Photo Clinic

Left Partial Anomalous Pulmonary Venous Connection and Vertical Vein

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A 41-year-old man with the complaint of exertional dyspnea and atypical chest pain was admitted to the emergency department. The patient's electrocardiogram showed right bundle branch block (RBBB) and T-wave inversion in *leads II* and *III*. Transesophageal echocardiography revealed normal left ventricular systolic function, mild right ventricular enlargement, and a left partial anomalous pulmonary venous connection (PAPVC) with the left pulmonary vein draining into the innominate vein via a large vertical vein (Figure 1). Subsequently, multislice spiral computed tomography (CT) angiography displayed a connection between the left superior pulmonary vein and the left brachiocephalic vein via the vertical vein and mild enlargement of the right atrium and ventricle (Figure 2). Coronary artery angiography also showed normal coronary arteries. Therefore, the patient was referred for surgical correction. The surgical correction was successfully performed on cardiopulmonary bypass (Figure 3).

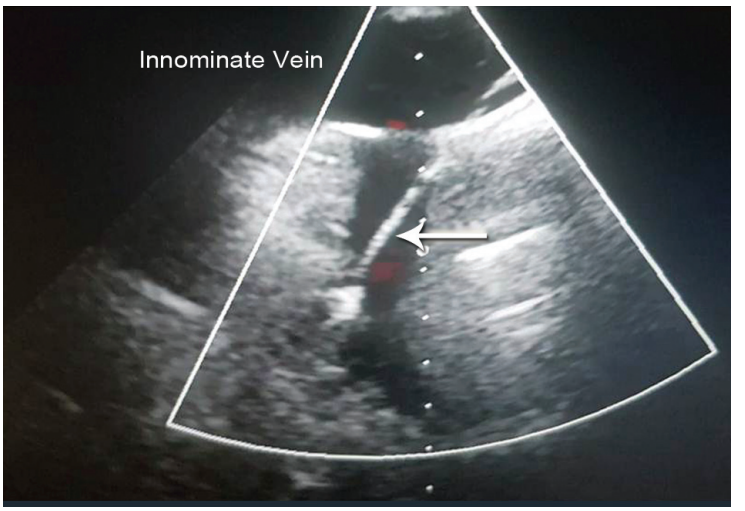

Figure 1. The image depicts transthoracic echocardiography in the suprasternal view. The arrow demonstrates the abnormal drainage of a vertical vein into the innominate vein.

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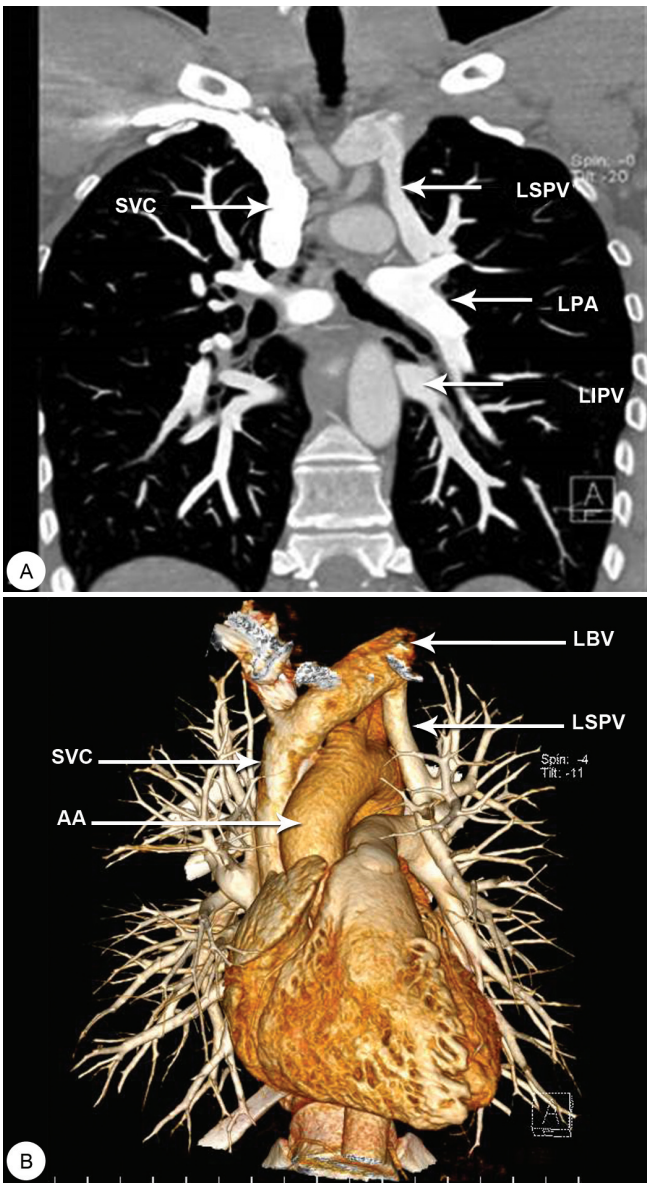

