CR (Figures 1-2). Increase in LVEF 5% had a significant impact on survival both on patients that underwent aortic valve intervention (log rank p=0.03) and those who underwent medical management (log rank p=0.01), as opposed to presence of SVFR (log rank p=0.234 and p=0.708 respectively).

Conclusions During LDDSE in LFLGAS normalised FR, not SVFR, is a better determinant of TSAS, whereas assessment of LVEF change instead of SVFR determines CR.

Valve Disease/Pericardial Disease/ Cardiomyopathy

125 EVALUATION OF TITIN CARDIOMYOPATHY IN PATIENTS WITH DILATED CARDIOMYOPATHY REVEALS A BLUNTED HYPERTROPHIC RESPONSE, AN EARLY ARRHYTHMIC RISK AND A SIGNIFICANT INTERACTION WITH ALCOHOL

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Background Titin truncating variants (TTNtv), found in~10%–20% of dilated cardiomyopathy (DCM), are notable for variable penetrance and expressivity. We evaluated whether TTNtv DCM patients had distinct phenotypic features, which may influence disease outcomes.

Methods Prospectively recruited DCM patients underwent comprehensive clinical evaluation, cardiac MRI and TTN sequencing.

Results Overall, 572 subjects, 388 men (67.8%), mean age 53.5±14.4 years, were recruited. TTNtv were found in 56 patients (9.8%) and were associated with lower indexed LV mass (LVMi) and thinner LV walls, in the absence of differences in LV volumes after adjusting for clinical covariates (LVMi 83.1 vs. 94.0g/m^2, p=0.008; max. LV wall thickness 9.1 vs. 10.1 mm, p=0.003; indexed LV end diastolic volume 122.7 vs. 131.3mls/m^2, p=0.07).

196/572 patients (34%) had atrial fibrillation or ventricular arrhythmia at recruitment. Adjusting for age, gender, baseline ventricular function, and left atrial volume, TTNtv independently associated with early arrhythmic burden (adjusted OR 2.90, CI 1.48 to 5.77, p=0.002). On sensitivity analysis, this association remained significant after exclusion of 12 patients with rare LMNA variants (adjusted OR 2.88, CI 1.45 to 5.81, p=0.003).

TTNtv alone did not predict LVEF but in the presence of a history of alcohol excess, LVEF was reduced by 17.5% (p<0.0001), independently of other predictors of LVEF (age, gender, NYHA class, mid-wall fibrosis, and a family history of NIDCM).

Conclusions These data demonstrate that DCM due to TTNtv is associated with a blunted hypertrophic response, highlighting possible disease mechanisms. We also demonstrate that TTNtv are independently predictive of early arrhythmia and show a significant gene-environmental interaction between TTNtv and alcohol, which may inform risk stratification.

126 DOES HYPERTROPHIC CARDIOMYOPATHY GENOTYPE AFFECT TISSUE DOPPLER IMAGING PARAMETERS OVER 3-YEAR FOLLOW-UP PERIOD?

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Introduction Recent studies suggest that tissue Doppler imaging (TDI) have prognostic value in hypertrophic cardiomyopathy (HCM). We aimed to identify if there was a difference in rate of deterioration of TDI values of diastolic function and longitudinal systolic function according to genotype status.

Methods Aprospective, single-centre observational study over 33 months was undertaken. Twenty-six HCM patients were assigned a group according to genotype status (G+ve (n=14) vs G-ve (n=12)). Differences in baseline and follow-up TDI

Abstract 126 Table 1 Changes in TDI parameters according to HCM genotype

Value		Baseline, mean (SD)	Follow up, mean (SD)	Change	p value for Change (G+ve vs G-ve)
Medial E	G+ve	6.33 (0.50)	7.85 (0.75)	+1.52	**0.002**
	G-ve	6.57 (0.48)	5.06 (0.46)	−1.51	
Lateral E	G+ve	9.68 (0.84)	9.86 (0.84)	+0.18	**0.034**
	G-ve	9.13 (0.73)	6.94 (0.73)	−2.19	
Average E	G+ve	8.00 (0.61)	8.86 (0.75)	+0.86	**0.003**
	G-ve	7.85 (0.53)	6.00 (0.50)	−1.85	
Medial S	G+ve	7.57 (0.48)	8.11 (0.69)	+0.54	0.200
	G-ve	7.19 (0.50)	6.68 (0.34)	−0.51	
Lateral S	G+ve	9.89 (1.01)	8.65 (0.61)	−1.24	0.387
	G-ve	9.15 (0.62)	7.76 (0.43)	−1.39	
Medial E/E	G+ve	12.59 (1.66)	9.73 (1.07)	−2.86	**0.004**
	G-ve	14.98 (2.09)	18.00 (2.58)	+3.02	
Lateral E/E	G+ve	8.33 (1.04)	7.23 (0.79)	−1.10	0.0003
	G-ve	9.98 (1.31)	14.05 (2.25)	+4.07	
Average E/E	G+ve	10.46 (1.31)	8.48 (0.87)	−1.98	**0.0002**
	G-ve	12.48 (1.66)	16.02 (2.30)	+3.54	

parameters were compared using an independent samples t-test.

Results The mean follow-up was 32.8±2.5 months. A standard 16-gene panel was performed in all patients. Fourteen patients (54%) were G+ve (4 MYBPC3, 3 M7H7 and 7 others). G +ve patients were more likely to be male (57%), younger (39 vs 66 years, p=0.0003) and have a family history of HCM (43 vs 8%, p=0.048). There was no difference in baseline diastolic septal diameter (G+ve 16.1±5.3 mm vs G-ve 17.4 ±7.3 mm, p=0.6) or ejection fraction (G+ve 64±2% vs G-ve 63±2%, p=0.697).

