1 COMPARISON OF RIGHT VENTRICULAR (RV) SIZE AND FUNCTION BY TWO DIMENSIONAL ECHOCARDIOGRAPHY (2DTTE) AND CARDIAC MAGNETIC RESONANCE IMAGING (CMR)

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The aim of this study was to compare the right ventricular diameter (RVD1) and tricuspid annular plane systolic excursion (TAPSE) measured by 2DTTE with volumes and EF derived from CMR.

Method and results We studied 50 patients (mean age 53 ± 17 years, 36 males and 14 females) who underwent TTE and CMR within 6 months of each other. RVD1 and TAPSE were measured by 3 independent observers (inter observer variability = 5.6% (95% CI: 4–6.8%). RV volumes were measured by 2 independent observers blinded to the TTE data (inter observer variability =5% (95% CI: 4–6.8%). For RVD1, an upper reference value of 4.2 cm was used to indicate RV dilatation and for TAPSE a lower reference value of 1.6 cm was used for systolic impairment as per current recommendations. Upper reference value for end diastolic volume (EDV) indexed to BSA, age and gender was used as the CMR cut off for RV dilatation and lower reference value for EF based on age and gender was used for systolic impairment.

RVD1 showed a statistically significant correlation with RVEDV ($r = 0.475$, $p < 0.01$). The sensitivity and specificity of RVD1 >4.2 cm for predicting RV dilatation was 57% and 83% respectively. A cut off value of RVD1 >4.1 cm improved the sensitivity to 71% with no change in specificity (ROC analysis: AUC = 0.81). TAPSE did not correlate with EF ($r = 0.174$, $p = 0.248$). The sensitivity and specificity of TAPSE <1.6 cm for predicting systolic impairment was 81% and 13% respectively (ROC analysis: AUC 0.43). Adjusting the cut off of TAPSE did not improve this.

Conclusions Our data suggests that the current value of RVD1 > 4.2 cm is a poor predictor of RV dilation and lowering the value to 4.1 increases the sensitivity. TAPSE does not correlate with EF by CMR and is a poor predictor of global RV systolic impairment at any cut off value.

2 ROLE OF CARDIAC MAGNETIC RESONANCE (CMR) IN THE IDENTIFICATION OF ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY/DYSPLASIA (ARVC/D) PHENOCOPIES

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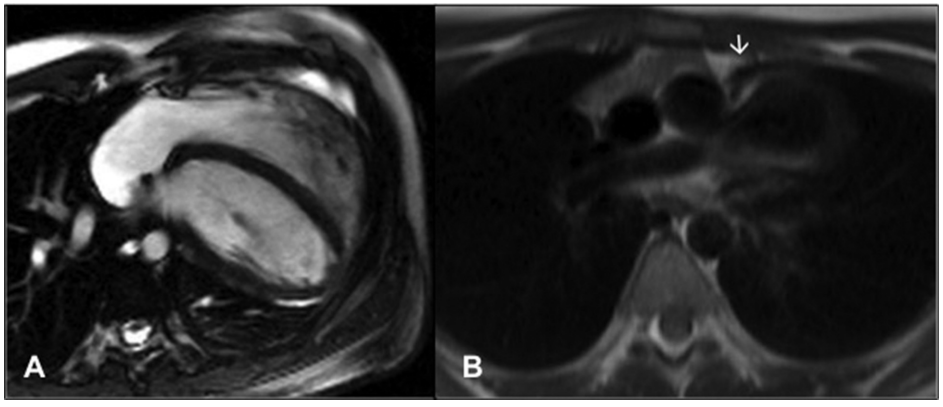
Introduction ARVC/D is an autosomal dominant genetic heart disease. As part of the diagnostic tools included in the 2010 diagnostic Task Force Criteria, patients are often referred to CMR to rule out the diagnosis. However, the diagnosis is often challenging due to pathologies mimicking ARVC/D.

Aim To assess the prevalence of ARVC/D phenocopies in patients referred to CMR for suspected ARVC/D.

Methods We retrospectively analysed the registry data of patients with suspected ARVC/D referred to CMR in a large UK tertiary centre from January to December 2014. We identified 125 patients (56% male, median age 40 years) with suspected ARVC/D on the basis of symptoms and clinical presentation, family history of ARVC/D or sudden cardiac death, abnormal electrocardiogram or transthoracic echocardiography. A comprehensive CMR protocol (including cine and late gadolinium enhancement sequences) was performed in all patients.

Results ARVC/D phenocopies were identified in 12 patients (9.6%): 5 patients had ischaemic heart disease and 7 had non-ischaemic heart disease (Table 1). In the latter group, congenital absence of pericardium (Figure 1), idiopathic dilated cardiomyopathy, left ventricular non compaction, arrhythmogenic left ventricular cardiomyopathy (ARVC/D variant), anomalous venous return, atrial septal defect with left to right shunting, and asymmetric pectus excavatum distorting right ventricular (RV) morphology were identified.

Conclusions CMR shows a remarkable role, both in diagnosing ARVC/D and ruling out its mimics. Correct diagnosis of the underlying pathology in patients with suspected ARVC/D is fundamental, given the non-negligible prevalence of phenocopies (9.6% in our population) and its subsequent impact on clinical management.



Abstract 2 Figure 1 Typical features of congenital absence of pericardium: heart displacement within the chest, with distorted RV morphology (A), and left lung interposition between the aorta and the pulmonary artery (B, arrow).