SCIENTIFIC LETTER

QUADRICUSPID AORTIC VALVE: A CASE REPORT IN A REFERRAL HOSPITAL IN SOUTHERN COLOMBIA

ABSTRACT

A quadricuspid aortic valve is a rare congenital cardiac anomaly that is often diagnosed incidentally and may progress to significant valvular dysfunction during adulthood. We present the case of a 65-year-old woman admitted with retrosternal chest pain, fever, and hemoptysis, whose diagnostic evaluation revealed severe aortic regurgitation and significant coronary artery disease. The patient underwent surgical treatment with aortic valve replacement and coronary artery bypass grafting, during which a quadricuspid aortic valve was identified as an incidental intraoperative finding. The postoperative course was favorable, without major complications. This case highlights the importance of considering uncommon congenital valvular abnormalities in patients with severe aortic insufficiency and emphasizes the role of timely surgical management in achieving satisfactory clinical outcomes.

Keywords: *aortic valve, quadricuspid valve, congenital heart disease, echocardiography, cardiovascular surgery.*

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Received: 02/16/2026 ***Accepted: 03/21/2026*** ***https://doi.org/10.55200/raccv.v24.n2.0129***

INTRODUCTION

A quadricuspid aortic valve (QAV) is a rare congenital cardiac anomaly characterized by the presence of four aortic cusps instead of the normal tricuspid configuration. Since its first description in the nineteenth century, QAV has been considered an uncommon entity frequently identified incidentally during imaging studies, cardiac surgery, or autopsy examinations.[1] Improvements in cardiovascular imaging techniques, particularly transthoracic and transesophageal echocardiography, have increased the recognition of this condition in recent decades.[2]

Although a patient with a QAV may remain asymptomatic for many years, progressive valvular dysfunction commonly develops with advancing age, especially aortic regurgitation resulting from abnormal cusp coaptation and structural degeneration.[3] In some patients, the anomaly may also coexist with other congenital or acquired cardiovascular disorders, including coronary artery abnormalities and structural heart disease, which may influence clinical presentation and surgical management.[4]

Because of its low prevalence and variable clinical manifestations, QAV continues to represent a diagnostic and therapeutic challenge. Early identification is important to prevent progressive cardiac remodeling and to determine the appropriate timing for surgical intervention in patients with significant valvular dysfunction.[5]

We present the case of a patient with severe aortic regurgitation and concomitant coronary artery disease in whom a quadricuspid aortic valve was identified incidentally during cardiac surgery at the Hospital Universitario Hernando Moncaleano Perdomo, a tertiary referral center in southern Colombia.

CASE REPORT

A 65-year-old patient from the municipality of Pitalito was admitted with a chronic clinical presentation characterized by retrosternal chest pain radiating to the dorsal region, rated 6/10 on the subjective pain scale, intermittent, self-limited, and unrelated to physical exertion or emotional stress. During the previous 10 days, the symptoms had become associated with unquantified febrile episodes and cough, initially dry and later accompanied by hemoptysis.

The patient initially sought medical attention at a secondary-level healthcare institution, where a diagnosis of acute bronchitis was established. Antibiotic therapy with ampicillin/sulbactam was initiated, and further diagnostic studies were requested, revealing severe aortic regurgitation. Consequently, the patient was referred to a higher-level institution for evaluation and management by the cardiovascular surgery service.

Medical history

The patient reported a medical history significant for type 2 diabetes mellitus, currently managed with insulin glargine and metformin; arterial hypertension treated with losartan and amlodipine; left inguinal herniorrhaphy; exploratory laparotomy secondary to a stab wound; hysterectomy; and a history of tobacco use.

Clinical findings

Upon admission, the patient was in fair general condition, alert, oriented to person, place, and time, and asymptomatic. Vital signs were as follows: blood pressure 120/53 mmHg, heart rate 67 beats per minute, respiratory rate 18 breaths per minute, oxygen saturation 97%, temperature 36.7 °C, and blood glucose level 152 mg/dL.

