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REPORTE DE CASO / CASE REPORT

# Válvula aórtica cuadricúspide: reporte de caso y breve revisión

*Quadricuspid aortic valve: case report and brief review*

Tamar Gaprindashvili, MD[a]✉; Zaza Katsitadze, PhD[a]; Rusudan Agladze, PhD[a]; Nino Nersezashvili, MD[a].

[a] Bokhua Memorial Cardiovascular Center, Tbilisi, Georgia.

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**RESUMEN**

La válvula aórtica cuadricúspide (VAC) es una anomalía congénita poco común, generalmente asociada con insuficiencia aórtica. Presentamos el caso de un hombre de 64 años con disnea progresiva, diagnosticado con VAC mediante ecocardiografía transesofágica intraoperatoria. Se realizó un reemplazo valvular aórtico con bioprótesis debido a su preferencia y factores de riesgo. El objetivo de este artículo es presentar un caso raro de válvula aórtica cuadricúspide asociada a una arteria coronaria derecha anómala, destacando los desafíos diagnósticos, las implicaciones quirúrgicas y las lecciones de manejo.

**ABSTRACT**

Quadricuspid aortic valve (QAV) is a rare congenital anomaly, often associated with aortic regurgitation. We present the case of a 64-year-old man with progressive dyspnea, diagnosed with QAV through intraoperative transesophageal echocardiography. A bioprosthetic aortic valve replacement was performed due to patient preference and risk factors. The objective of this article is to present a rare case of quadricuspid aortic valve associated with an anomalous right coronary artery, highlighting diagnostic challenges, surgical implications, and management lessons.

✉ Autor para correspondencia
Correo electrónico: tamunaphd@gmail.com

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## INTRODUCTION

Congenital quadricuspid aortic valves (QAV) are a very rare condition, with approximately two hundred cases reported in the literature since the first case was reported in 1862[1], whereas bicuspid valves are relatively common, occurring in 1-2% of the population, and are well studied[2,3]. The functional status of QAV is predominantly pure aortic regurgitation. All clinical manifestations of the disease depend on the functional status of the valve and concomitant disorders. The condition is predominantly revealed in the fifth to sixth decade of life. Stenotic QAV is very rare and is seen in almost 0.7% of the patients. Due to the growing progress of echocardiography and other imaging diagnostic techniques, QAVs are increasingly reported[4]. As Anand D. Jagannath[5] has noted, unlike patients with Bicuspic aortic valves (BAV), whose aortic valves often have associated enlargement of the ascending aorta secondary to medial disarray[5], adult patients with "QAV" in their series did not have an enlarged ascending aorta (size 26-34 mm). In addition, ascending aortic enlargement has only been reported three times in the literature[6,7].

Two classification systems are widely used. Hurwitz and Roberts have described up to eight morphological subtypes based on cusp size[8], while Nakamura et al. categorize QAV according to the position of the supernumerary cusp[9]. These classifications are clinically relevant, as different morphologies have distinct hemodynamic implications and may explain the variability in presentation, ranging from asymptomatic cases to severe regurgitation requiring surgery.

One more clinical concern is the possible linkage of QAV with other developmental disorders, such as anomalies of the coronary arteries. The QAV with coronary artery anomalies is very rare, with only a handful of cases published, thus reinforcing the exceptional nature and great clinical importance of the case at hand. In the available literature, very few cases have been documented that demonstrate this association, thus making the case presented more interesting and adding to the scarcity of literature. Though rare, these associations are crucial, as they may hinder diagnosis and greatly increase the risk during operative procedures.

This article aims to demonstrate the uniqueness of QAV with an absent right coronary artery, noting the challenges in diagnosis, the hemodynamic consequences, and the surgical approach to the case. By highlighting this combined anomaly, it is our aim to contribute to the understanding of QAV beyond its rarity and provide lessons for clinical practice.

### QAV classification

Two types of classification of QAV are established in the literature: the Hurwitz & Roberts[8] classification and the Nakamura et al. classification[9]. The first of the classifications relates to the relative dimension of the extra cusp, classifying QAVs into seven types from A to G, having been enhanced to eight types by Vali et al.[10] (Figure 1)[11]. Types A, B, and C account for more than 85% of the cases, while type D is very rare [5]. Nakamura et al.[9]

