

Asymptomatic Pulmonary Artery Aneurysm with a Bicuspid Pulmonic Valve

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An active 78-year-old woman with a lifelong asymptomatic heart murmur underwent primary care evaluation. Her physician characterized a grade 3/6, crescendo-decrescendo murmur heard best at the left sternal border, with an increased split in S₂ and increased intensity with inspiration. A transthoracic echocardiogram showed elevated velocities across the pulmonic valve (PV), suggesting stenotic disease. Right-sided heart catheterization revealed a right ventricular systolic pressure of 69 mmHg, a main pulmonary artery (PA) systolic pressure of 30 mmHg, and a mean gradient across the PV of 26 mmHg.

The patient was referred to our center for cardiac magnetic resonance. A crescent-shaped orifice was consistent with a bicuspid PV (Fig. 1). The main PA was substantially dilated (maximum diameter, 5.15 cm) (Fig. 2). The right ventricle was mildly

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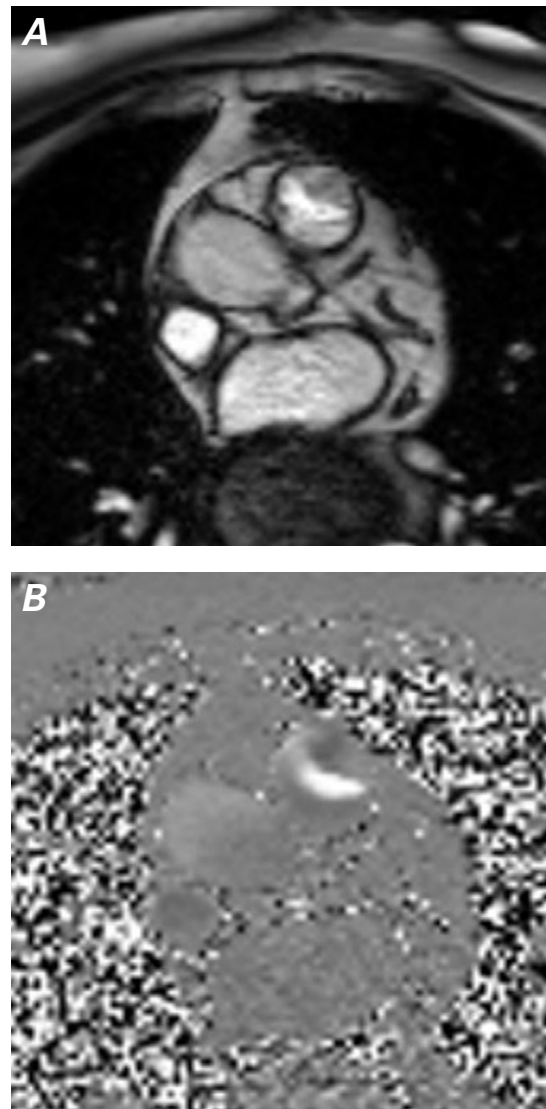


Fig. 1 Cardiac magnetic resonance images. **A)** Steady-state free precession sequence shows the opening of the bicuspid pulmonic valve. **B)** Phase contrast sequence shows a crescent-shaped flow pattern across the valve.

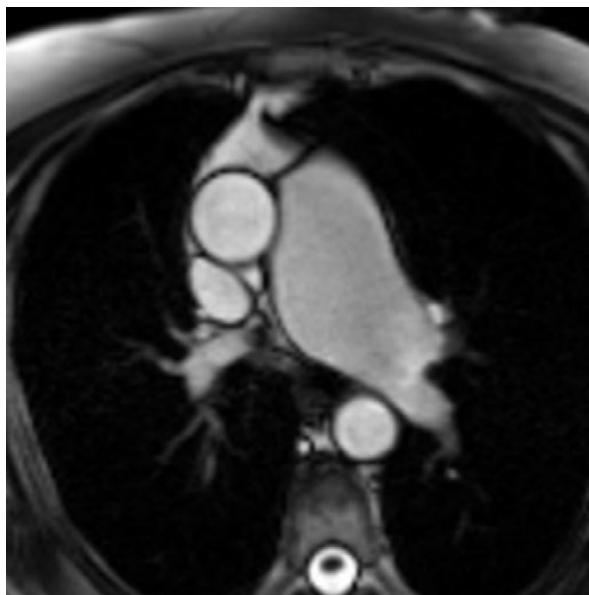


Fig. 2 Cardiac magnetic resonance image (steady-state free precession sequence in axial view) shows the aneurysmal, dilated main pulmonary artery.

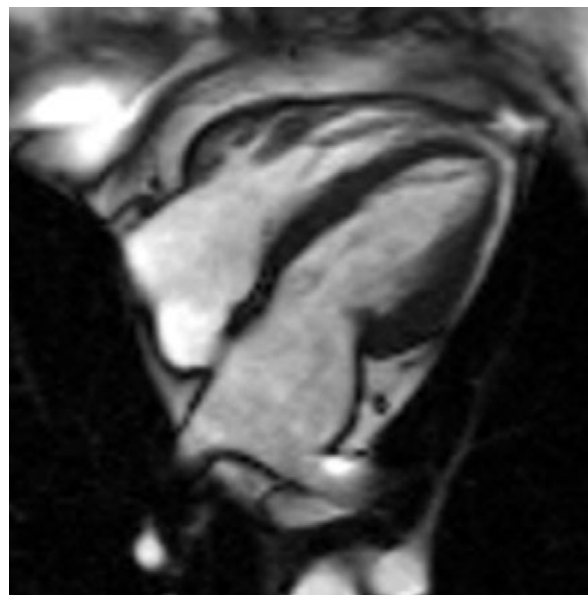


Fig. 3 Cardiac magnetic resonance image (steady-state free precession sequence in 4-chamber view) shows mild right ventricular enlargement without obvious free-wall hypertrophy.

dilated without obvious free-wall hypertrophy (Fig. 3); the ejection fraction was 0.59. The left ventricular ejection fraction was 0.77. The aorta's size was normal, and the aortic valve was trileaflet. The patient had no symptoms and had a PA pressure <50 mmHg, so clinical monitoring was maintained.

Comment

Pulmonary artery aneurysms are rare, having been described in approximately 1 of 14,000 autopsies.¹ Usually, PA aneurysms occur in clinical situations such as congenital heart disease and pulmonary hypertension.² Bicuspid PVs have been associated with dilation of the pulmonary trunk. In patients who have a PA aneurysm and a bicuspid PV, the most likely origin is abnormal migration of the neural crest cells.³

Clinically, bicuspid PVs with PA aneurysms do not pose the same clinical risk as do their systemic counterparts, because of the lower pressure in the PAs. The greatest risk for a PA dissection from a PA aneurysm is an absolute pressure limit >50 mmHg,⁴ so we recommended clinical monitoring for our patient.

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