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Quadricuspid Aortic Valve: A Rare Congenital Anomaly Detected on Routine Echocardiography

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I. INTRODUCTION

Quadricuspid aortic valve (QAV) is an uncommon congenital malformation in which the aortic valve consists of four cusps of equal or unequal size. It is clinically important, as many patients eventually develop progressive regurgitation [1]. The mean age at diagnosis is reported to be around the fourth decade of life, although cases are increasingly being recognized in younger individuals with the availability of high-quality echocardiography.[2]

While bicuspid and unicuspid valves are more commonly recognized congenital anomalies, QAV is clinically important because of its association with AR, infective endocarditis, and, in some cases, aortic dilatation.[1,3] Recognition of QAV remains challenging; it is often missed on transthoracic echocardiography (TTE) and sometimes diagnosed only intraoperatively or post-mortem. [4]

We present the case of a 21-year-old male with incidentally detected QAV on routine evaluation, who had been misdiagnosed as having a trileaflet valve with trivial AR across several years of follow-up. This case underscores the diagnostic difficulty and highlights the importance of accurate recognition for long-term surveillance and prevention of complications.

II. CASE REPORT

A 21-year-old Indian male presented to the cardiology outpatient clinic for a routine health check-up. He was asymptomatic at the time of evaluation.

He had a past history of being evaluated for a febrile illness in childhood, during which an echocardiogram suggested a trileaflet aortic valve with trivial aortic regurgitation. Since then, he had been kept under periodic echocardiographic follow-up.

Between 2018 and 2022, the patient underwent six serial transthoracic echocardiograms, all of which consistently reported a trileaflet aortic valve with trivial to mild regurgitation. In October 2024, however, transthoracic echocardiography clearly demonstrated a quadricuspid aortic valve (QAV) in the parasternal short-axis view, with four cusps. Parasternal short-axis echocardiography revealed a quadricuspid aortic valve with unequal cusps, corresponding to Hurwitz and Roberts' Type D classification (Fig. 1). During systole, the valve demonstrated a rectangular opening consistent with quadricuspid morphology (Fig. 2). Color Doppler confirmed central turbulence at the point of cusp closure, in keeping with mild aortic regurgitation (Fig. 3).

Table 1: Serial Echocardiogram Findings (2018–2024)

Date	Findings
10/02/2018	Trileaflet AV; trivial–mild AR; grade I MVP of AML & PML; trivial MR; preserved LV size and function (EF 66%); no AS, no PAH, no shunt lesions
13/08/2018	Trileaflet AV; trivial AR; likely congenital
20/09/2019	Trileaflet AV; trivial AR; no AS
09/04/2021	Trileaflet AV; trivial AR
17/12/2021	Trileaflet AV; trivial AR; normal biventricular function
29/06/2022	Trileaflet AV; trivial AR; congenital aortic valve disease
02/10/2024	Quadricuspid AV (Type D); mild AR (PHT 613 ms); normal aortic annulus (16 mm) and ascending aorta (20 mm); preserved LV systolic (EF 74%) and diastolic function; trivial MR; RA/RV dilation; tricuspid valve prolapse with mild TR; normal PA pressures; trivial PR; normal RV function (TAPSE 2.6 cm)

Associated findings included mild AR (pressure half-time 613 ms), normal aortic annulus (16 mm) and ascending aorta (20 mm), preserved left ventricular systolic and diastolic function (EF 74%), and trivial mitral regurgitation. The right atrium and right ventricle were mildly dilated with evidence of tricuspid valve prolapse and mild tricuspid regurgitation, but pulmonary artery pressures remained normal. No vegetations, pericardial effusion, or thrombus were noted.

The patient remained clinically stable and asymptomatic. He was counselled regarding the risk of infective endocarditis, advised prophylaxis before invasive procedures, and instructed to promptly treat any trivial infections. Periodic echocardiographic surveillance was recommended to monitor valve function and chamber dimensions.

