ACHD/Valve disease/Pericardial disease/ Cardiomyopathy

1 THE ROLE OF THE ELECTROCARDIOGRAPHIC PHENOTYPE IN RISK STRATIFICATION FOR SUDDEN CARDIAC DEATH IN CHILDHOOD HYPERTROPHIC CARDIOMYOPATHY

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Introduction Sudden cardiac death is the most common cause of mortality in childhood onset hypertrophic cardiomyopathy. Identifying individuals at highest risk is therefore an essential part of clinical care but remains challenging. The 12 lead electrocardiogram (ECG) has been proposed as a useful tool for risk stratification and an ECG risk score has been proposed. However, this has not been independently validated and the ECG phenotype of childhood HCM has not been previously described. The aim of this study was to describe the ECG phenotype of childhood HCM in a large, international, multi-centre cohort and investigate its role in risk prediction for arrhythmic events.

Methods Participants with an available baseline resting 12-lead ECG were identified from a large, international, multi-centre, retrospective cohort of patients aged less than 16 years fulfilling the diagnostic criteria for HCM (n=1029). Resting baseline ECG was evaluated and ECG variables were extracted. In addition, the ECG risk score based on 8 parameters (deviation in QRS axis, pathological T-wave inversion in limb or precordial leads, ST-segment depression, dominant S-wave in V4, limb-lead amplitude sum, 12-lead amplitude

duration product and QTc) was calculated as previously described. The primary study endpoint was a composite outcome of major cardiac events (MACE) defined as SCD, resuscitated cardiac arrest, appropriate implantable cardioverter defibrillator therapy, or sustained ventricular tachycardia (VT) with haemodynamic compromise. The discriminatory performance of using an ECG risk score >5 to identify patients at increased risk of MACE at 5 years was determined using Harrell's C-index.

Results Of 356 patients with childhood HCM (68.9% male, mean age at presentation 10.1 ± 4.5 years), 347 (97.5%) had baseline ECG abnormalities such as: repolarization abnormalities (n=277, 77.8%), left ventricular hypertrophy (n=240, 67.6%), abnormal QRS axis (n=126, 35.4%) or QT prolongation (n=131, 36.8%). Over a median follow up of 3.9 years (IQR 2.0-7.7), 25 (7%) had an arrhythmic event, with an overall annual event rate of 1.38 (95% CI 0.93-2.04). No ECG variables were associated with 5-year MACE on univariable or multivariable Cox regression analysis. Of the 164 participants with an ECG score >5, 153 (93.3%) did not have a MACE within 5 years. Harrell's C-index (the probability of correctly distinguishing between high and low risk patients using an ECG risk score threshold of >5) was 0.60 (95% CI 0.484-0.715) at 5 years. The corresponding positive and negative predictive values were 6.7% (95% CI 4.7 – 9.4%) and 96.9% (95% CI 94.2 – 98.4%).

Conclusions In a large, international, multi-centre cohort of children with HCM, ECG abnormalities are common. No ECG characteristic, either in isolation or combined in the ECG risk score, was associated with 5-year MACE risk. This suggests that the role of the baseline ECG phenotype in improving risk stratification in childhood HCM is limited.

Conflict of Interest None

2 DIFFERENTIAL EFFECTS OF LEFT VENTRICULAR HYPERTROPHY ON CORONARY HAEMODYNAMICS IN AORTIC STENOSIS AND HYPERTENSION

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Background Hypertension and aortic stenosis(AS) are the commonest causes of left ventricular hypertrophy (LVH) and share similar pathophysiological features. Whilst an increase in resting coronary blood flow (per gram of LV) has been observed in AS, reduced resting coronary blood flow (per gram of LV) has been observed in hypertension.

Aim We aimed to compare coronary flow patterns in subjects with left ventricular hypertrophy and aortic stenosis, in subjects with left ventricular hypertrophy and hypertension, and in subjects without left ventricular hypertrophy or hypertension.

Methods We recruited 31 subjects (mean age 63, 18 female). 10 subjects had LVH and severe AS, 11 had LVH and hypertension and 10 had no LVH and no AS, with LVH defined on echocardiography.

Simultaneous invasive pressure and Doppler velocity measurements in each of the left coronary arteries were taken. We performed 'wave intensity analysis', which is a method for