TDI parameters during follow up revealed the magnitude of deterioration in medial E, medial E/E, lateral E and lateral E/E was greater in G-ve compared to G+ve patients. There was also a trend for a greater decrease in medial and lateral S in G-ve patients.

During follow up we did not observe any significant differences in the change of LA diameter (G+ve+3.29(±6.29) vs G-ve+3.08(±5.50), p=0.932) or LV max wall thickness (G+ve −1.07±5.05 vs G-ve+0.20±2.61, p=0.440). Overall changes in sudden cardiac death (SCD) risk scores remained similar (G+ve+0.27(±0.48) vs G-ve+0.75(±0.64), p=0.378). There were no sudden cardiac deaths or shockable events.

Conclusion In this single-centre HCM clinic, despite no change in SCD risk scores or measurements of cardiac morphology, G-ve patients have a greater rate of deterioration of TDI parameters with a significant reduction in diastolic filling and a trend towards reduction in longitudinal systolic function. Although G-ve patients were older than G+ve, the rate of decline in TDI velocities over a 33 month period is greater than what would be expected in the normal ageing population.

127 RELATIONSHIP BETWEEN PLASMA CONCENTRATIONS OF B-TYPE NATRIURETIC PEPTIDE AND EXERCISE CAPACITY IN HYPERTROPHIC CARDIOMYOPATHY

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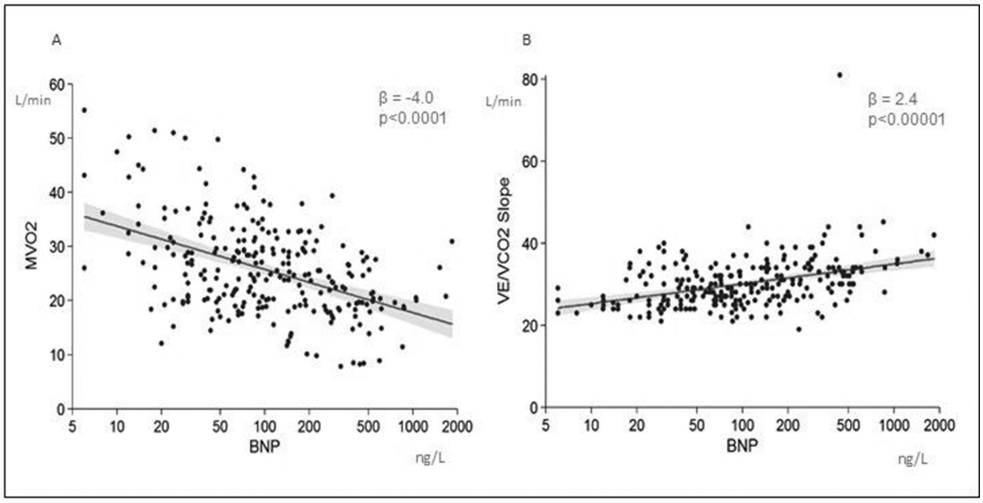
Background Hypertrophic cardiomyopathy (HCM) is characterised by increased left ventricular wall thickness leading to exercise intolerance and heart failure. Peak oxygen consumption (peak VO2) on cardiopulmonary exercise testing (CPET) is often used as a functional marker alongside B-type natriuretic peptide (BNP). However, CPET is complex and time-consuming, access may be limited, and peak VO2 can be influenced by physical deconditioning and motivation. Our aims were to determine the association between BNP and submaximal exercise parameters, ventilatory efficiency (VE/VCO2) and anaerobic threshold; and to determine whether f these were effective surrogate markers of functional status in patients with HCM.

Methods We retrospectively reviewed data for all patients with HCM in our local cardiomyopathy service. A diagnosis of HCM was based on LV hypertrophy >15 mm as per current ESC guidelines. Patients underwent contemporaneous echocardiography, CPET using maximal treadmill ergometry and BNP measurement at sequential annual clinic visits [1 (18%), 2 (52%), 3 (20%) or ≥4 visits (10%)] yielding data from 252 observations over 8 years for 119 individual patients. Univariable and multivariable linear regression was used to investigate variables associated with the results of CPET. Robust standard errors were used to correct for multiple observations from the same patients.

Results In our cohort of 119 patients (mean age 49±17 years, 66% men), mean LVEF was 75±7%, maximal wall thickness was 19±5 mm, and left ventricular outflow tract obstruction was present in 15%. The mean peak V02 was 26±8 L/min, mean VE/VCO2 slope was 30±6 and median BNP was 93.5 ng/L (IQR 43.5–232.5). On univariable analysis, several markers were found to predict peak VO2; age (β^2=−4.5; 95% CI −6.1 – −3.0; p<0.00001), male sex (β^2=5.2; 95% CI 2.3 – 8.1; p<0.001), LV ejection fraction (β^2=−2.1; 95% CI −3.6 – −0.6; p<0.01), BNP (β^2=−4.0; 95% CI −5.8 – −2.2; p<0.0001) and echocardiographic markers of diastolic dysfunction, for example, E/E lateral ratio (β^2=−3.4; 95% CI −4.8 – −2.0; p<0.00001). Predictors of VE/VCO2 were age (β^2=1.5; 95% CI 0.6 – 2.3; p<0.01), BNP (β^2=2.4; 95% CI 1.5 – 3.4; p<0.00001) and similar echocardiographic markers of diastolic dysfunction. Multivariable analysis revealed that BNP was associated with peak VO2 (β^2=−2.0; 95% CI −3.0 – −1.1; p<0.0001) but also age, male sex,



Abstract 127 Figure 1 Association between BNP and peak VO_2(A) and VE/VCO_2 (B) inpatients with HCM.