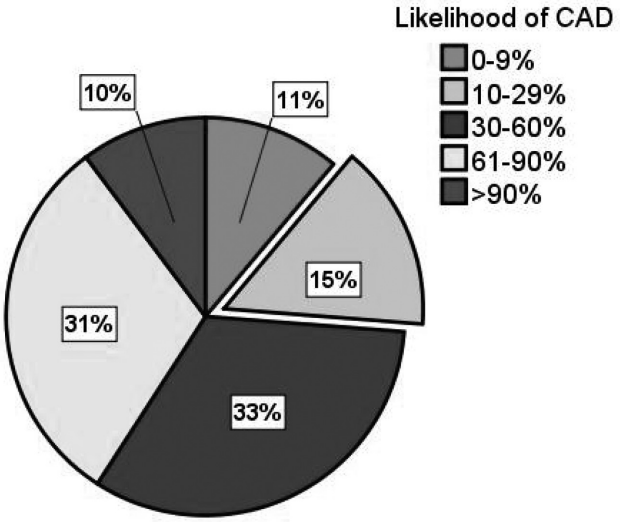

Abstract 133 Figure 1 Estimated likelihood of coronary artery disease according to the NICE guideline (CG95)

134 NON-INVASIVE INTERROGATION OF MYOCARDIAL DISARRAY IN HYPERTROPHIC CARDIOMYOPATHY

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10.1136/heartjnl-2016-309890.134

Introduction Hypertrophic cardiomyopathy (HCM) is an inheritable cardiomyopathy characterised by left ventricular hypertrophy (LVH). In contrast to other aetiologies of LVH, the HCM myocyte microstructure displays disarray, the ability to detect disarray non-invasively could therefore assist the diagnosis in equivocal cases.

In vivo Diffusion tensor imaging (DTI) is a novel cardiovascular magnetic resonance technique, which exploits the fact that intra-myocardial water diffusion reflects the shape and orientation of the underlying microstructure. DTI can characterise the myocardial microstructure with novel parameters: Myocyte orientation can be depicted by the helical angle gradient (HAG); the degree of myocardial organisation can be assessed by fractional anisotropy (FA); and mean diffusivity (MD) quantifies the ease of myocardial water passage. We sought to determine whether DTI could detect myocardial micro-structural differences compared with patients with hypertension and healthy controls.

Methods We recruited 25 patients with HCM, 13 patients with HTN and 14 healthy, age matched controls for DTI at 3T. Three short axis mid-ventricular slices were acquired during the systolic pause, with a diffusion-weighted stimulated echo sequence, as previously described. Data was post-processed to calculate global FA, HAG and MD values. Statistical comparison between the 3 cohorts was made with one way ANOVA, followed by students t-tests were ANOVA was significant.

Results There was no significant difference in FA between cohorts (HCM: 0.44 ± 0.05, HTN: 0.47 ± 0.06 and Control: 0.46 ± 0.05, p = 0.20). MD was greater in HCM compared to controls (ANOVA p = 0.002, HCM 1.07 ± 0.13 vs Controls: 0.91 ± 0.13×10–3mm^{2}s^1, p < 0.01), however there was no difference in MD between HCM and HTN (HCM: 1.07 ± 0.13 vs HTN: 0.97 ± 0.14×10–3mm^{2}s^1, p = 0.15). The HAG was less in HCM compared to both HTN and Controls (HCM: -6.2 ± 1.0, HTN: -7.4 ± 0.9 and Controls: -8.6 ± 0.8, p < 0.01). The HAG was plotted against maximal wall thickness for all patients combined: correlation coefficient 0.723, p < 0.001.

Summary DTI was unable to detect a significant disarray in HCM via fractional anisotropy. Explanations include the limited resolution of our sequence, the small study sample and potential regionality of disarray. The higher MD observed in HCM compared to controls may be attributable to a greater extracellular volume as a result of fibrosis. Technical factors such as myocardial strain and heterogeneity of SNR may also have contributed. The HAG was correlated with maximal wall thickness, with reduction in the rate of angular change as the wall thickness increases.

Conclusion DTI provides novel myocardial tissue characterisation, however in its current form it is unable to detect disarray in HCM. Further assessment following technical development is required.

135 DETECTING PROGRESSION OF DIFFUSE INTERSTITIAL FIBROSIS IN ALSTROM SYNDROME

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10.1136/heartjnl-2016-309890.135

Introduction Alstrom Syndrome (ALMS) is a rare inherited disorder caused by a mutation in the ALMS1 gene. The syndrome is a multi-system disorder with exaggerated features of the metabolic syndrome and although rare, provides a monogenic model for end-organ fibrosis and as a paradigm for the effects of severe metabolic syndrome. Adults with ALMS have a high risk of death from heart failure in their twenties due to a cardiomyopathy which (in the small post-mortem series available) is characterised by coarse fibrosis on histology. Our previous work has identified expansion of the extracellular space (ECV) consistent with diffuse interstitial fibrosis in over half of asymptomatic ALMS patients compared to controls. The aim of this study was to investigate the longitudinal change in ECV and assess the impact on ventricular structure and function.

Methods A prospective longitudinal cohort study of patients attending the national service for ALMS at the Centre for Rare Disease in Birmingham from 2012. At referral and on annual follow up, all subjects underwent comprehensive LV and RV assessment with cardiac MRI (CMR 1.5T Siemens Avanto). The presence of diffuse interstitial myocardial fibrosis was assessed using native myocardial T1 relaxation mapping and extracellular volume (ECV) in the LV septum (MOLLI) using cvi42 (Circle Cardiovascular Imaging). Coarse replacement fibrosis was assessed using standard late gadolinium enhancement imaging.

Results In total 14 patients (male gender 71%, age 28 ± 8years) had baseline and follow up data (median 1.7 [1.1–2.8] years). CMR data is presented in Table 1. The native LV myocardial T1 values and ECV were increased in the septum at basal and mid levels at follow up. Left ventricular mass increased (54 ± 9 g/m^2 vs. 62 ± 12 g/m^2) but with a reduction in septal myocardial intracellular volume (ICV 0.74 ± 0.06 vs. 0.68 ± 0.04, p < 0.05) suggesting ECV expansion rather than myocyte hypertrophy was the driver. There were no differences in LV or RV volumes or RVEF. Four patients