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Two cases of quadricuspid aortic valve

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Bicuspid pulmonary valve

Abstract Quadricuspid aortic valve is a rare congenital malformation of the aortic valve. Most cases are discovered incidentally at autopsy or at aortic valve replacement. Recent advances in echocardiography epecially transesophageal echocardiography led to diagnosis of more cases before surgery. Two cases of quadricuspid aortic valve are presented.
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Case 1

A 31-year-old man presented with a history of dyspnea on exertion that began seven years ago. Physical examination showed central cyanosis, clubbing and a 4/6-grade systolic ejection murmur at all areas accompanied by a thrill. Transthoracic echocardiography (TTE) showed biventricular enlargement, right ventricular hypertrophy, and a large perimembranous ventricular septal defect (VSD). A maximum gradient of 107 mmHg was present across the pulmonary valve. Transesophageal echocardiography revealed a large perimembranous VSD, secundum type atrial septal defect (ASD) and a quadricuspid aortic valve (QAV). Aortic valve cusps were equal in size, opening was normal. There was mild aortic regurgitation. The thicknesses of the pulmonary valves were increased, with a limitation in the movement. Cardiac catheterization confirmed the diagnosis. The patient was referred to surgery. VSD and ASD were repaired. As the pulmonary valve turned up to be bicuspid and stenotic, the operations valvotomy, myectomy and myotomy were performed. Surgery was not performed for the aortic valve.

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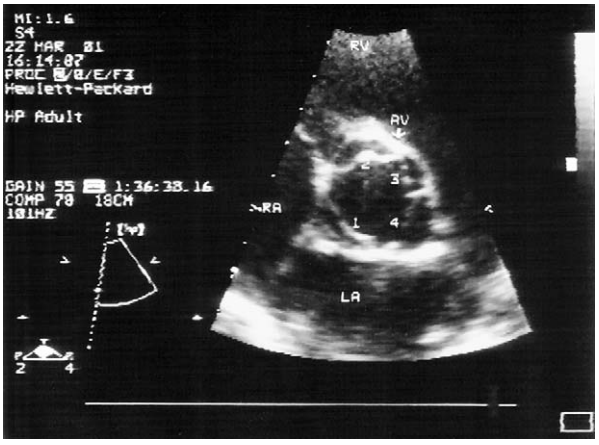


Figure 1 Systolic transthoracic echocardiographic view of quadricuspid aortic valve with four approximately equal-sized freely opening cusps in Case 2.

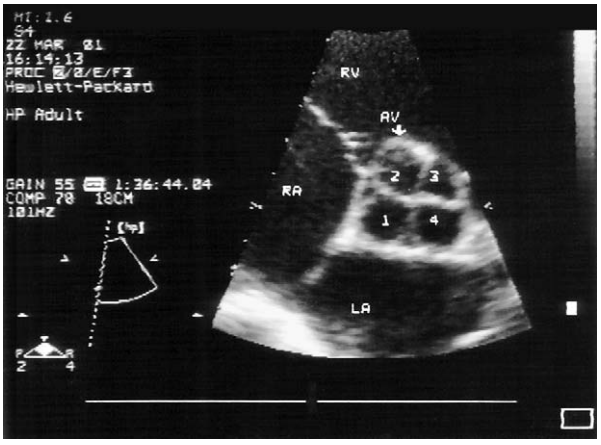


Figure 2 Diastolic transthoracic echocardiographic view of quadricuspid aortic valve with four equal cusps in Case 2. On the short axis view of the aortic valve, the commissural lines formed by the adjacent cusps resulted in an ''X'' configuration rather than the ''Y'' configuration of the normal tricuspid aortic valve.

Case 2

A 24-year old man was admitted with dyspnea. His physical examination was normal. TTE showed a QAV with four equal cusps on both systolic and diastolic frames (Figs. 1 and 2).

Autopsy series have reported an incidence of QAVs between 0.008% and 0.033%.[1] QAVs may be isolated or associated with other congenital cardiac abnormalities. A case of quadricuspid pulmonary valve and bicuspid aortic valve has been reported.[2] The cause of the combined abnormalities might be both an abnormality of mesenchymal proliferation in common trunk and aberrant fusion of the aortopulmonary septum. To our knowledge this is the first case of complex cardiac abnormality where QAVs are associated with ASD, VSD, pulmonary valve stenosis, and bicuspid pulmonary valve.

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