Figure 2. The images present the cardiac computed tomography angiography performed on the patient.
A) The image depicts the coronal maximum intensity projection. B) The volume-rendered images reveal an anomalous connection between the LSPV and the LBV as a vertical vein, indicating a partial anomalous pulmonary venous return.
LSPV, Left superior pulmonary vein; LBV, Left brachiocephalic vein, SVC, Superior vena cava; AA, Ascending aorta; LIPV, Left inferior pulmonary vein; LPA, Left pulmonary artery

PAPVC is a rare congenital heart disease in that it comprises 0.6% of all cases of congenital heart diseases. The incidence in adulthood without correction is also extremely rare (0.007%). PAPVC with an intact atrial septum can go undetected into adulthood, as was the case in our patient. PAPVC can be associated with other cardiac lesions, most commonly with atrial septal defects. The most common type is the connection of the right pulmonary veins to the right superior vena cava, while the least frequent type is the connection between the left pulmonary veins and the left innominate vein or the coronary sinus. The diagnosis in adults is *generally* aided by symptoms suggesting right heart failure and exercise intolerance and confirmed by imaging modalities (CT or magnetic resonance imaging) and echocardiography. A differential diagnosis of PAPVC is pulmonary varices (stenosis or atresia of 1 of the 4 pulmonary veins). Additionally, the differential diagnosis of an enlarged vertical vein is a left superior vena cava or an enlarged left superior intercostal vein. An isolated PAPVC usually remains asymptomatic until symptom development due to pulmonary hypertension or left-to-right shunting. The management of an isolated PAPVC is conservative medical treatment or surgical repair based on clinical symptoms, shunt fractions (the Qp/Qs ratio), concurrent indications for cardiac surgery, and recurrent pulmonary infections.[1-4] Finally, the rare occurrence of PAPVC does not diminish its significance; therefore, it should always be considered in cardiac physical examinations.

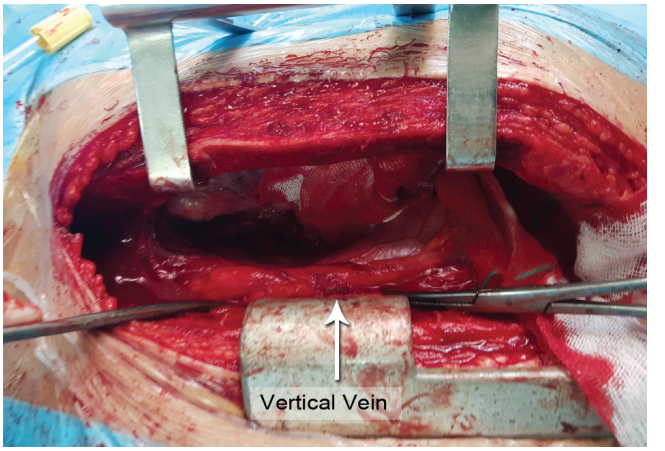

Figure 3. The intraoperative image shows the vertical vein (arrow) after sternal retraction during the surgical repair of the left partial anomalous pulmonary venous connection.

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