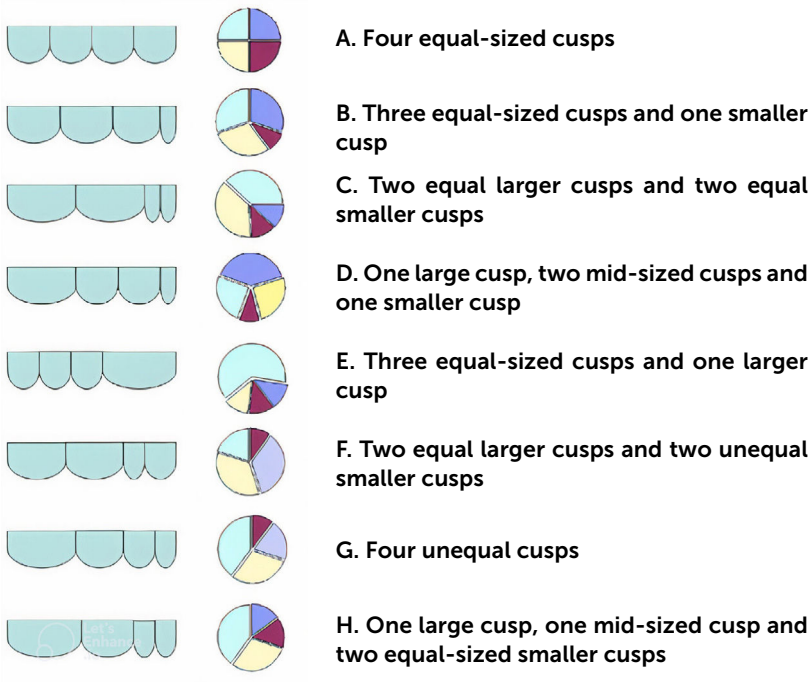


**Figure 1.** Classification of quadricuspid aortic Valve with Vali

Hurwitz & Roberts[8] classification of quadricuspid aortic Valve with Vali et al[10]. Supplement (reprinted from: Yuan SM.[11])

proposed a shortened classification focused on the position of the supernumerary cusp: type I, supernumerary cusp between the left and right coronary cusps; type II, supernumerary cusp between the right and non-coronary cusps; type III, supernumerary cusp between the left and non-coronary cusps; and type IV, unidentified supernumerary cusp as two equal-sized smaller cusps (Figure 2).

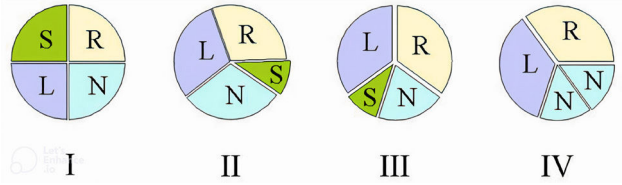


**Figure 2.** Simplified classification of quadricuspid aortic valve

Nakamura et al[9] . simplified classification of quadricuspid aortic valve. L=left coronary cusp; N=non-coronary cusp; R=right coronary cusp; S=supernumerary cusp (reprinted from: Yuan SM.[11]).

## CASE REPORT

A 64-year-old man was referred with a 2-month history of dyspnea (NYHA class II); he had no history of endocarditis or rheumatic disease other than grade II hypertension. By the time he was admitted to our hospital to develop a treatment strategy, he was receiving medical therapy for his hypertension (perindopril 8 mg, amlodipine 10 mg) and diuretics for volume overload (furosemide 80 mg)).

On physical examination, HR was 67 bpm; his blood pressure was 160/80 mmHg, and cardiac auscultation revealed a high-pitched, early diastolic decrescendo murmur heard at the 3rd intercostal space on the left (Erb's point) at end expiration with the patient sitting up and leaning forward. A 12-lead electrocardiogram showed a sinus rhythm, a regular and incomplete left bundle branch block, and signs of left ventricular hypertrophy. During transthoracic echocardiography (TTE), it was possible to confirm concentric left ventricular hypertrophy (LV mass: -405 g, RWT: -0.52, LV mass index: 201 g/m$^2$), but left ventricular cavity enlargement was not revealed (left ventricular end diastolic dimension (LVEDD): 58 mm [42.0-58.4], LVESD 31 mm [25.0-39.8])[12], left ventricular ejection fraction (EF) was mildly abnormal (48% [41-51])[12]; A severe aortic regurgitation diagnosis was established (large central jet, vena contracta 5-8 mm, jet/LVOT ratio -65%, EROA - 35 mm$^2$)[13], according to the patient's BSA (2.062 m$^2$) evidence of dilatation of the sinus of valsalva (42 mm) and ascending aorta dilatation (41 mm) was found; with no history of endocarditis or rheumatic disease. Given the severe central aortic regurgitation in the absence of any history of rheumatic or infectious endocarditis, together with leaflet thickening and deformation, a congenital valvular anomaly — including the possibility of a QAV — was suspected. TTE visualization and valvular deformation did not allow distinguishing the quantity of cusps.

