

# Quadricuspid aortic valve: a rare cause of aortic regurgitation and stenosis

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Quadricuspid aortic valve is a rare congenital condition that occurs not only as an isolated anomaly, but also with other cardiac defects. We describe a 10-year-old boy whose aortic stenosis was diagnosed during infancy. Transthoracic echocardiography revealed dilation of the left ventricle, valvular and subvalvular aortic stenosis, bicuspid aortic valve, aortic regurgitation, and mitral valve prolapse. The results of cardiac catheterization and aortography showed severe aortic regurgitation, an aortic valve gradient of 76 mmHg, a bicuspid aortic valve, a subaortic membrane, and an ascending aortic aneurysm. The patient underwent elective valve replacement with a mechanical prosthesis, and during surgery, the valve was noted to be quadricuspid. The patient was diagnosed as having a quadricuspid aortic valve associated with aortic regurgitation, severe aortic stenosis, and an ascending aortic aneurysm.

*Key words:* quadricuspid aortic valve, aortic regurgitation, aortic stenosis.

Quadricuspid aortic valve (QAV) is a very rare congenital condition that occurs not only as an isolated anomaly but also with other cardiac defects<sup>1,2-4</sup>. Most QAVs were discovered as an incidental finding during aortic valve surgery or at autopsy. Because of the advances in echocardiography, more cases are now being diagnosed preoperatively<sup>5</sup>.

We report a patient with a QAV associated with aortic regurgitation, aortic stenosis, and an ascending aortic aneurysm.

## Case Report

A 10-year-old boy with aortic stenosis, who had been diagnosed at another medical center during infancy, was admitted to our hospital for routine check-up. On physical examination, his blood pressure was 100/60 mmHg. A grade-2 systolic ejection murmur was audible in the second right sternal border. A chest radiograph showed cardiomegaly, with cardiothoracic ratio of 0.60, and an electrocardiogram revealed signs of left ventricular hypertrophy. Transthoracic echocardiography showed moderate dilation of the left ventricle, bicuspid aortic valve with valvular aortic stenosis, grade-2 to grade-3 aortic

regurgitation, and subaortic membrane that caused a peak systolic gradient of 40 mmHg. Cardiac catheterization and aortography showed severe aortic regurgitation (grade 3), valvular aortic stenosis with a peak systolic gradient of 76 mmHg, a subaortic membrane, a bicuspid aortic valve, and an ascending aortic aneurysm (Fig. 1). The patient underwent elective valve replacement with mechanical prosthesis and Bentall procedure. During surgery, the aortic valve was noted as quadricuspid (Fig. 2). The patient was discharged on the fifth postoperative day with no complications.

## Discussion

Quadricuspid aortic valve is a rare cardiac anomaly, which has been identified in 0.008% to 0.03% of the population<sup>1</sup>. A review of a modern echocardiography database shows the prevalence of that condition to be somewhat higher (from 0.013% and 0.043%) depending on the years reviewed<sup>1</sup>.

Hurwitz and Roberts<sup>2</sup> suggest that there are 7 types of QAV (type A to type G) that are defined according to the relative size of the 4 cusps. The most common type of QAV

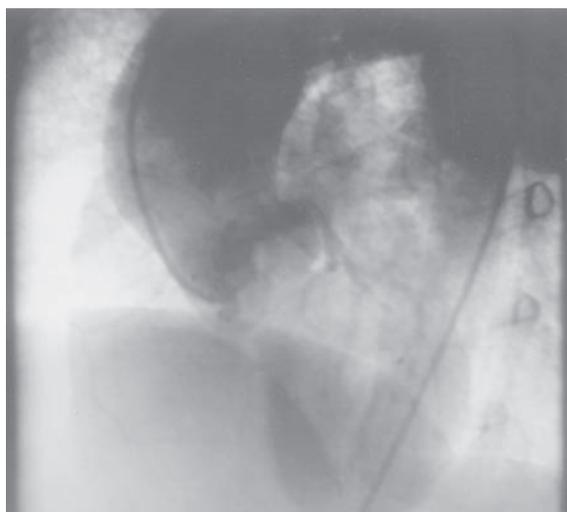


Fig. 1. Postoperative view of the quadricuspid aortic valve cusps.



Fig. 2. Quadricuspid aortic valve.

consists of 3 equally sized cusps and 1 smaller (or hypoplastic) cusp (type B). This was the type of QAV exhibited in our patient.

In many cases, a QAV is an isolated anomaly, but it may also be associated with other congenital malformations, such as ventricular septal defects, patent ductus arteriosus, subaortic fibromuscular stenosis, or hypertrophic non-obstructive cardiomyopathy<sup>2-4</sup>.

Quadricuspid aortic valve is seen most frequently with significant aortic regurgitation and mild-to-moderate stenosis<sup>1,3,6-8</sup>. There have been documented cases of endocarditis that affected isolated QAV patients, but the risk of endocarditis in patients with QAV has not been established<sup>1,2, 9-12</sup>.

Valvular incompetence in QAV is rare in infants or young children<sup>2,13</sup>. In most of those cases, aortic regurgitation tends to deteriorate in adulthood. Valve replacement is frequently required during the fifth or sixth decade of life<sup>5</sup>. Aortic root dilation in patients with QAV has been reported in only 3 cases<sup>14,15</sup>. Interestingly, despite his young age, moderate dilation of the aortic root was apparent in our patient.

In the past, QAVs were diagnosed at autopsy or during aortic valve surgery. The advent of echocardiography has enhanced our ability to diagnose that condition; however, it may not be possible to visualize the aortic leaflets adequately with transthoracic echocardiography. With the advent of transesophageal echocardiogram, preoperative diagnosis has become commonplace<sup>5</sup>. Egred and colleagues<sup>16</sup> reported the important role of cardiac magnetic resonance imaging in establishing and confirming the diagnosis of a QAV. The preoperative diagnosis of a QAV is important because that anomaly can be associated with an abnormally placed coronary ostium. Therefore, preoperative coronary angiography is necessary when a QAV is diagnosed<sup>17</sup>. For patients with QAV because of severe valvular regurgitation and stenosis, aortic valve replacement is usually the preferred treatment<sup>4,13</sup>.

In conclusion, QAV is a rare congenital malformation that can be diagnosed by echocardiography (primarily transesophageal), and it may be associated with aortic regurgitation and stenosis.

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