

## Female Adolescent With Quadricuspid Aortic Valve

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**Quadricuspid aortic valve is a rare congenital heart defect. The authors present a case of this anomaly in a young, asymptomatic adolescent who presented to the echocardiography laboratory for further evaluation of a cardiac murmur detected on routine physical examination. Imaging revealed a quadricuspid aortic valve with aortic regurgitation. This case highlights the importance of auscultation in detecting cardiac abnormalities.**

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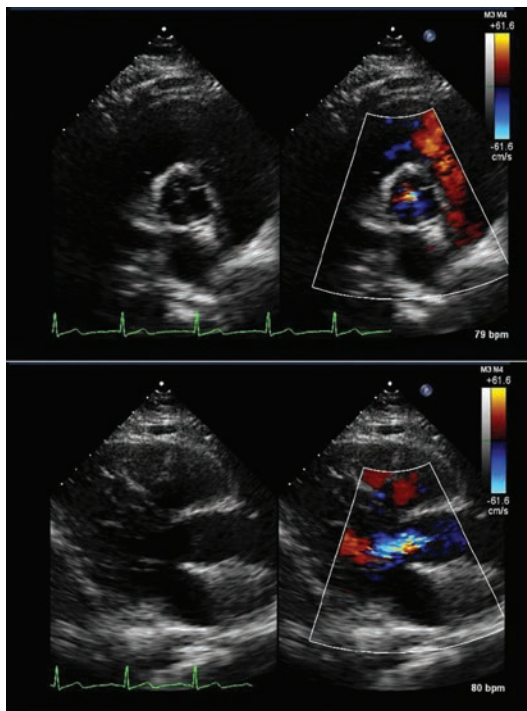
A quadricuspid aortic valve (QAV) is a congenital heart defect that has been increasingly reported as imaging techniques have advanced.<sup>1</sup> Nonetheless, it remains a rare anomaly, with nearly 200 cases reported in the literature since the first case was discovered in 1862.<sup>2</sup> We report a case of QAV in a young patient who presented for echocardiographic evaluation of a cardiac murmur detected on routine physical examination.

### Report of Case

A 17-year-old female adolescent was referred to our echocardiography laboratory for evaluation of a cardiac murmur. She had no reported history of congenital heart defect. On examination, her blood pressure was 111/70 mm Hg and her pulse rate was 85/min. Two-dimensional echocardiography revealed a QAV with moderate aortic regurgitation, necessitating further evaluation with transesophageal echocardiography (*Figure 1* and *Figure 2*). Her aortic regurgitation pressure half-time was 397 cm/s<sup>2</sup>. Her left ventricular end diastolic dimension was 4.4 cm and end systolic dimension was 3.1 cm, ejection fraction was 56%, and left ventricular wall thickness was in the upper limit of normal. The rest of the cardiac structures were all normal in size and function. Ongoing follow-up was recommended to monitor for evidence of left ventricular systolic and structural dysfunction and the need for surgical replacement of the QAV using 2-dimensional echocardiography.

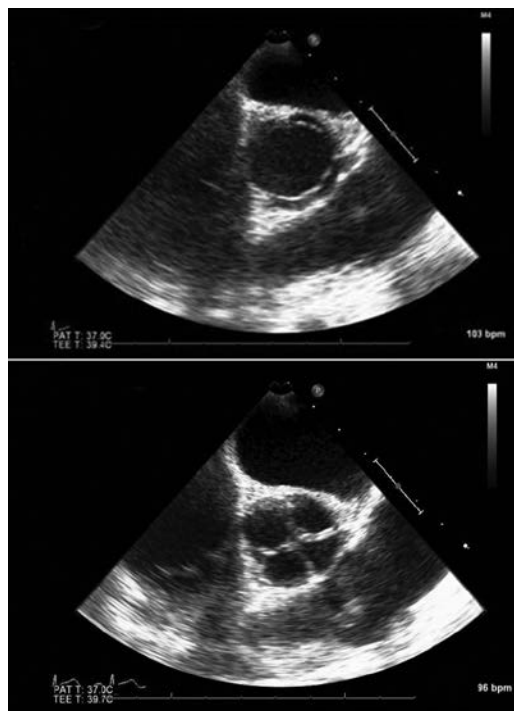
### Discussion

Important physical examination findings of aortic regurgitation include an early decrescendo diastolic murmur detected on auscultation. This murmur is generally high pitched and heard best at the third left intercostal space along the sternal border, also known as Erb point. Other important clinical clues include wide pulse pressure, Corrigan pulse, brisk carotid upstroke on palpation of the carotid artery, and Quincke nail bed pulsation. In severe longstanding aortic regurgitation with left ventricular failure, S3 or S4 may be auscultated.