Intraoperative transesophageal echocardiography showed, in the mid-esophageal right ventricular inflow-outflow view (ME RV inflow-outflow) and mid-esophageal aortic valve short axis view (ME AV SAX), a quadricuspid aortic valve with the presence of four cusps of equal size and a "+-shaped" commissural aspect in diastole (Figure 3). (Type A by Hurwitz & Roberts[8] classification and Type I by Nakamura et al.[9] classification). There was a raphe between the S-L and R-N cusps (Figure 4).

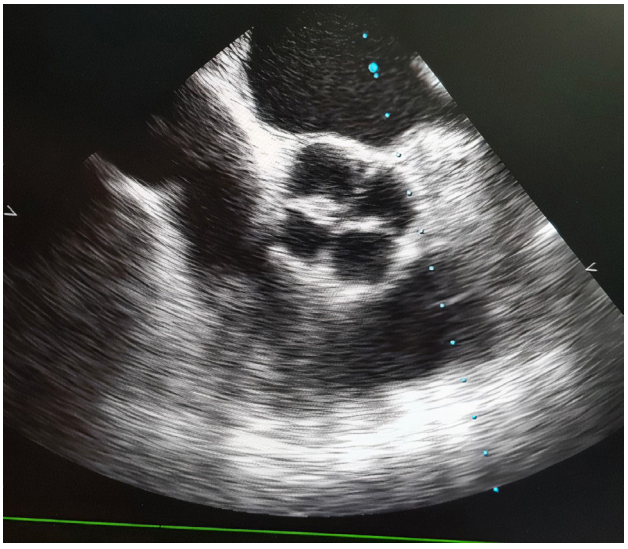

**Figure 3.** SMid –esophageal aortic valve in diastole short axis view (ME AV SAX)

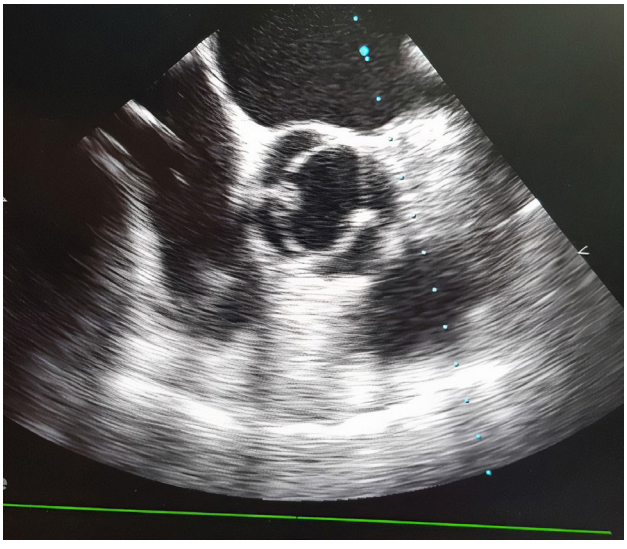

**Figure 4.** SMid –esophageal aortic valve in systole short axis view (ME AV SAX)

There was an incomplete diastolic coaptation insufficiency and a central, severe aortic regurgitation jet. The sinus valsalva and proximal ascending aorta were enlarged (diameters: 42 mm and 41 mm, respectively)[12].

The intraoperative TEE findings of four equal cusps and a raphe were confirmed by direct surgical inspection (Figure 5) and subsequent pathology, which reported four independent cusps. This confirmed the valve as type A by Hurwitz & Roberts[8] and type I by Nakamura et al[9]. classifications. Coronary angiography was normal except for an abnormal displacement of the ostium of the right coronary artery from the right anterior coronary sinus, inferring an anomalous right coronary artery (RCA), which was confirmed during surgery.

The patient was informed about the choice of the aortic prosthesis options, taking into consideration the risk of anticoagulation-related bleeding and thromboembolism with a mechanical valve versus the risk of structural valve deterioration with a bioprosthesis. After a thorough discussion between the informed patient, cardiologists, and surgeon regarding the patient's lifestyle and preferences and taking into consideration the constraints regarding the patient's unwillingness and compliance problems regarding a future good long-term anticoagulant control, a bioprosthesis was chosen. The patient underwent elective aortic valve replacement with a biological prosthesis (Hancock II Ultra Bioprosthesis T505; diam. 25 mm); intraoperative assessment confirmed a quadricuspid aortic valve (Figure 5) and the displacing of a right coronary artery. The diameter of the ascending aorta was reduced (aortoplasty). The pathology report stated the presence of four independent cusps with hyalinosis, fibrosis, and calcification.

