

Quadricuspid Aortic Valve: A Rare Cause of Aortic Regurgitation



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INTRODUCTION

A quadricuspid aortic valve (QAV) is a rare congenital anomaly that occurs with an incidence of <1%.¹

Over time, leaflet fibrosis and thickening may lead to leaflet malcoaptation and aortic regurgitation.²⁻⁵ Aortic stenosis is a less common finding.^{2,4} Usually the diagnosis of QAV is made by echocardiography, although other imaging modalities may be needed if imaging quality is unsatisfactory.^{2,6} Patients with long-standing aortic regurgitation may have hemodynamic consequences such as left ventricular chamber dilation or a decrease in left ventricular systolic function.^{6,7} Surgery is indicated in patients who are symptomatic, have left ventricular systolic dysfunction, or are undergoing cardiac surgery for another reason.⁶

CASE PRESENTATION

A 48-year-old Egyptian man with a history of a murmur presented with symptoms of progressive dyspnea on exertion, chest pressure, and palpitations for the preceding 3 weeks. Upon initial evaluation, the patient was afebrile, and blood pressure was 122/58 mm Hg with a normal heart rate. Cardiac examination revealed regular rate and rhythm and 3/4 diastolic murmur best heard at the left lower sternal border.

Because of his progressive dyspnea the patient was referred for transthoracic echocardiography, which revealed normal left ventricular systolic function, a dilated left ventricular chamber (Figures 1 and 2), and severe aortic regurgitation by color Doppler with a vena contracta width of 0.7 cm² (Figure 3).

The morphology of his aortic valve was not well visualized, and he was referred for transesophageal echocardiography (TEE). TEE showed a QAV with central leaflet malcoaptation and mild dilatation of the aortic root measuring 4 cm (Figures 4-6, Videos 1-3).

Cardiac catheterization before planned valve replacement surgery showed normal coronary arteries.

Intraoperative TEE again confirmed the QAV with severe aortic regurgitation. The patient underwent minimally invasive aortic valve replacement surgery and received a 25-mm Edwards pericardial valve. His postoperative course was complicated by a short episode of atrial fibrillation that was self-limited, with no recurrence on

VIDEO HIGHLIGHTS

Video 1: Two-dimensional transesophageal echocardiographic long-axis view of the left ventricle with color Doppler showing central aortic regurgitation.

Video 2: Two-dimensional TEE. Zoomed-in image clearly showing four aortic valve leaflets with incomplete central closure. There is a small independently mobile lesion seen attached to the anatomic noncoronary leaflet, likely representing tissue degeneration.

Video 3: Two-dimensional TEE with color Doppler showing leaflet malcoaptation and central aortic regurgitation in the aortic valve short-axis view.

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follow-up. Pathology of the valve showed four leaflets with fibrosis, focal thickening, and myxoid degeneration.

DISCUSSION

QAV is a rare finding with an incidence of 0.05% to 1% for those receiving aortic valve replacements for aortic regurgitation and 0.01% to 0.04% in echocardiographic and autopsy data.^{1,4}

Additional cardiac abnormalities were present in up to 32% of the patients with confirmed QAV.^{1,4} Some of the other frequent abnormalities include ventricular septal defect, pulmonary valve stenosis, subaortic fibromuscular stenosis, and coronary ostial malformation and displacement.^{1,2} The most frequently discovered abnormality in these patients is coronary artery and coronary ostial abnormalities.¹

The mechanisms of development are not well known, but it has been suggested that there is anomalous septation of the conotruncus and abnormal septation of one of the endocardial cushions due to an inflammatory event, though some studies have shown this to be less likely.^{2,3,8} It has also been suggested that the development of QAV is from abnormal division of one of the three mesenchymal ridges that later become the aortic valve cushions.^{2,3,8}

There are several classifications of QAV based on the sizes of the cusps and the location of the accessory cusp. Hurwitz and Roberts⁹ classified QAV into subtypes A to G on the basis of the relative sizes of the valve leaflets. The most commonly identified morphologies are type A (four equally sized cusps) and type B (three equally sized cusps and a smaller accessory cusp).³ Type B has been thought to have a high likelihood of aortic regurgitation because of the stress distribution between the unequally sized cusps.¹⁰

Patients with QAVs may present with symptoms of aortic regurgitation such as dyspnea on exertion and angina and some clinical signs such as widened pulse pressure and a diastolic murmur.^{2,7}

