

## Images in Geriatric Cardiology

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# Quadricuspid Aortic Valve With Four Equal Cusps in a Quinquagenarian

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We report a case of a patient with quadricuspid aortic valve (QAV), a rare isolated congenital abnormality, found incidentally on a transthoracic echocardiogram (TTE) ordered for a clinically unrelated indication. Our patient had a less common QAV variant, with four equal-sized cusps. Although it is believed that this QAV variant is more likely to have preserved valvular function, our patient nonetheless had moderate aortic regurgitation.

A 55-year-old woman was admitted to the intensive care unit following a suicide attempt. She had jumped out of a third-story window, suffered multiple bone fractures, and later developed deep venous thrombosis in her right leg. A TTE was ordered to evaluate the right ventricular function when bilateral pulmonary emboli were visualized on computed tomography.

TTE revealed that the global right ventricular systolic function was preserved despite significant pulmonary hypertension (50 mm Hg as assessed by the peak velocity of the tricuspid regurgitant jet). Left atrium and left ventricular size and function were normal.

Color Doppler imaging demonstrated moderate aortic regurgitation. No apparent aortic valve (AV) leaflet abnormalities were seen on the parasternal long axis; however, the short-axis view showed four AV cusps of equal size in both systole and diastole. At end diastole, the commissural lines formed by the adjacent AV cusps resulted in an “X” configuration rather than the “Y” configuration of the normal tricuspid AV (Figure 1 and Figure 2). In addition, there was incomplete coaptation of the AV leaflets, resulting in a regurgitant orifice located in the center of the AV.

Based on these findings, the diagnosis of congenital QAV was established. Antibiotic prophylaxis before bacteremia-induced procedures and annual echocardiograms were recommended. One year later, the patient is asymptomatic and no longer has pulmonary hypertension.

### DISCUSSION

Our case demonstrates the typical way in which the QAV is diagnosed, namely as a rare isolated asymptomatic congenital abnormality found incidentally on a TTE ordered for a clinically unrelated indication.

In 1862, Balington<sup>1</sup> described the first case of QAV on autopsy. The reported incidence is between 0.008% (two cases in 25,556 necropsies)<sup>2</sup> and 0.013% (eight cases in 60,446 echocardiograms).<sup>3</sup> Its true incidence may be underestimated, however, because QAV may be overlooked unless it is specifically sought. QAV may be more prevalent in men than women (1.6:1.0).<sup>4</sup>

The overall incidence of QAV is much less than that of other congenital abnormalities affecting the number of AV cusps. Bicuspid AV, the most common congenital cardiac abnormality in humans, occurs in approximately 0.9%–2% of all live births,<sup>5</sup> while the unicuspid AV has a reported incidence of 0.02%.<sup>6</sup> Interestingly, quadricuspid pulmonary valves are nine times more common than AVs.<sup>7</sup>

The four cusps of QAV are referred to as posterior, left coronary, right coronary, and anterior supernumerary cusp. They vary in size, thickness, and pliability. Hurwitz and Roberts<sup>8</sup> have described seven anatomic variants, with the most common type exhibiting three equal-sized cusps and one smaller cusp.

Aortic insufficiency is the predominant complication of QAV, occurring in up to 75% of the cases<sup>4</sup>



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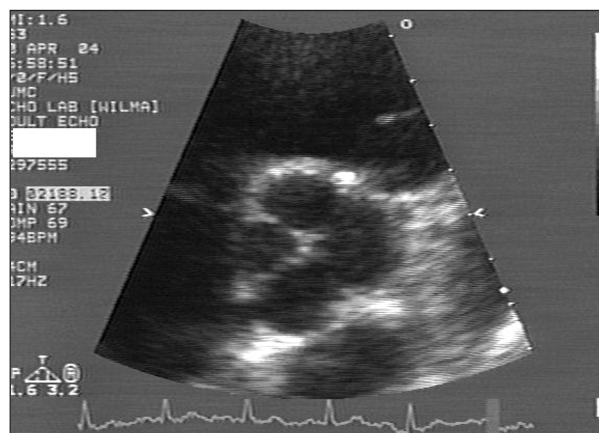


Figure 1. Transthoracic short-axis view showing four aortic valve cusps of equal size at end diastole with the characteristic "X" configuration formed by the commissural lines

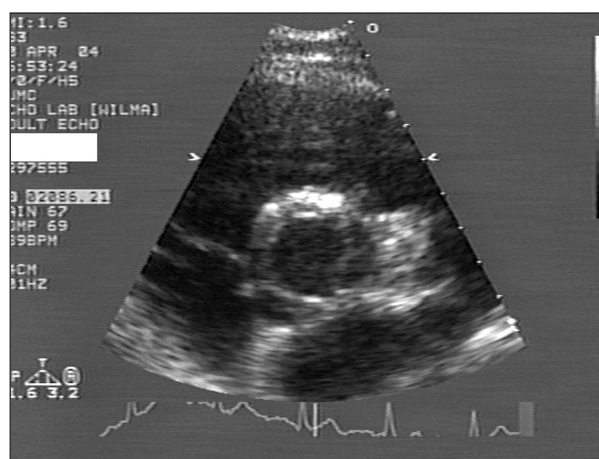


Figure 2. Transthoracic short-axis view showing four aortic valve cusps of approximately equal size at end systole

and is due to fibrous thickening and partial non-coaptation of the leaflets. AV replacement is often needed by the fifth or sixth decade of life.<sup>9</sup>

The risk of endocarditis in QAV has not been clearly established. At least three confirmed cases of endocarditis affecting a QAV have been reported, and endocarditis prophylaxis is advisable for all patients with documented QAV.<sup>3</sup>

As of July 2004, 186 cases of QAV have been reported,<sup>4</sup> and here we report one more. Our patient is unusual, however, in that she has a less common QAV variant with four equal-sized cusps. Although this variant has been reported to have a higher likelihood of functioning normally,<sup>8</sup> our patient nonetheless had moderate aortic regurgitation.

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