Detection of progressive hypertrophy and apical aneurysm formation in symptomatic apical hypertrophic cardiomyopathy

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Introduction
Age related penetrance of hypertrophic cardiomyopathy (HCM) can result in so called "late-onset HCM", but significant increases in existing ventricular hypertrophy is rare in older ages. Cardiac magnetic resonance (CMR) imaging can accurately identify phenotypic progression in patients with apical HCM, identifying discreet apical aneurysm and guide subsequent therapies.

Description of the clinical case
A 73-year-old gentleman with apical HCM underwent serial CMR imaging between 2014 and 2017, prompted by worsening symptoms of dyspnoea and postural dizziness. The 2017 imaging demonstrated a significant increase in maximal wall thickness at the lower mid-cavity and apical levels, increasingly diffuse presence of late gadolinium enhancement (LGE) in the affected segments, and for the first time a small apical aneurysm that was not clearly identified on two-dimensional echocardiography. The development of this more severe phenotype was accompanied by worsening symptoms of advanced heart failure.

Discussion
Patients with apical HCM may develop a discreet apical chamber progressing to apical aneurysm, due to systolic contractile persistence, localised ischaemia, and evidence of replacement fibrosis inferred by LGE. The continued radial hypercontractility at the mid-level ventricle provides the substrate for left ventricular mid-cavity obstruction (LVMCO) and development of even higher pressures at the dyskinetic apical chamber. Patients with LVMCO often experience severe drug-refractory symptoms, with limited therapeutic options available beyond medical therapy. Use of CMR to accurately define the phenotypic progression of severe hypertrophy and LGE expansion in this patient contributed to a re-evaluation of risk of sudden cardiac death. On this basis, the patient was offered a primary prevention implantable cardioverter-defibrillator (ICD) which they accepted. During the ICD implant, a haemodynamic pacing study was performed to see if the introduction of localised dyssynchrony via pacing from either right or left ventricle could attenuate the LVMCO gradient that was considered to be a significant contributor to exertional symptoms (Figure 1).

Conclusion and implications/lessons to clinical practice
CMR is an important tool for the assessment of apical HCM having implications for both sudden cardiac death risk stratification and symptom management strategy. In this case of apical HCM with mid-cavity obstruction and apical aneurysm, CMR imaging helped to define the phenotype and plan invasive therapy using LV pacing in a severely symptomatic patient. A randomised blinded cross-over trial of LV pacing in this population is currently recruiting to help determine if this is a viable treatment option in a group that are otherwise difficult to manage.
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Figure 1: 3 chamber image demonstrating severe hypertrophy and small apical aneurysm (A), diffuse LGE at the apex (B), and near circumferential perfusion defect in the short axis (C). The patient was referred for ICD implantation and haemodynamic pacing study: LV pacing onset, reduction in mid-cavity gradient and maintenance of aortic pressure (D) and offset (E) with immediate return of significant intracavity gradient.