

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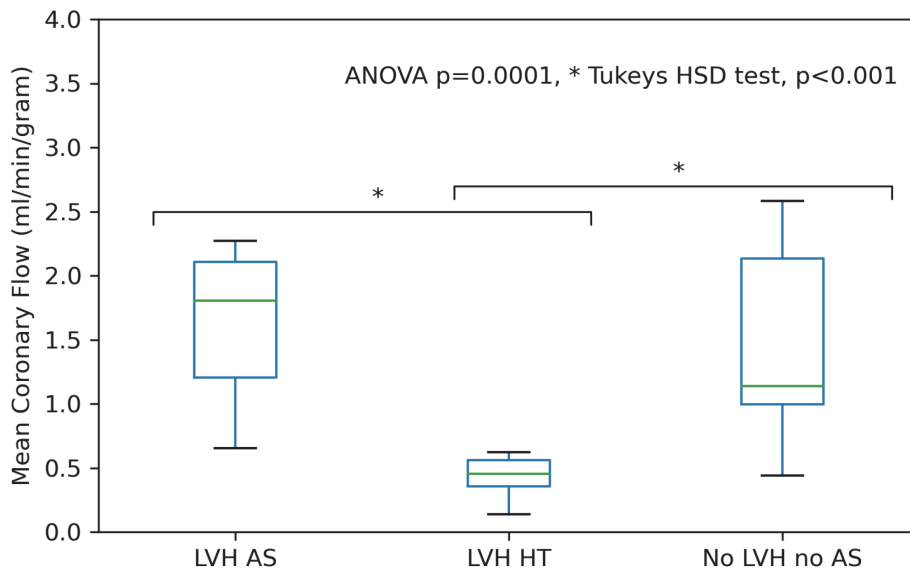
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Background Hypertension and aortic stenosis(AS) are the commonest causes of left ventricular hypertrophy (LVH) and share similar pathophysiological features. Whilst an increase in resting coronary blood flow (per gram of LV) has been observed in AS,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

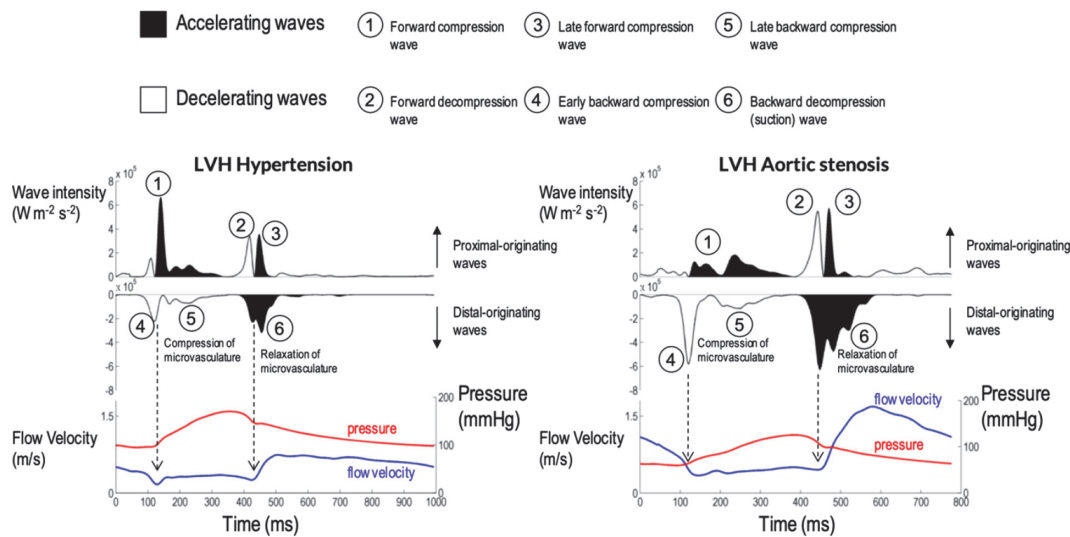
Aim We aimed to compare coronary flow patterns in subjects with left ventricular hypertrophy and aortic stenosis, in subjects with left ventricular hypertrophy and hypertension, and in subjects without left ventricular hypertrophy or hypertension.

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



Abstract 2 Figure 1 Mean coronary flow per gram of LV for each group



Abstract 2 Figure 2 Intracoronary wave intensity in a participant with LVH secondary to hypertension (left) and in a participant with LVH secondary to aortic stenosis (right)

separating the coronary flow pattern in terms of ‘waves’ that are generated proximally (by the aorta and systemic arteries) and distally (by the myocardial microcirculation).

Results Mean resting coronary flow per gram of tissue (figure 1) was increased in participants with LVH secondary to AS ($1.62 \pm 0.60 \text{ ml/min/g}$) and reduced in participants with LVH secondary to HT ($0.49 \pm 0.27 \text{ ml/min/g}$), compared to participants with no LVH and no AS ($1.47 \pm 0.73 \text{ ml/min/g}$).

Wave 6 (figure 2) is the backwards decompression wave (BDW) and is particularly important for myocardial perfusion. The BDW corresponds to the diastolic ‘suction’ of blood down the coronary arteries during myocardial relaxation.

The energy of the BDW was increased in LVH secondary to AS ($31.1 \times 10^3 \text{ Wm}^{-2}\text{s}^{-2}$) but was reduced in LVH secondary to HT ($12.3 \times 10^3 \text{ Wm}^{-2}\text{s}^{-2}$) ($p < 0.05$), compared to participants with no LVH and no AS ($14.3 \times 10^3 \text{ Wm}^{-2}\text{s}^{-2}$).

The energy of the BDW correlated with LV cavity pressure ($r = 0.84$, $p < 0.001$) and diastolic time ($r = -0.62$,

$p < 0.001$) only in LVH secondary to AS participants. In contrast, the BDW correlated with LV mass ($r = -0.49$, $p = 0.03$) in participants with LVH secondary to HT and with no LVH and no AS, but not in participants with LVH secondary to AS.

Conclusions In hypertension, LVH is associated with reduced mean coronary flow and reduced myocardial ‘suction’ during diastole.

However, in AS, the large pressure gradient between the LV cavity pressure and the aorta results in a large contractile force which is generated in systole and then released in diastole. This large diastolic force overwhelms any local impairment caused by the hypertrophied myocardium and contributes to high resting coronary flow in LVH that is secondary to AS, compared to LVH that is secondary to hypertension.

Conflict of Interest None