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APICAL VERSUS NON-APICAL HYPERTROPHIC CARDIOMYOPATHY (HCM): INSIGHT FROM CARDIAC MAGNETIC RESONANCE IMAGING

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10.1136/heartjnl-2016-309890.144

Background Hypertrophic cardiomyopathy (HCM) is the most common genetic cardiac disorder, and the most common cause of sudden cardiac death (SCD) in young adults. The 3 main phenotypes are asymmetric, concentric or apical, with asymmetric being the most common. Literature suggests apical HCM to be a rare variant (variable prevalence) with better prognosis but the data is limited.

Aims Provide a contemporary prevalence and characteristics of apical HCM in a large tertiary clinical CMR service.

Methods Approximately 3,100 CMR scans were reviewed from our CMR registry (Jan 2014 to Mar 2015). comprehensive CMR protocol was used including cines, early and late gadolinium enhancement imaging. 114 consecutive HCM patients were identified. A Asymmetric HCM was defined as: septal to free wall thickness ratio of > 1.3 ; apical HCM as apical wall thickness of > 15 mm or apical to basal LV wall thicknesses ≥ 1.3 – 1.5 ; and concentric HCM as symmetrical hypertrophy of ventricular wall without any regional preferences. Non-apical HCM group (comprising of asymmetric and concentric phenotypes) were compared with apical HCM. Fisher's exact t-test and unpaired t-test were performed for statistical significance. P-value < 0.05 was statistically significant. Univariate and multivariate logistic regression analyses were performed to determine the CMR predictors of apical HCM.

Results The final study sample consisted of 104 patients with HCM with median age 60years (IQR = 54–70) and 70% male, (10 patients excluded due to uncertain diagnosis) 70% non-apical HCM; the remainder 30% apical HCM. In the non-apical HCM group, 5 patients had concentric HCM and the rest had asymmetric HCM. The. The mean maximum LV wall thickness, mean indexed LV mass, mean indexed stroke volume, prevalence of LVOTO and SAM were significantly greater in non-apical group. Table 1 The presence of LGE was high in both groups ($>85\%$) and was not statistically different. The univariate predictors of apical HCM included maximum LV wall thickness, indexed stroke volume, LVOT obstruction whereas in the multivariate model maximum LV wall thickness remained the only significant predictor.

Conclusions Our study suggests that in the era of CMR, the prevalence of apical HCM to be almost 1/3rd of all observed HCM cases. The study also demonstrates that the prevalence of LGE was high also in the apical HCM group suggesting that the better prognosis that apical HCM is thought to have based on the absence of myocardial fibrosis should be reconsidered. Further large prospective multi-centre trials are needed to establish the key differences thereby understanding the pathophysiology.

Abstract 144 Table 1 CMR characteristics of Apical vs non-Apical HCM

CMR findings	Total Cohort (n = 104)	Non-apical (n = 73)	Apical (n = 31)	P-value
Mean LVEF (%)	69.7	68.4	72.6	0.0552
Mean LVEDVI (mL m ⁻²)	73.7	76.7	66.8	0.0718
Mean LVESVI (mL m ⁻²)	23.8	25.5	20.1	0.1177
Mean indexed stroke volume	53.1	55.9	46.4	0.0333
Mean max. LV wall thickness (mm)	18.2	19.3	15.6	0.0001
Mean indexed LV mass	93.5	98.4	82.4	0.0102
LVOTO	35.2	41.1	12.5	0.0403
SAM	31.4	38.9	6.25	0.0143
LGE%	86.9	85.7	89.7	0.8063

LVEF, left ventricular ejection fraction; LVEDVI, left ventricular end diastolic volume index; LVESVI, left ventricular end systolic volume index; LVOTO, Left ventricular outflow tract obstruction; SAM, systolic anterior valve motion; LGE, late gadolinium enhancement.

Cardiac Rhythm Management

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RISK STRATIFICATION IN HYPERTROPHIC CARDIOMYOPATHY: EVALUATION OF THE EUROPEAN SOCIETY OF CARDIOLOGY SUDDEN CARDIAC DEATH RISK SCORING SYSTEM

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10.1136/heartjnl-2016-309890.145

Introduction Implantable cardio-defibrillators (ICDs) have proven benefit in treating lethal ventricular arrhythmias and preventing sudden death (SD) in hypertrophic cardiomyopathy (HCM), making risk stratification essential. We retrospectively evaluate the effectiveness of the 2014 European Society of Cardiology (ESC) risk scoring system in our cohort of HCM patients.

Methods We evaluated the ESC risk scoring system which employs mathematical and statistical modelling of 7 disease variables to predict SD risk over 5 years, with a recommendation for ICD implant if SD risk $\geq 6\%$. From our cohort of HCM patients previously evaluated at our centre, we retrospectively calculated the ESC 5 year SD risk score at point of implant and measured it against ICD outcome. Decision of ICD implant, prior to the introduction of the ESC scoring system, was based on clinical history and number of conventional risk markers as defined by the American College of Cardiology and Heart Association.

