Risk stratification in hypertrophic cardiomyopathy: Time to think about the electrocardiogram

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Dear Editor,

The review article in this journal by Norrish and Kaski focuses on risk of sudden cardiac death in childhood hypertrophic cardiomyopathy and declare it is *time to solve the mystery*.[1] The authors make a reasoned plea for large collaborative projects to try to solve the mystery of how to risk stratify children with hypertrophic cardiomyopathy (HCM) for risk of sudden death. Let's do it. But in designing such projects let us not forget the electrocardiogram (ECG). This article is one of many which has overlooked the interesting work from Sweden exploring the use of the 12 lead ECG in risk stratification. This work has been published in journals of high reputation and yet has not gained any traction, nor (to my knowledge) been assessed in any other cohorts. In 2010 in the European Heart Journal, Ostman Smith *et al.* found that an ECG risk score was superior to traditional risk scores.[2] In 2017, in Open Heart, the same group produced a study of the Swedish national cohort of children,[3] 155 children followed up carefully for an average of 11 years. The best ECG score alone predicted a 5 year risk of sudden cardiac death of 31%, and the authors conclude that the ECG score should be included as part of risk stratification. The ECG score includes assessment of repolarisation (ST segment depression, QT interval) and of voltage. It not only makes intuitive sense that electrocardiographic features reflecting myocardial mass and ischaemia may be markers of disease severity and arrhythmic risk, it is also partly supported by other data correlating, for example, QTc with risk of ICD discharge in HCM[4] and QTc with the presence of single nucleotide polymorphisms in channelopathy genes previously linked to risk of sudden death.[5] It certainly seems like a hypothesis worthy of further evaluation.

As Norrish and Kaski suggest, it is time to develop a multinational prospective study of childhood HCM. Those of us who can should collaborate, and this should include serial 12 lead ECGs as well as traditional risk factors.

References

1. Norrish G, Kaski JP. Risk of sudden cardiac death in childhood hypertrophic cardiomyopathy: Time to solve the mystery. Cardiogentics 2018;8:7201.
2. Ostman-Smith I, Wisten A, Nylander E, et al. Electrocardiographic amplitudes: a new risk factor for sudden death in hypertrophic cardiomyopathy. Eur Heart J 2010;31:439-49.
3. Ostman-Smith I, Sjoberg G, Rydberg A, et al. Predictors of risk for sudden death in childhood hypertrophic cardiomyopathy: the importance of the ECG risk score. Open Heart 2017;4:e000658.
4. Gray B, Ingles J, Medi C, et al. Prolongation of the QTc interval predicts appropriate implantable cardioverter-defibrillator therapies in hypertrophic cardiomyopathy. JACC Heart Failure 2013;1:149-55.
5. Earle N, Ingles J, Bagnall RD, et al. NOS1AP Polymorphisms Modify QTc Interval Duration But Not Cardiac Arrest Risk in Hypertrophic Cardiomyopathy. J Cardiovasc Electrophysiol 2015;26:1346-51.