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Clinical Image

CLINICAL IMAGE

Rare congenital quadricuspid pulmonary valve stenosis evaluated by CMR

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CASE DESCRIPTION

A 41-year old Caucasian female with a history of congenital pulmonary stenosis diagnosed at the age of 6 months was referred for a cardiovascular magnetic resonance (CMR) scan to better evaluate the degree of pulmonary valve (PV) stenosis. She had been complaining of intermittent palpitations and limited exercise tolerance. Clinical examination revealed a harsh ejection systolic murmur at the left upper sternal border and a split second heart sound. Electrocardiogram showed incomplete right bundle branch block. Transthoracic echocardiogram showed moderate pulmonary stenosis with a peak velocity of 3 m/s, peak gradient of 37 mmHg and mean gradient of 22 mmHg; however, the PV was not well visualized.

CMR revealed the unusual appearance of a quadricuspid pulmonary valve (QPV) with four cusps of almost equal size (Fig. 1a). The leaflets were mobile with moderate-severely restricted central opening (valve area 0.9–1 cm^2; Supplementary Video 1) and moderate flow acceleration (Fig. 1b). There was also mild pulmonary regurgitation. The PV annulus was non-dilated. The main and left pulmonary arteries were moderately dilated (32 and 28 mm, respectively), with high-normal right pulmonary artery dimensions (21 mm). Biventricular volumes and function were normal. There was no evidence of other congenital cardiac or vascular abnormality.

QPV is a rare congenital abnormality, occurring in isolation or with other cardiac anomalies. QPV tends to be under-diagnosed as it is not commonly associated with significant clinical manifestations, and PV anatomy is often difficult to evaluate in adults with echocardiography. It is mainly discovered on post-mortem specimens with a higher than expected reported incidence of between 1 in 400 and 1 in 1000. QPV with severe stenosis is very rare [1]. Although cases with mildly stenotic quadricuspid pulmonary valves characterized with CMR have been previously reported [2–4], this is the first description of significant quadricuspid pulmonary valve stenosis evaluated by CMR.

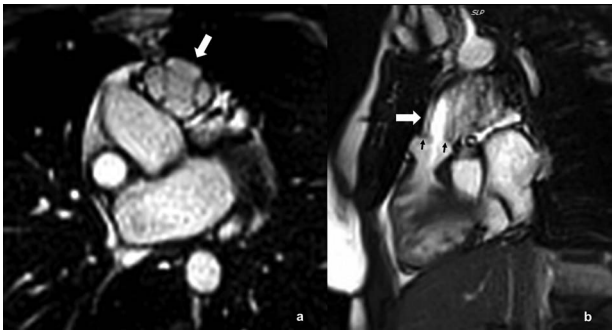

Figure 1: CMR steady-state free precession images of the pulmonary valve in short-axis view showing the four cusps in diastole (arrow, panel 1a), and of the right-ventricular outflow tract in sagittal view showing restricted pulmonary valve tips (small black arrows) with systolic flow acceleration into a dilated main pulmonary artery (white arrow, panel 1b).

SUPPLEMENTARY MATERIAL

Supplementary material is available at the *Journal of Surgical Case Reports* online.

CONFLICTS OF INTEREST STATEMENT

None declared.

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CONSENT

Written consent for submission and publication of this case report including images and associated text has been obtained from the patient.

GUARANTOR

The nominated Guarantor is Dr Chrysovalantou Nikolaidou.

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