

Quadricuspid Aortic Valve with Patent Ductus Arteriosus

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A 30-year-old woman was referred to our hospital for evaluation of increasing dyspnea on exertion. On physical examination, a continuous Gibson murmur was heard at the upper left sternal border. Results of electrocardiography and chest radiography were normal. Transthoracic echocardiography revealed a quadricuspid aortic valve with mild aortic regurgitation; color-flow Doppler imaging showed a narrow mosaic jet; and continuous-wave Doppler imaging showed high-velocity left-to-right flow. There was no atrial or ventricular dilation, and systolic function was normal. Aortography showed shunting from the aorta into the pulmonary artery (Fig. 1). Transesophageal echocardiography showed the X-shaped commissural pattern of a quadricuspid aortic valve (Fig. 2A) and a patent foramen ovale (Fig. 2B). A patent ductus arteriosus (PDA) was closed percutaneously with use of an AMPLATZER® duct occluder (AGA Medical Corporation; Plymouth, Minn).

Comment

Quadricuspid aortic valve is a very rare congenital malformation, far less common than unicuspid or bicuspid aortic valve.¹ Most cases have been discovered incidentally during surgery or at autopsy. The incidence at autopsy is reportedly between 0.008% and 0.043%.^{2,3} In accordance with the classification system of Hurwitz and Roberts, our patient had a “type C” quadricuspid aortic valve (2 equal larger cusps and 2 equal smaller cusps).¹

Quadricuspid aortic valve in association with other congenital anomalies is extremely rare. Aortic regurgitation appears to be the most common hemodynamic abnormality associated with quadricuspid aortic valve.⁴ Aortic regurgitation develops chiefly as a result of fibrosis and incomplete coaptation, and surgery is frequently required in a patient’s 5th or 6th decade of life.¹ Coronary artery anomalies have been seen in 10%

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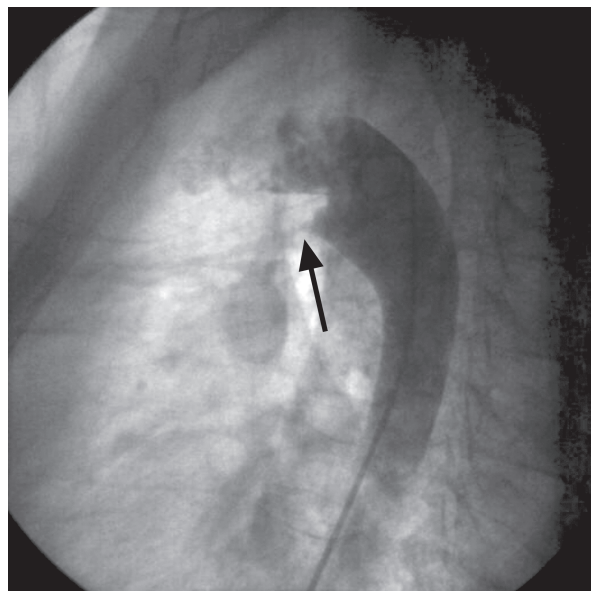


Fig. 1 Aortogram shows shunting from the aorta into the pulmonary artery (arrow).

Real-time motion image is available at www.texasheart.org/journal.

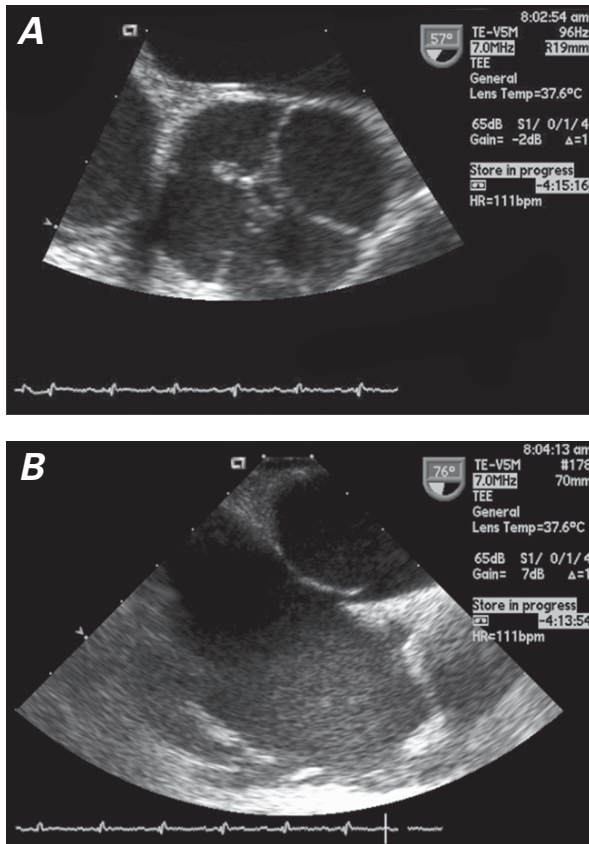


Fig. 2 Transesophageal echocardiography shows **A**) a quadricuspid aortic valve and **B**) a patent foramen ovale.

Real-time motion image of Figure 2A is available at www.texasheart.org/journal.

of cases of quadricuspid aortic valve.³ Quadricuspid aortic valves have been reported in association with non-obstructive cardiomyopathy, pulmonary valve stenosis, ventricular septal defect, and fibromuscular subaortic stenosis.^{1,4} Infective endocarditis is a potential complication.⁵

Echocardiography is increasingly useful in detecting quadricuspid aortic valve. If this anomaly is found incidentally, continual follow-up is recommended, in view of the eventual requirement of valve replacement. Patients with quadricuspid aortic valve should also be carefully evaluated for other congenital abnormalities.

In 1923, Simonds described a postmortem finding of quadricuspid aortic valve with PDA.⁶ To the best of our knowledge, ours is the 1st report of quadricuspid aortic valve in association with PDA in a living patient.

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