Isolated partial anomalous pulmonary venous return with intact atrial septum: a rare but treatable cause of pulmonary hypertension in adults

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A 56-year-old woman presented with a 1-year history of progressive dyspnoea on exertion. Echocardiography and right heart catheterization demonstrated severe right ventricular enlargement and pulmonary hypertension (74/40 mmHg) without any evidence of atrial septal defect (ASD). Cardiac magnetic resonance (CMR) imaging was performed to identify possible causes. Cine MR showed severe right ventricular enlargement right ventricular end-diastolic volume index (RVEDVI) (160 mL/m²) with moderate systolic dysfunction right ventricular ejection fraction (RVEF) (35%) (Panel A, white arrows, and see Supplementary data online, Video S1). Gadolinium-enhanced MR angiography demonstrated partial anomalous pulmonary venous connections (PAPVC) with two right pulmonary veins draining into the superior vena cava (SVC) (Panel D, white arrows, and see Supplementary data online, Video S2) and the left superior pulmonary vein draining into the innominate vein (Panel E, white arrows, and see Supplementary data online, Video S2). Phase-contrast MR demonstrated 125 mL/beat of pulmonic flow (Panel B) and 56 mL/beat of aortic flow (Panel C) giving a Qp/Qs of 2.2. The patient underwent successful surgical repair with reimplantation of the left anomalous pulmonary vein into the left atrium. The anomalous right pulmonary veins were tunnelled through the right atrium and attached to the left atrium using an autologous pericardial patch.

Causes of right ventricular enlargement in adults include: pulmonary hypertension, pulmonary regurgitation (particularly in repaired Tetralogy of Fallot), tricuspid regurgitation, arrhythmogenic right ventricular cardiomyopathy, right ventricular infarction, ASD, and PAPVC.1,2 PAPVC has been described in 0.4–0.7% of autopsy series. In most cases, a right pulmonary vein drains to the SVC. Less frequently, there is drainage to the right atrium, innominate vein, inferior vena cava, or coronary sinus. PAPVC is associated with an ASD in 80–90% of cases, most commonly of the sinus venosus type. However, an isolated PAPVC with an intact atrial septum has been described and can lead to right ventricular enlargement and pulmonary hypertension in adults, particularly when more than one pulmonary vein is anomalous.2 Surgical repair is typically indicated in symptomatic patients with large shunts.2 Multimodality CMR can make the diagnosis and identify the anatomy and haemodynamic severity of this condition.

Supplementary data are available at European Heart Journal — Cardiovascular Imaging online.

References

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