Successful repair of obstructed pulmonary venous confluence by the descending aorta in a 43-year-old patient: pre- and post-operative images

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A 43-year-old female patient presented with a 3-year history of progressive dyspnea (NYHA III). She underwent pulmonary artery banding at the age of 6 months followed by ventricular septal defect repair at 6 years and mitral valve repair at the age of 7 years. On physical examination, the patient was dyspneic on mild effort with a soft pansystolic murmur over lower left sternal edge.

Trans-thoracic echocardiography revealed severe tricuspid valve regurgitation and turbulent flow at the site of entrance of the pulmonary veins (PVs) in the left atrium (LA). (Panel A).

Contrast-enhanced CT scanning revealed abnormal PV drainage with obstruction at the level of both left-sided PVs and PV confluence connection to the LA (Panels B and C). A small persistent left superior vena cava (LSVC) drained into the LA (Panel D).

Surgical correction involved enlargement of the left-sided PV ostium and of the confluence to the LA, ligation of LSVC, and repair of the tricuspid valve. CT scanning 1 month after surgery revealed wide open pulmonary connection to the LA (Panels D and E). The patient was symptom free at the 6-month follow-up.

(Panel A) Apical four chambers view of transthoracic echocardiography showing stenosed common orifice of the PVs and turbulent blood flow at the site of entrance in LA. (B and C) Pre-operative CT with axial and reconstructed images showing that both the left superior and the inferior PVs join together forming a narrow compressed channel that joins another right common channel, both open by a narrow opening with the LA. (D) Pre-operative coronal image showing persistent LSVC. (E and F) Post-operative axial and reconstructed images showing wide open entry of all PVs into the LA.