IMAGING VIGNETTE

CMR for the Diagnosis of Right Heart Disease

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RIGHT HEART DISEASE CAN PRESENT A CLINICAL DIAGNOSTIC CONUNDRUM as a number of pathologies can present with a variety of often nonspecific symptoms. Transthoracic echocardiography (TTE) remains the main noninvasive diagnostic imaging test for right heart disease, yet it has several limitations. Visualization of the right ventricular (RV) free wall and its trabeculations by TTE can be difficult, with no accurate method of quantifying RV volumes and function. Cardiac magnetic resonance (CMR) allows more reliable assessment of the right heart because of its unrestricted field of view and free and reproducible imaging planes. Therefore, CMR is considered the reference standard for the calculation of right heart volumes, mass, and function. In addition, the multiparametric properties of CMR allow for tissue characterization to aid the differential diagnosis of right heart pathologies, as demonstrated in this case series (Figs. 1, 2, 3, 4, 5, and 6).

Figure 1. Case #1
A 54-year-old asymptomatic male was referred to cardiology following sudden cardiac death in a first-degree relative. An echocardiogram suggested a dilated right heart, but views were suboptimal. Cardiac magnetic resonance (CMR) was requested to rule out arrhythmogenic right ventricular cardiomyopathy (ARVC). Cine images in short axis (SA) (A) and horizontal long axis (HLA) (B, Online Video 1) orientations revealed a dilated (end-diastolic volume index [EDVi]: 133 ml/m²) and severely impaired right ventricular (RV) (ejection fraction [EF]: 38%) with regional thinning and akinesia of the RV free wall (B, arrow). Late-gadolinium enhanced (LGE) images showed corresponding areas of hyperenhancement, consistent with fibrous replacement (C and D, arrow). Combined with the family history, the CMR findings allowed for a diagnosis of ARVC to be made on the basis of 2 major criteria. The 2010 Task Force diagnostic criteria for ARVC include RV EDV of >110 ml/m² (males) and >100 ml/m² (females) or a reduced EF ≤40% as major diagnostic criteria. Although fibrofatty replacement of the myocardium may be shown by T1-weighted and LGE CMR (as in this case) with a typical regional pattern, associated wall motion abnormalities, and thinning of the RV myocardium in advanced cases, it does not form part of the diagnostic criteria.

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A 66-year-old male presented with a non-ST-segment elevation acute coronary syndrome. Coronary angiography showed a chronic total occlusion (CTO) of the right coronary artery (RCA) with nonobstructive atheroma in the left coronary system. CMR was requested to determine inferior wall viability before an attempt at revascularizing the RCA. Cine images showed no regional wall motion abnormalities (RWMA) in the left ventricle (LV) (A and B), but reduced longitudinal function of the RV free wall and right ventricular outflow tract. A corresponding area of increased signal intensity on T2-weighted imaging (C, arrow) was suggestive of myocardial oedema and the LGE images showed subendocardial hyperenhancement indicative of an RV infarction (D, red arrow). The inferior LV wall was viable (blue arrow) and the patient had successful CTO revascularization. CMR is a very useful test for the detection of RV infarction, a diagnosis that is frequently missed, but is associated with worse outcome. The typical pattern of acute RV infarction is impaired RV function, a RWMA with corresponding oedema and hyperenhancement on T2W and LGE images respectively. Abbreviations as in Figure 1.

A 44-year-old male presented with chest pain and palpitations. A 24-h electrocardiogram (ECG) had shown episodes of ventricular tachycardia, but subsequent coronary angiography was normal. A transthoracic echocardiography (TTE) suggested thickening of the RV but no other abnormalities. On cine CMR images, the RV wall was clearly hypertrophied (A, SA; B, HLA arrows). RV volumes (Online Video 2) confirmed normal cavity size (EDV 85 ml/m²) and normal systolic function (EF 60%). LGE images showed extensive areas of hyperenhancement in the RV wall (C, SA arrow) and areas of the LV septum (D, HLA arrow). Mediastinal lymph nodes were present on the T1-weighted black blood transaxial stack (not shown). A diagnosis of sarcoidosis was suggested and subsequently confirmed on high-resolution computed tomography and endomyocardial biopsy. Cardiac involvement in sarcoidosis rarely produces symptoms (5%) but can result in sudden cardiac death due to ventricular arrhythmias. The diagnosis of cardiac sarcoidosis by echocardiography is challenging, and LGE CMR in particular is a useful tool to detect this complication. Predominant involvement of the RV in cardiac sarcoidosis as in this case is rare, but shows typical findings of preserved RV function with widespread infiltrates on LGE. Abbreviations as in Figures 1 and 2.

A 38-year-old male was admitted with chest pain and breathlessness. Investigations showed anterior T-wave inversion on the ECG and a raised troponin I (0.19 ng/ml). Coronary angiography demonstrated minor left anterior descending atheroma. Inpatient CMR was requested before echocardiography to distinguish between a plaque event and myocarditis. The first ventricular long axis (VLA) cine image demonstrated a focal area of low signal intensity in the right pulmonary artery (RPA), suggestive of a thrombus (A, arrow). On further imaging, the RV was found to be dilated with reduced systolic function with a prominent septal bounce suggesting RV pressure overload (SA, B, arrow Online Video 3). T2-weighted (C) images showed no evidence of RV or LV myocardial edema and LGE images were normal (not shown). Pulmonary artery cine images revealed further mobile masses in the right (D, arrow), and left (Online Video 4) pulmonary arteries. A diagnosis of bilateral pulmonary emboli was made, and anticoagulation therapy was initiated. CMR is a frequently used test for the differentiation between infarction and myocarditis, but in this case, an unexpected diagnosis of pulmonary embolus (PE) was made. The typical findings of acutely raised RV pressure can also be demonstrated with echocardiography, but CMR as shown can directly visualize the embolic source. A pulmonary magnetic resonance (MR) angiogram can further aid the diagnosis, but was not performed in this patient because the cine images were diagnostic. Abbreviations as in Figures 1, 2, and 3.
A 23-year-old female was referred with exertional breathlessness. TTE identified a dilated RV with no obvious cause. CMR showed an enlarged RV (RVEDVi 115 mls/m²) (A), and a sinus venous atrial septal defect (ASD) measuring 7 x 11mm (B, arrow). Further multiplanar imaging showed anomalous drainage of the right pulmonary vein into the right atrium (C, arrow), which was further demonstrated by contrast-enhanced MR angiography (D, arrow). Contrast velocity imaging of aortic and pulmonary arteries (not shown) revealed an intracardiac left-to-right shunt by stroke volume difference of 2.2. CMR is a frequently used test to further characterize ASD following initial echocardiographic assessment, in particular to identify associated pathology such as anomalous pulmonary venous drainage and to quantify shunt ratios to plan therapy. Abbreviations as in Figures 1 and 3.

**Figure 6. Diagnostic Flow Chart for the Investigation of Right Heart Disease.**

CTPA − computed tomography of the pulmonary arteries; EGE − early gadolinium enhancement; MRA − pulmonary magnetic resonance angiography; MRI − magnetic resonance imaging; PV − pulmonary vein; RWM − regional wall motion; T2 STIR − short tau inversion time; VLA − ventricular long axis; other abbreviations as in Figures 1, 2, and 4.

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**APPENDIX**

For supplementary videos and their legends, please see the online version of this article.