A bad shortcut: partial anomalous pulmonary venous return

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DESCRIPTION

A woman in her 70s with a longstanding history of 'heart murmur' presented with worsening shortness of breath. Transthoracic echocardiogram demonstrated a severely dilated and dysfunctional right ventricle (RV), severe pulmonary hypertension and a dilated coronary sinus (video 1). A CT scan demonstrated a large sinus venosus-type atrial septal defect with partial anomalous pulmonary venous return (PAPVR) involving veins draining the right-upper and right-middle lobes into the superior vena cava (SVC; figure 1 and video 2).

Right heart catheterisation demonstrated flow-mediated pulmonary hypertension, with saturations rising from 52% to 99% when measured high versus low in the SVC, consistent with PAPVR. Her pulmonary to systemic blood flow ratio (Qp:Qs) was calculated as 2.7:1 (normal 1:1), with a normal right atrial pressure of 7 mm Hg, elevated mean pulmonary artery pressure of 40 mm Hg, and a normal pulmonary artery wedge pressure of 13 mm Hg. Cardiac output was markedly elevated at 12 L/min (normal ~5 L/min).

Despite the presence of severe pulmonary hypertension and the echocardiographic finding of severe RV dysfunction, her pulmonary vascular resistance (PVR) was less than 3 Wood units and her right atrial pressure was normal, suggesting a compensated physiological state. The patient was felt to be a good surgical candidate due to the relatively low PVR and high Qp:Qs. She declined surgical intervention citing her age as the main factor and expired a few months later from her cardiac condition.

Video 1 Transthoracic Echocardiogram.

Video 2 CT Review of Anatomy.

Figure 1 Three-dimensional CT reconstruction of vascular anatomy (SVC, superior vena cava; PV, pulmonary venous).

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Learning points

- ▶ Although the shunt fraction is a risk factor for the development of pulmonary vascular disease, this case demonstrates that even patients with longstanding high-flow shunts may not develop pulmonary arterial hypertension.
- ▶ The prevalence of partial anomalous pulmonary venous return is most often reported between 0.1% and 0.7%, though this may represent an underestimate, as many patients may be completely asymptomatic.^{1 2}
- ▶ The magnitude of the left-to-right shunt, the presence of symptoms and the pulmonary vascular resistance all factor into decisions regarding treatment.³

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