Objectives We used CMR with extracellular volume fraction (ECV) measurement to characterise cardiac involvement in relation to outcome in ATTR amyloidosis. 

Methods Subjects comprised 263 patients with cardiac ATTR amyloidosis corroborated by grade 2–3 99mTc-DPD cardiac uptake, 17 with suspected cardiac ATTR amyloidosis (grade 1 99mTc-DPD) and 12 asymptomatic individuals with amyloidogenic transthyretin (TTR) mutations. Fifty patients with cardiac AL amyloidosis acted as disease controls.

Results In contrast to AL amyloidosis, asymmetric septal hypertrophy was present in 79% of ATTR patients (70% sigmoid septum and 30% reverse septal curvature), whilst symmetric left ventricular hypertrophy (LVH) was present in only 18%; 3% of patients has no LVH. In patients with cardiac amyloidosis, the pattern of LGE was always typical for amyloidosis (29% subendocardial, 71% transmural) including right ventricular LGE (96%). 63 patients died during follow-up (19 ±14 months). ECV independently correlated with mortality and remained independent after adjustment for age, N-terminal pro-brain natriuretic peptide, ejection fraction, E’/E and left ventricular mass index (hazard ratio, 1.164; 95% confidence interval, 1.066–1.271; p<0.01).

Conclusions Asymmetric hypertrophy, traditionally associated with hypertrophic cardiomyopathy, is the commonest pattern of ventricular remodelling in ATTR amyloidosis. LGE imaging is typical in all patients with cardiac ATTR amyloidosis. ECV correlates with amyloid burden and provides incremental information on outcome even after adjustment for known prognostic factors.

Abstract 026 DIMENSIONAL FLOW Cardiovascular Magnetic Resonance: Two-Centre, 1.5T, Phantom and In-Vivo Validation Study

Background Validation of four-dimensional (4D) flow CMR accelerated acquisition methods is needed to make them more robust for clinical applications. Our aim was to compare three widely-used acceleration methods in 4D flow CMR: 4D segmented fast-gradient-echo (4D- turbo-field-echo, 4D-TFE), 4D non-segmented gradient-echo with echo-planar imaging (4D- EPI) and 4D-k-t Broad-use Linear Acquisition Speed-up Technique accelerated TFE (4D-k-t BLAST).

Methods CMR was performed in two institutions on identical 1.5T systems. Acceleration methods were compared in static/flow phantom setup.

Abstract 026 Figure 1 Illustration to demonstrate static and pulsatile flow phantom setup.
The CMR protocol included: cines, 2D phase contrast (PC) at the aortic valve (AV) and mitral valve (MV) and three whole-heart free-breathing (no respiratory motion correction) 4D flow CMR pulse sequences. Field-of-view, slices, phases (30), voxel size and VENC were the same for each subject. In volunteers, net acquisition time for each 4D flow sequence was recorded, as well as a visual grading of image quality on a four-point scale: 0, no artefacts to 3, non-evaluable.

Results For the pulsatile phantom experiments, the mean error against the reference flow by time beaker measurements for 4D-TFE was 4.9%±1.3%, for 4D-EPI 7.6%±1.3% and for 4D-k-t BLAST 4.4%±1.9%. In vivo, acquisition time was shortest for 4D-EPI at 7 min59s±2 min30s. 4D- EPI and 4D-k-t BLAST had minimal artefacts, while for 4D-TFE, 40% of AV and MV assessments were non-evaluable because of phase dispersion artefacts. Peak velocity assessment using 4D-EPI demonstrated best correlation to 2D PC (AV: r=0.78, p<0.001; MV: r=0.71, p<0.001). Coefficient of variability (CV) for net forward flow (NFF) volume was least for 4D-EPI (7%) (2D PC:11%, 4D-TFE: 29%, 4D-k-t BLAST: 30%, respectively) (Figure 2, 3).

Conclusion Of the three 4D flow CMR methods tested, 4D-EPI demonstrated the least susceptibility to artefacts, good image quality, modest agreement with the current reference standard for peak intra-cardiac velocities and the highest consistency of intra-cardiac flow quantifications.

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Abstract 026 Figure 2 Bland Altman analysis and scatter plots for the assessment of peak velocity using all the acceleration methods.

A22

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