
Any deviation in QRS-axis		1 point
Pathological T-wave inversion limb leads		1 point
Pathological T-wave inversion precordial leads*		2 points
ST-segment depression $\geq$ 2 mm		2 points
Dominant S in V_4		2 points
Limb-lead QRS-amplitude sum	$\geq$7.7 mV	1 point
	$\geq$10.0 mV	2 points
	$\geq$12.0 mV	3 points
12-lead amplitude-duration product	$\geq$2.2 mV.s	1 point
	$\geq$2.5 mV.s	2 points
	$\geq$3.0 mV.s	3 points
QTc	$\geq$440 ms	1 point
		Max score=14

*Total score available for T-wave abnormalities is 2 points, i.e. 1 limb lead point is not added on top of precordial points. QTc = corrected QT-interval (Eur Heart J 2010;31:439-49).

METHODS: All Sweden´s centres of paediatric cardiology collaborated on identifying all patients that had died suddenly from HCM between birth and 19 years of age in the last 30 years. 33 patients were identified, with 29 patients where ECGs had been recorded prior to death (mean follow-up 9.8$\pm$5.9 (SD) years). For comparison we used ECGs from the total geographical cohort of HCM-patients age 0-19 years from West Götaland region where systematic cascade screening has been used to identify symptom-free children with HCM (n=48, follow-up 6.8$\pm$8.1 years).

RESULTS: A high risk ECG-score limit set at 6 points or above gives a relative risk for sudden death of 24.3 [95% CI 3.5-169; p <0.0001], sensitivity of 96% [80-100%] and specificity of 78% [62-89]. The positive predictive value was calculated within the West Götaland cohort where there were 7 deaths (giving an annual sudden death mortality of 2.1%). Sensitivity was here 100% [54-100%], positive predictive value 40% [16-68%] and negative predictive value 100% [89-100%].

CONCLUSIONS: The ECG risk score is applicable to children, and the hitherto most specific individual risk factor described.