

Multimodal imaging in a rare malformation of bilateral supracardiac partial anomalous pulmonary venous connection

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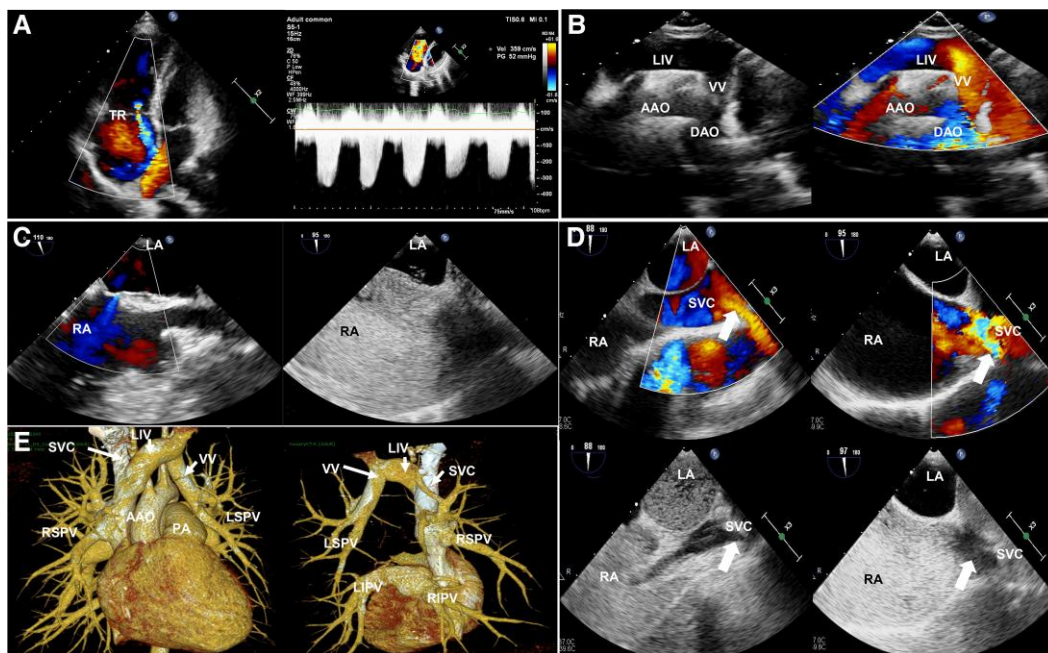


Figure 1 (A) Transthoracic echocardiography revealed enlarged right heart chambers with moderate tricuspid regurgitation, and the velocity of tricuspid regurgitation. (B) The suprasternal axis view showed an abnormal laminar continuous venous flow draining into the LIV. (C) TEE revealed a PFO. Colour Doppler revealed a left–right shunting, and right-heart agitated saline study shown an immediate passage of saline bubbles from RA to LA through the PFO. (D) TEE with colour Cooper, and negative saline contrast study revealed two separate bloodstreams that drained into the SVC. (E) A contrast-enhanced computed tomography image of PAVC. AA, aortic arch; AAO, ascending aorta; LA, left atria; LIPV, left inferior pulmonary vein; LIV, left innominate vein; LSPV, the left superior pulmonary vein; PA, pulmonary artery; PAPVC, partial anomalous pulmonary venous connection; PFO, patent foramen ovale; RA, right atria; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein; SVC, superior vena cava; TEE, transesophageal echocardiography; VV, vertical vein.

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A 40-year-old female presented to our hospital with complaints of chest tightness after exercise. Transthoracic echocardiography (TTE) revealed enlarged right heart chambers and moderate tricuspid regurgitation (Figure 1, panel A), indirect evidence of moderate pulmonary artery hypertension, with an estimated right ventricular systolic pressure of 60 mmHg and good biventricular function. The suprasternal axis view showed an abnormal laminar continuous venous flow draining into the left innominate vein (LIV) (panel B). Transesophageal echocardiography (TEE) revealed a patent foramen ovale (PFO) with left-to-right shunting (panel C). After the administration of agitated saline, there was an immediate passage of saline bubbles from the right atrium (RA) to the left atrium (LA) through the PFO after a Valsalva manoeuvre (panel C). The right inferior pulmonary vein (RIPV) and the left inferior pulmonary vein (LIPV) drain in to the LA. However, two separate bloodstreams drained into the superior vena cava (SVC) (panel D, [Supplementary material online, Video S1](#)). Negative contrast effects of agitated saline were observed in the draining area in SVC, relatively (panel D, [Supplementary material online, Video S2](#)). A contrast-enhanced computed tomography was performed and showed a partial anomalous pulmonary venous connection (PAPVC) with the right superior pulmonary vein (RSPV) to the SVC with two sites, and the left superior pulmonary vein (LSPV) to the LIV via an anomalous vertical vein (VV) (panel E). The PFO was confirmed. The patient underwent successful surgical correction which verified our diagnosis.

PAPVC is relatively uncommon, being observed in 0.4–0.7% of autopsies. Bilateral anomalous connection is a rare anatomical finding, occurring in 0.9–1.6% of the reported cases, of which no case report has described RSPV with two sites drained into the SVC. In patients with elevated right-sided pressures and possible shunt not clearly noted

on TTE, TEE with saline contrast echocardiography and cardiac computed tomography imaging can provide pertinent clues and help clinch the diagnosis.

Supplementary material

[Supplementary material](#) is available at *European Heart Journal – Case Reports*.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [supplementary data](#).

Consent: Consent for publication has been obtained from the patient in line with COPE guidelines.

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Data availability

The data underlying this article are available in the article and in its online [supplementary material](#).