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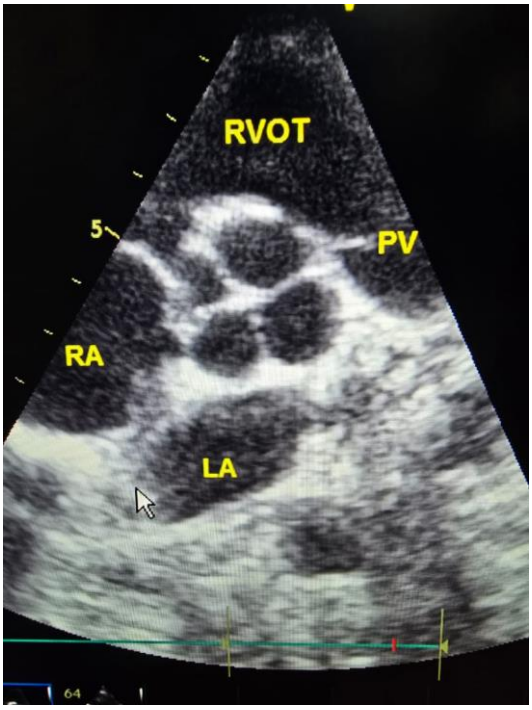

Figure 1. Parasternal short-axis echocardiographic view demonstrating a quadricuspid aortic valve (QAV) with one small cusp, one large cusp, and two intermediate cusps, corresponding to Hurwitz and Roberts' Type D classification

Parasternal short axis view demonstrating qav with one small cusps, one lrage and 2 intermediate cusps – class D (Fig. 1)

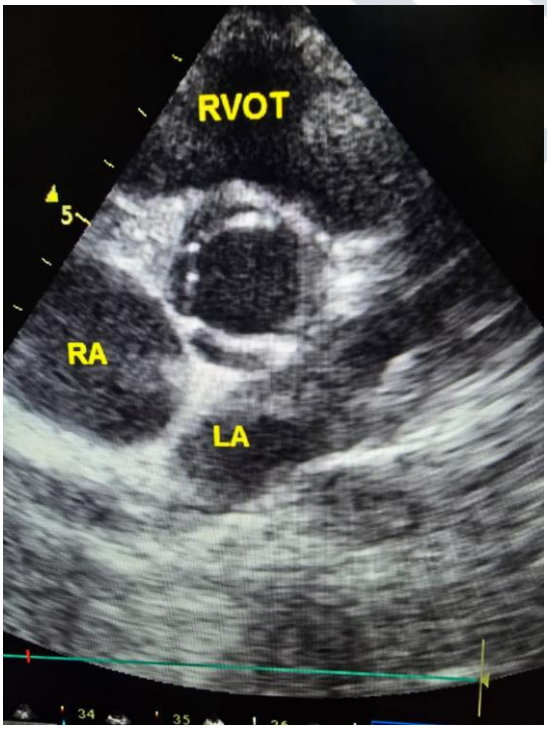

Figure 2. Systolic frame showing the rectangular opening of the aortic valve, confirming the presence of a quadricuspid morphology

During systolic aortic valve opening is rectangular in shape confirming QAV (Fig. 2)

