A 35-year-old, previously healthy man, presented with a 6-month history of progressive exercise intolerance. Routine laboratory values and ECGs at rest and during exercise were inconspicuous. Trans-thoracic echocardiography hinted at moderate enlargement of right-sided cavities of the heart without apparent atrial septal defect (ASD).

Hence, cardiovascular magnetic resonance (CMR) imaging was performed to elucidate right-ventricular enlargement: on cine imaging right-ventricular end-diastolic volume amounted to 295 mL while regional and global RV-function were normal (Panel B, see Supplementary material online, Movie S1). Subsequent contrast-enhanced pulmonary CMR angiography revealed a partial anomalous pulmonary venous connection (PAPVC) of the right upper pulmonary vein with drainage into the superior vena cava (white arrows in Panel A, see Supplementary material online, Movie S2).

Cardiovascular magnetic resonance determined left-to-right shunting yielded a Qp:Qs = 1.89 (shunt fraction, 47%). Consequently, surgical repair was recommended: an autologous pericardial patch was used to redirect the blood flow from the orifice of the anomalous pulmonary vein into the left atrium via a surgically created ASD (Warden procedure with intracardiac baffle; outlined arrow in Panel C, see Supplementary material online, Movie S3). At 3-month follow-up, the symptom-free patient presented with improved exercise capacity and CMR imaging highlighted already normalized right-ventricular dimensions (Panel D, see Supplementary material online, Movie S1).

Partial anomalous pulmonary venous connection as a rare congenital cardiac defect represents an uncommon cause of right-ventricular enlargement. While in most cases, PAPVC is associated with an ASD (sinus venosus type), an isolated PAPVC with an intact atrial septum is a very rare finding and difficult to detect on echocardiography. In order to clarify the diagnosis of patients with unexplained right-ventricular enlargement, CMR imaging consistently constitutes the method of choice.

Supplementary material is available at European Heart Journal online.

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