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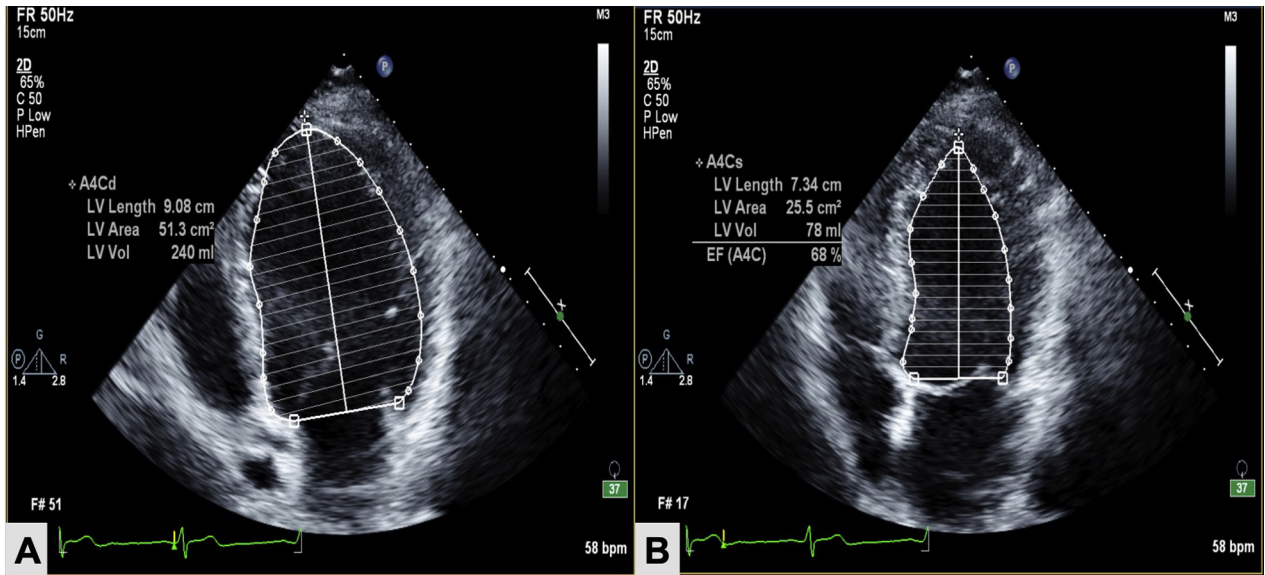


Figure 1 Transthoracic echocardiographic image showing a dilated left ventricle with an end-diastolic volume of 240 mL (A) and left ventricular end-systolic volume of 78 mL (B).

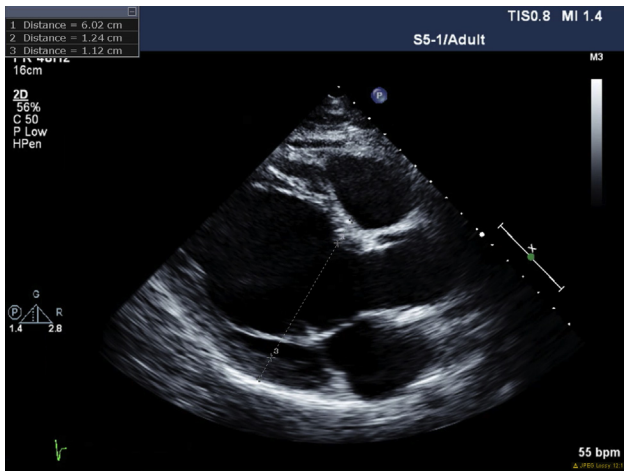


Figure 2 Transthoracic echocardiographic image, parasternal long-axis view, of the left ventricle showing a dilated left ventricle.

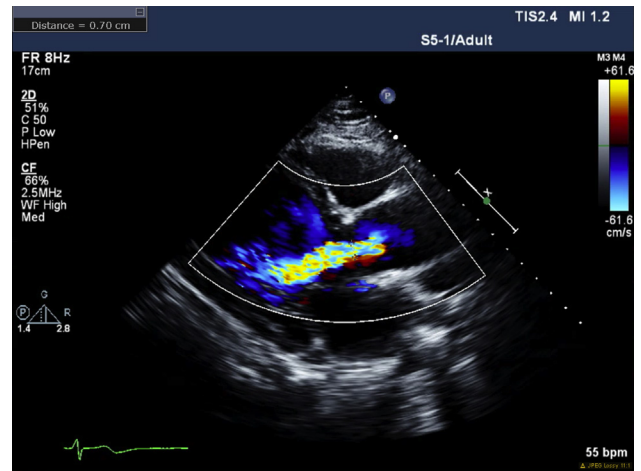


Figure 3 Transthoracic echocardiographic parasternal long-axis view with color Doppler showing severe aortic regurgitation with a vena contracta width of 0.7 cm².