Results 52 out of 199 HCM patients (mean age 51 ± 13 yrs) underwent ICD implantation for primary prevention, with 8 (15%) having appropriate therapy for sustained ventricular tachycardia/fibrillation (VT/VF) over an average follow up period of 6.2 ± 4.9 yrs. There was no difference in the ESC risk scores between patients with or without device therapy ($4.79\% \pm 1.5$ vs $5.37\% \pm 3.3$, $p = 0.68$) (Table 1). 5 of 8 (62%) patients with appropriate therapies for VT/VF had scores ranging from 3.08–5.05% and would not have reached the threshold for an ICD recommendation. In two an ICD

would not be recommended and may be considered in the other three.

Conclusion The current ESC scoring system potentially leaves many high-risk patients unprotected or with ambiguous ICD implant guidance. Lowering the current threshold may improve accuracy.

Imaging

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CONTRIBUTION OF CONDUCTION AND REPOLARISATION ABNORMALITIES TO THE TYPE I BRUGADA PATTERN: A STUDY USING NON-INVASIVE ELECTROCARDIOGRAPHIC IMAGING

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10.1136/heartjnl-2016-309890.146

Introduction In Brugada Syndrome (BrS), the substrate location and underlying electrophysiological mechanisms that contribute to the characteristic ECG pattern are still debated. Using non-invasive electrocardiographical imaging (ECGi), we study whole heart conduction and repolarisation patterns following an ajmaline challenge in individuals with concealed Type I BrS.

Methods 13 participants (mean age 44 ± 12 yrs; 8 males), 11 concealed Type I BrS and 2 controls, underwent an Ajmaline infusion with ECGi and ECG recordings for a research study.

ECGi technology reconstructs >1000 electrograms (EGMs) from 252 surface electrode vest and projects this mathematically onto a 3D cardiac image created using a CT scan. Activation time points were determined as the QRS (dP/dtmin) and repolarisation time as (dP/dtmax) for positive T waves and (dp/dtmin) for negative or biphasic T waves, annotated using a custom built semi-automated software off-line. From these data, the local activation recovery interval (ARI), a surrogate of action potential duration, and activation timings across the right ventricle (RV) body, out flow tract (RVOT), and left ventricle (LV) were computed for all participants (Figure 1a). Changes in AT timings and ARI across the RVOT, RV and LV with ajmaline were calculated, and correlated with peak ST elevation (STE) derived from the ECG at the same time point.

Results Following ajmaline administration, the greatest median increase in conduction delay was noted in the RVOT than in the RV or LV (5[3–8] ms vs 1[0–4]ms vs 1[0–2] ms; $p < 0.0001$) (Figure 1b). Prolongation of ARI was also observed to have increased the most in the RVOT (68[53–99] ms vs 35[23–46] ms vs 25[9–30] ms; $p < 0.01$). In the two control patients, no STE was noted with minimal rise in conduction delay or ARI prolongation noted in the RVOT, RV and LV. Only conduction delay in RVOT with ajmaline correlated to amount of STE (Pearson R 0.8, $p < 0.001$) (Figure 1c), but not in the RV or LV (Pearson 0.3 and 0.2 respectively; $p = \text{ns}$). No significant correlation was also seen between STE and ARI prolongation in the RVOT, RV or LV (Pearson 0.5, 0.4, 0.1 respectively; $p = \text{ns}$).

Conclusion Magnitude of STE in the Type I BrS pattern is attributed to degree of conduction delay in the RVOT and not prolongation in repolarisation time.

Abstract 145 Table 1 Summary of risk factors in (primary prevention) patients with and without ICD therapy. Mean $\pm$ SD and proportion shown

	Appropriate therapy (n=8)	No therapy (n=44)	
ESC 5 year risk score (%)	4.79 $\pm$ 1.5	5.37 $\pm$ 3.3	p=ns
Average no. of risk markers (ACC/AHA)	2	1.6	
Age at implant (yrs)	51 $\pm$ 13	47 $\pm$ 13	
LVH (mm)	23 $\pm$ 7	24 $\pm$ 6	
LVOT gradient (mmHg)	8 $\pm$ 3	28 $\pm$ 32	
LA diameter (mm)	44 $\pm$ 4	43 $\pm$ 8	
Non-sustained VT	6/8 (75%)	30/44 (68%)	
Unexplained syncope	0/8 (0%)	8/44 (18%)	
Family history of SD	3/8 (38%)	13/44 (30%)	
Family history of multiple (≥ 2) SD in 1 st and 2 nd degree relative	2/8 (25%)	3/44 (7%)	
Abnormal BP to exercise	3/5 (60%)	9/32 (28%)	
Massive LVH (≥ 35 mm)	1/8 (12.5%)	3/44 (6.8%)	
AF	2/8 (25%)	9/44 (20%)	
Follow up period (years)	6.5 $\pm$ 0.5	6.2 $\pm$ 5.3	

ESC — European Society of cardiology; ACC — American college of cardiology; AHA — American Heart Association; LVH — Left Ventricular Hypertrophy; LVOT — Left Ventricular Outflow Obstruction; LA — left atrium; VT — ventricular tachycardia; SD — sudden death; BP — blood pressure; AF — atrial fibrillation)