Dilated right cardiac chamber delusion solved by cardiovascular magnetic resonance

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A previously healthy 59-year-old woman complained of palpitations and breathlessness. Two relatives had sudden cardiac death, including a sibling with dilated cardiomyopathy. Her electrocardiogram showed sinus rhythm and incomplete right bundle branch block. Transthoracic echocardiography (TEE) revealed a dilated right ventricle (RV) with normal systolic function and moderate tricuspid regurgitation (estimated pulmonary artery systolic pressure 45 mmHg); no other alterations were found.

Suspicion of arrhythmogenic RV cardiomyopathy (ARVC) was raised and a cardiovascular magnetic resonance (CMR) was performed. Steady-state free precession CINE images confirmed a dilated RV (indexed end-diastolic volume 159 ml/m^2), with no regional akinesia or dyskinesia (Fig. 1A and 1B). No focal myocardial fibrosis was detected (Fig. 1C). Phase contrast sequences acquired at the aortic root and pulmonary trunk uncovered a significant left-to-right cardiac shunt (Qp / Qs = 2). CINE sequences directed to the inter-auricular septum revealed an ostium secundum atrial septal defect (ASD), measuring approximately 13 – 11 mm with a shunt of 39 ml (Fig. 2). MR-angiography identified partial anomalous pulmonary venous connec-

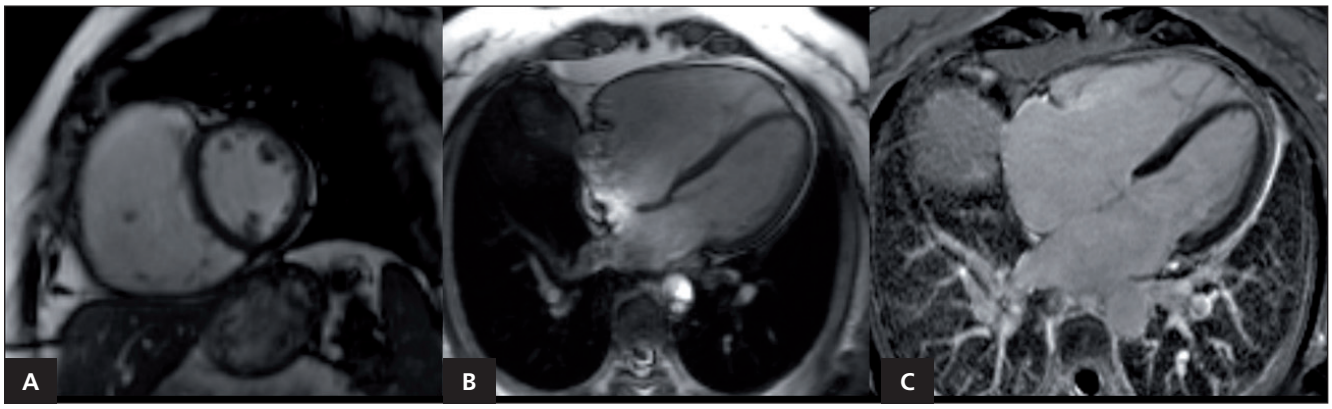

Fig. 1 – CINE imaging in short-axis (A) and horizontal long-axis (B) end-diastolic image frames. Severe dilation of the RV is noted (256 ml; 159 ml/m^2) without regional akinesia or dyskinesia or systolic dysfunction (EF: 62%). Late gadolinium enhancement PSIR imaging (C) shows no evidence of myocardial fibrosis. EF – ejection fraction; PSIR – phase-sensitive inversion recovery; RV – right ventricle.

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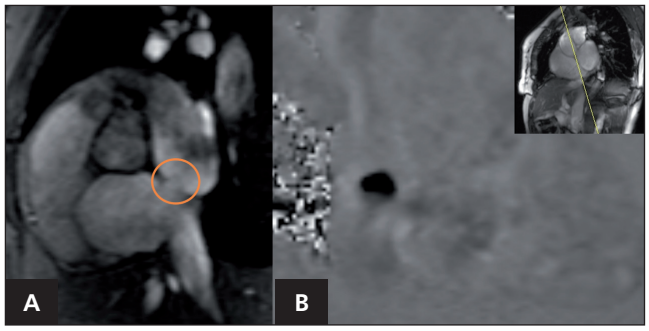

Fig. 2 – CINE imaging in sagittal oblique axis (A) showing an ostium secundum ASD (circle). Through-plane PC-MRI imaging demonstrates significant flow through the ASD. ASD – atrial septal defect; PC-MRI – phase-contrast magnetic resonance imaging.

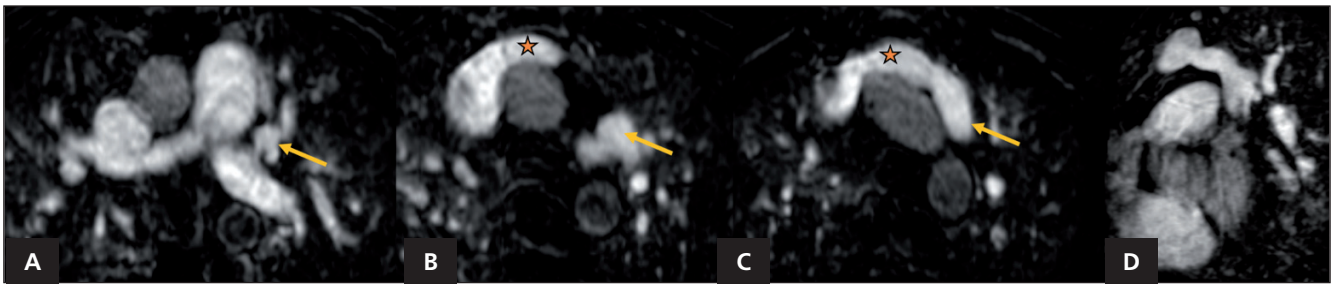

Fig. 3 – MR-angiography imaging in the axial plane (A–C) shows abnormal communication of the left superior pulmonary vein (arrows) to the left brachiocephalic vein (asterisk). Sagittal-oblique MPR (D) showing the partial anomalous pulmonary venous return. MPR – multiplanar reformation.

tion (PAPVC) of the superior left pulmonary vein draining in the left brachiocephalic vein (Fig. 3).

Right chambers dilatation is a common manifestation in several pathologies, namely ARVC and congenital heart disease with left-to-right shunting.[1] When first line echocardiography is equivocal or inconclusive, CMR can provide additional information.[2] In this particular patient, suggestive findings of anomalous drainage, such as dilated superior vena cava and fewer than four pulmonary veins connecting to the left atrium, were inconspicuous. Moreover, ASD – the most typical finding associated with PAPVC – was also unrecognized.

This clinical case highlights CMR use in the study of dilated right cardiac chambers, and the relevance of a systematic evaluation of Qp/Qs.

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