

## Images in Congenital Cardiac Disease

### So-called “absence” of the leaflets of the pulmonary valve

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**A** NEONATE WHO HAD SYSTOLIC AND DIASTOLIC murmurs was referred to our institution for investigation of moderate cyanosis without respiratory distress. Cross-sectional echocardiography

showed a large ventricular septal defect opening beneath the ventricular outlets. The short axis parasternal view (Fig. 1), revealed that the subpulmonary infundibulum, arising from the right ventricle (RV), coursed through a hypoplastic ventriculo-pulmonary junction (PV) to capacious left (LPA) and right (RPA) pulmonary arteries. Colour Doppler interrogation revealed severe pulmonary regurgitation. Three-dimensional echocardiography was performed using the paediatric matrix probe (2–7 MHz). The same view (Fig. 2) characterized the specific anatomy of the rudimentary formation of the leaflets of the pulmonary valve, an arrangement usually described as “absent pulmonary valve”. Viewed from below, the remnants of the valve were seen to be composed of thick nodular tissue, without demarcation of true

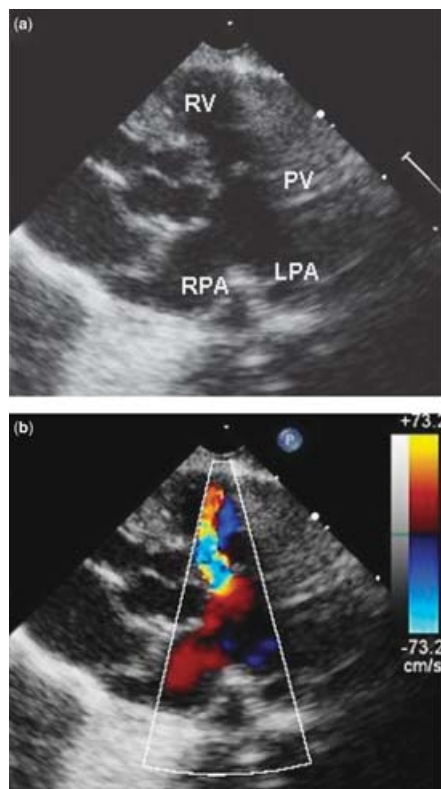


Figure 1.

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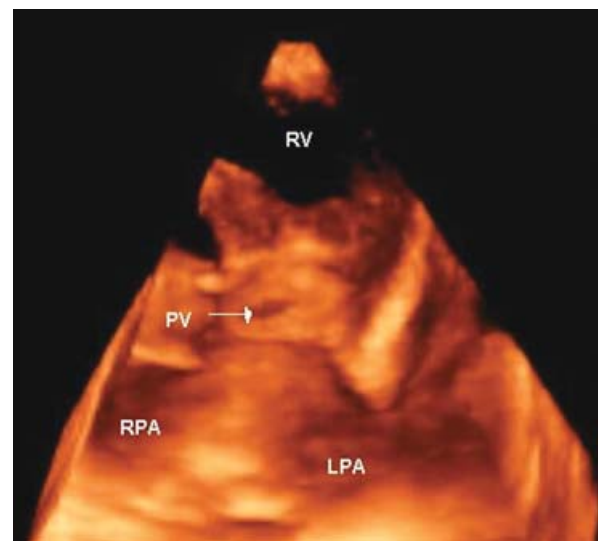


Figure 2.

leaflets or zones of apposition. The arrow indicates the eccentric and regurgitant orifice. The left (LPA) and right (RPA) pulmonary arteries were massively dilated. This patient, who is waiting for surgical correction, illustrates well the features of absent pulmonary valve syndrome,<sup>1</sup> in reality rudimentary formation of the valvar tissue, which can be

demonstrated well by transthoracic three-dimensional echocardiography.

### Reference

1. Zucker N, Rozin I, Levitas A, Zalstein E. Clinical presentation, natural history, and outcome of patients with the absent pulmonary valve syndrome. *Cardiol Young* 2004; 14: 402–408.