**Figure 1.**

Parasternal short axis showing a predominate central aortic regurgitation on transthoracic echocardiogram (top). Parasternal long axis showing aortic regurgitation on transthoracic echocardiogram (bottom).



**Figure 2.**

Transesophageal echocardiogram at mid-esophageal aortic valve short axis view in diastole (top). Transesophageal echocardiogram at mid-esophageal aortic valve short axis view in systole (bottom).

A normal aortic valve consists of 3 identical cusps. Bicuspid aortic valve is the most common anomaly followed by unicuspid aortic valve. Quadricuspid aortic valve is exceedingly rare. Quadricuspid pulmonic valves have an estimated 9 times higher incidence compared with QAV.<sup>3,4</sup> Advances in imaging techniques, such as 2-dimensional echocardiography, cardiac computed tomography, and cardiac magnetic resonance imaging, have led to greater numbers of diagnosed cases. In the past, QAV was discovered during open heart surgery or at autopsy. Tutarel<sup>5</sup> identified 186 cases in his comprehensive review of QAV published in 2004. The incidence was reported to be between 0.008% and 0.043%, with a slight male predominance.<sup>3</sup> Morbidities associated with this condition are generally progressive worsening of aortic

regurgitation with development of left ventricular dysfunction and heart failure.

Seven variants of QAV were described by Hurwitz and Roberts in 1973.<sup>6</sup> Two of these variants account for about 75% of cases identified: type A has 4 equal-sized cusps and type B, 3 equal cusps and 1 smaller cusp. The remaining 25% of cases have type C, with 2 equal larger and 2 equal smaller cusps; type D, with 1 large, 2 intermediate, and 1 small cusp; type E, with 3 equal cusps and 1 larger cusp; type F, with 2 equal larger and 2 equal smaller cusps; and type G, with 4 unequal cusps.

The functional status of cases has been reported to be pure aortic regurgitation in nearly 75%, combined aortic regurgitation and stenosis in 8%, and normal functioning valves in approximately 16%.<sup>4,7</sup> More than half of patients needed valve surgery because of progressive

left ventricular dysfunction secondary to aortic regurgitation.<sup>8</sup> In current practice, the surgical approach involves open heart surgery with complete replacement of the QAV with either a bioprosthetic or a mechanical valve. The transcatheter aortic valve replacement approach is currently not recommended or studied in clinical trials for patients with severe regurgitation.

Regarding endocarditis prophylaxis for patients with QAV or any congenital valvular abnormalities, the American College of Cardiology/American Heart Association (ACC/AHA) 2008 update<sup>9</sup> on guidelines for infective endocarditis does not recommend prophylactic antibiotic treatment before dental, respiratory, genitourinary, or gastrointestinal invasive procedures in the absence of active infection.

Surveillance of and treatment for patients with aortic regurgitation depends on the ACC/AHA aortic regurgitation stages, ranging from mild (stage A) to most severe (stage D).<sup>10</sup> Each stage is defined by valve anatomy and valve hemodynamics, severity of left ventricular dilatation, left ventricular systolic function, and patient symptoms. Recommended follow-up intervals range from 3 months to annually.

## Conclusion

An important primary valvular disorder, QAV has the potential to cause substantial morbidity and mortality. Early detection and subsequent surveillance will allow optimal medical and surgical management that will give patients the best possible outcome. The current patient's diagnosis was type B variant QAV, with ACC/AHA stage B (progressive) aortic regurgitation. She will have annual follow-up evaluations for evidence of left ventricular dysfunction or development of heart failure symptoms necessitating surgical intervention.

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