Echocardiography is the standard diagnostic imaging modality used in patients with suspicion of valvular heart disease.^{6,11} TEE is a preferred diagnostic tool because of its ability to delineate the valve's morphology and functionality, and it can also assess aortic root size and identify coronary ostia.¹ However, despite the use of echocardiography, in some cases, the diagnosis is not made until the valve is directly visualized during surgery or autopsy.²

The most common indications for surgery are severe aortic regurgitation, severe aortic stenosis, or dysfunctional QAV with other le-

sions, such as an occluded left coronary ostium.¹ Most patients undergo aortic valve replacement for chronic severe aortic regurgitation.^{5,6} The most common repair technique is aortic valve "tricuspidization," in which the accessory cusp is resected and the remaining three cusps are conjoined. Similarly, "bicuspidization" has been reported, in which the valve commissural closures are sutured to form a bicuspid aortic valve.^{5,12}

It is unclear whether patients with QAVs have increased risk for infective endocarditis, though it has been reported.^{1,13} Some case re-



Figure 4 Two-dimensional transesophageal echocardiographic image showing short-axis aortic valve in systole with four distinct equally sized aortic valve cusps (Hurwitz and Roberts subtype A).

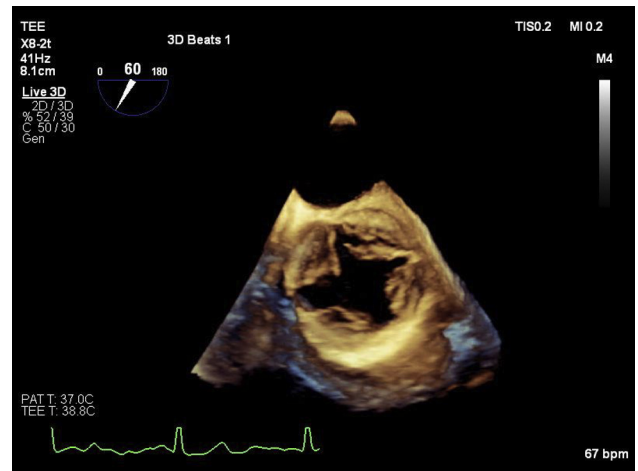


Figure 6 Three-dimensional transesophageal echocardiographic images of QAV in short axis showing four distinct aortic valve leaflets.

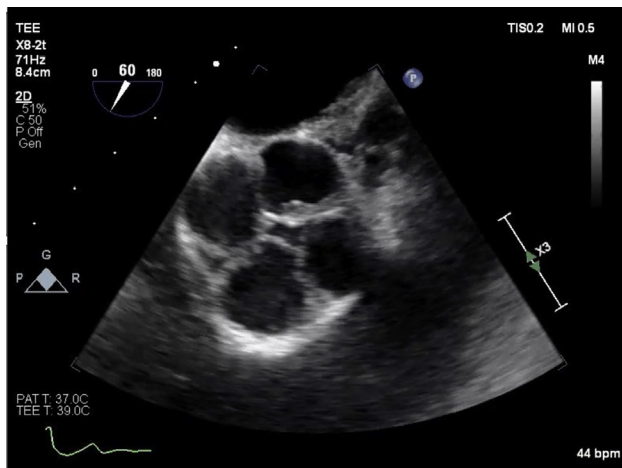


Figure 5 Two-dimensional transesophageal echocardiographic image showing the QAV closed in diastole. Seen is the X formation of the valve, instead of the upside down Y (Hurwitz and Roberts subtype A). See [Video 2](#).

ports have suggested that the risk is increased with valves with smaller accessory cusps because of an unequal stress distribution and abnormal leaflet coaptation.^{1,13} Currently, there is no recommendation for infective endocarditis prophylaxis for these patients.¹⁴

CONCLUSION

QAV is a rare cause of aortic regurgitation with an incidence of <1%. Echocardiography, especially TEE, is integral in identifying associated abnormalities such as a displaced coronary ostium. Patients with this anomaly often undergo aortic valve replacement for symptomatic valvular disease, left ventricular systolic dysfunction, or associated structural abnormalities found to be associated with the valve.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2020.04.002>.

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