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

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Background Hypertrophic cardiomyopathy (HCM) is the most common genetic cardiac disorder, and the most common cause of sudden cardiac death (SCD) in the 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.

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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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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