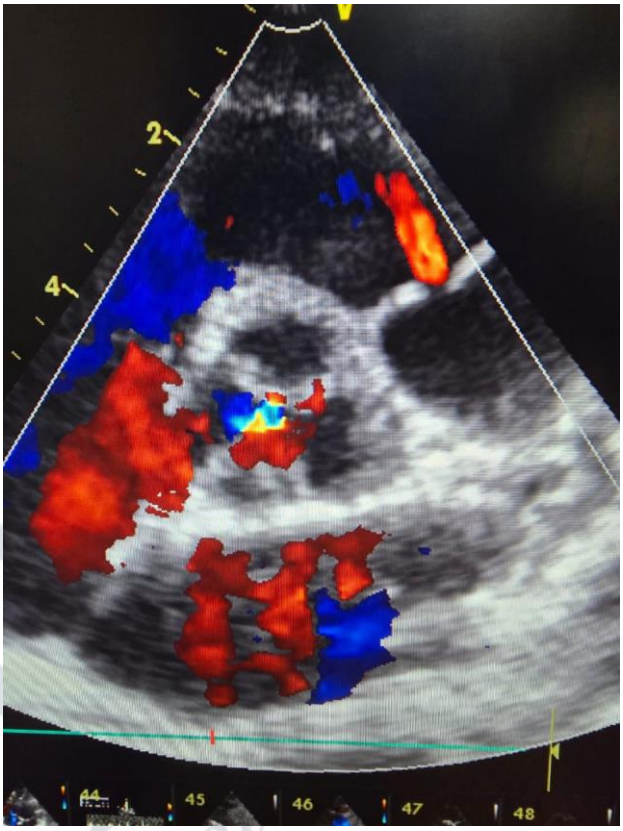

Figure 3. Color Doppler image demonstrating central turbulence at the point of cusp closure, consistent with mild aortic regurgitation

Colour Doppler reveal AR turbulence at the center of closure point (Fig. 3)

III. DISCUSSION

Quadricuspid aortic valve (QAV) is a rare congenital malformation, with an incidence of 0.01%–0.04% in echocardiographic and autopsy studies and up to 0.05%–1% among patients undergoing aortic valve replacement for aortic regurgitation (AR).[2,3] In many cases, the diagnosis is delayed or even missed, as the morphology may be difficult to appreciate on routine transthoracic echocardiography (TTE). In our patient, multiple echocardiograms performed between 2018 and 2022 consistently described a trileaflet valve with trivial AR, and the diagnosis of QAV was established only in 2024.

The reasons for this diagnostic challenge are multifactorial. First, routine TTE assessments often prioritize ventricular function and regurgitation severity rather than careful evaluation of cusp morphology. Second, operator dependency, older-generation echocardiography machines, and limited awareness of this rare anomaly likely contributed to misclassification. This limitation has been well recognized, as QAV can be missed even on high-quality TTE

and sometimes only diagnosed intraoperatively or post-mortem.[4] Advanced imaging techniques, including three-dimensional TTE and TEE, have been shown to significantly improve diagnostic accuracy in such cases. [5]

Accurate recognition of QAV is clinically important. While many patients remain asymptomatic in early adulthood, the condition carries a strong association with progressive AR, as observed in our case. Malcoaptation of unequal cusps results in progressive eccentric regurgitant jets, which may progress insidiously and eventually require surgical correction.

According to the 2023 ESC Guidelines for the management of endocarditis, routine antibiotic prophylaxis is not recommended for most congenital valve anomalies, except in high-risk patients such as those with prosthetic valves, prior IE, or specific cyanotic congenital lesions.[6] Although the absolute risk of infective endocarditis remains debated, cases of endocarditis in QAV have been reported, particularly when cusp malalignment leads to turbulent flow.[7] In light of this, our patient was advised endocarditis prophylaxis prior to invasive procedures and was counselled regarding the prompt management of even minor infections.

This case highlights the importance of maintaining a high index of suspicion for QAV in patients with unexplained or disproportionate AR, especially when serial echocardiograms yield inconsistent findings. Early and accurate diagnosis allows for tailored surveillance, appropriate counselling, and timely surgical referral if significant regurgitation develops.

IV. CONCLUSION

Quadricuspid aortic valve is a rare congenital anomaly that may be overlooked on routine transthoracic echocardiography, as seen in our patient who was repeatedly reported to have a trileaflet valve on serial studies. Recognition of this condition is clinically significant, since it is strongly associated with progressive aortic regurgitation and may predispose to infective endocarditis. Our case emphasizes the importance of meticulous echocardiographic assessment, particularly with parasternal short-axis views, to avoid misdiagnosis. Accurate identification allows for timely counselling, institution of preventive measures such as endocarditis prophylaxis, and appropriate surveillance of valve function and ventricular dimensions. Early detection is critical for guiding long-term management and surgical planning, thereby improving outcomes in these patients.

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