After 2 months, the patient remained asymptomatic.

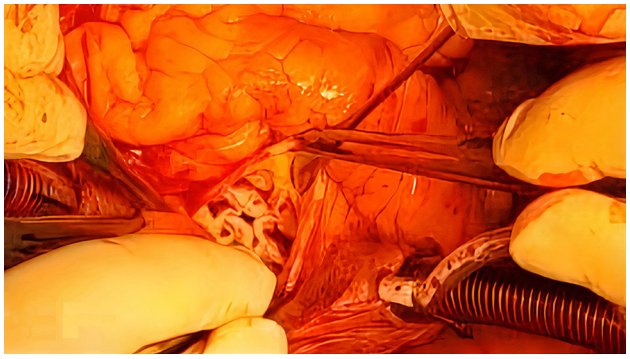

**Figure 5.** Intraoperative assessment of a quadricuspid aortic valve

## DISCUSSION

The case that we state in this article addresses the diagnostic difficulties, clinical features, and management of a rare case of congenital QAV. In our observation, the recently noted increase of QAV cases diagnosed in recent years seems to correlate with technological advances in imaging, specifically with ultrasounds and cardiac MRIs. Nowadays, all these means allow for better detection and differentiation of valve types, which in the past could only be made during surgery or autopsy[8,9].

This case contributes evidence supporting a possible association between QAV and coronary anomalies, a relationship rarely reported. Recognizing this link may prompt more systematic coronary screening in QAV patients and improve surgical planning.

As in our patient, QAV is most commonly seen with aortic regurgitation. The additional regurgitation seen is due to the poor leaflet coaptation and can result in overload of the left ventricle (LV) volume, dilatation, and eventually LV dysfunction. With regard to our patient, he had a considerable amount of LV hypertrophy but no dilatation, which could indicate compensatory mechanisms at play early on in the disease. This is in line with the literature, which states that in QAV, the natural history is highly variable, and some patients never become symptomatic for decades while others progress rapidly to severe valvular dysfunction that requires surgical correction[5]. Unlike bicuspid aortic valves, which frequently present with stenosis or mixed disease due to calcification and fibrosis, the functional status of QAV is predominantly pure aortic regurgitation. This is attributed to malcoaptation of the often-unequal cusps or leaflet redundancy, as seen in our patient. This difference in predominant pathophysiology is a key distinguishing feature between these two congenital defects. While QAV and BAV have comparable long-term results concerning valve malfunction and surgical timing, QAV is more frequently characterized by regurgitation rather than stenosis.

The surgical indication in this case was driven by the presence of severe symptomatic aortic regurgitation (NYHA Class II dyspnea) with evidence of left ventricular response (significant hypertrophy and a mildly reduced ejection fraction), in accordance with current valvular heart disease guidelines[13].

We can ascertain that the selection of surgical procedures continues to be patient-focused and is based on the overall health, age, and lifestyle factors of the patient. In our specific case, the use of anticoagulation therapy for long periods of time was not preferable for the patient; therefore, a bioprosthetic valve was selected instead of a mechanical valve. This treatment approach is in line with

recommendations for older patients with poor adherence to anticoagulation management[14]. Although bioprosthetic valves are not as durable as mechanical valves, they are associated with important quality-of-life benefits in some patients.

Moreover, the presence of an anomalous right coronary artery ostium in this patient is interesting because coronary anomalies are uncommon but important anomalies to have with congenital valve defects. The recognition and thorough assessment of such abnormalities are very important to the construction of the surgical strategy to minimize chances of complications during surgery[15].

This highlights the importance of detecting such anomalies prior to surgery as opposed to during surgery. Intraoperative-findings tend to raise complications, which is the reason for early diagnosis as the safest surgical strategies with the least risk of complications can be planned ahead of time. follow up, the necessity for endocarditis prophylaxis, and the counseling of patient on the possible progression of their condition may also be influenced by this.

To sum it up, this case serves as an example regarding the need for meticulous detailed, complex preoperative imaging, particularly for patients with QAV, to fully assess the cardiac valves and coronary arteries. Although the combination of QAV and an anomalous RCA is rather uncommon, it serves as an example of the complex embryological and surgical difficulties posed by congenital heart anomalies. The more longitudinal data which is collected on this patient population, the better the risk assessment, timing of surgery, and overall outcome.

*Declaración de Conflictos de Interés:*
*Los autores declaran que no existen conflictos de interés con respecto a la publicación de este artículo. The authors declare that there are no conflicts of interest with respect to the publication of this article.*

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