Physical examination revealed a normocephalic patient with moist mucous membranes. The neck was symmetric and mobile, without jugular venous distention or carotid pulsations. Cardiac auscultation revealed normal, regular heart sounds with a low-grade diastolic murmur at the aortic focus. Breath sounds were preserved bilaterally without adventitious sounds. The abdomen was soft and depressible, with no palpable masses or organomegaly and no signs of peritoneal irritation. The extremities were symmetric and eutrophic, with a capillary refill time of 2 seconds and palpable distal pulses, without motor or sensory deficits.

Diagnostic assessment

The patient was evaluated by the cardiovascular surgery service, which requested additional diagnostic studies to establish the appropriate management plan.

The patient's laboratory workup demonstrated preserved hematologic parameters without leukocytosis or thrombocytopenia. Renal function was consistent with stage II chronic kidney disease, while liver function tests showed hypoalbuminemia with otherwise unremarkable hepatic parameters. Hyperglycemia was also documented during admission. Serologic studies for hepatitis B virus, HIV, and syphilis were negative, and urinalysis was not suggestive of infection. The complete laboratory findings are summarized in *Table 1*.

Carotid Doppler ultrasound findings were within normal limits. Transesophageal echocardiography demonstrated mild left ventricular dilatation,

Parameter	Patient value	Reference range	Interpretation
Complete blood count			
Leukocytes	8,400/µL	4,000-11,000/µL	Normal
Neutrophils	5,500/µL	1,500-7,500/µL	Normal
Hemoglobin	14.5 g/dL	12-16 g/dL	Normal
Platelets	302,000/µL	150,000-450,000/µL	Normal
Electrolytes			
Sodium	142.4 mmol/L	135-145 mmol/L	Normal
Potassium	4.16 mmol/L	3.5-5.1 mmol/L	Normal
Chloride	104 mmol/L	98-107 mmol/L	Normal
Renal function			
Blood urea nitrogen	14.14 mg/dL	7-20 mg/dL	Normal
Creatinine	0.97 mg/dL	0.6-1.2 mg/dL	Normal
Estimated GFR	61.49 mL/min/1.73 m^2	>90 mL/min/1.73 m^2	Decreased
Liver function tests			
Total bilirubin	0.33 mg/dL	0.2-1.2 mg/dL	Normal
Direct bilirubin	0.12 mg/dL	0-0.3 mg/dL	Normal
Indirect bilirubin	0.21 mg/dL	0.2-0.8 mg/dL	Normal
AST	22.7 U/L	10-40 U/L	Normal
ALT	42 U/L	7-56 U/L	Normal
Total proteins	5.19 g/dL	6.0-8.3 g/dL	Decreased
Albumin	2.46 g/dL	3.5-5.0 g/dL	Decreased
Alkaline phosphatase	85 U/L	44-147 U/L	Normal
Prothrombin time	11.4 s	10-13 s	Normal
INR	1.04	0.8-1.2	Normal
Partial thromboplastin time	22.9 s	25-35 s	Slightly decreased
Metabolic and serologic tests			
Blood glucose	282.79 mg/dL	70-140 mg/dL	Increased
TSH	1.32 µIU/mL	0.4-–4.0 µIU/mL	Normal
HIV	Negative	Negative	Normal
Syphilis	Negative	Negative	Normal
Hepatitis B	Negative	Negative	Normal
Urinalysis			
Urinary sediment	No evidence of infection	Negative for infection	Normal

TABLE 1. Laboratory findings at admission.
ALT: alanine aminotransferase, AST: aspartate aminotransferase, GFR: glomerular filtration rate, HIV: human immunodeficiency virus, INR: international normalized ratio, TSH: thyroid-stimulating hormone.

normal-sized atrial chambers and right ventricle, with no intracardiac masses or thrombi. Adequate contractility of both ventricles was observed, with a left ventricular ejection fraction of 60%. Severe aortic regurgitation was identified, with an aortic annulus diameter of 3 cm. No sclerosis of the three valve cusps was reported, and the pericardium appeared normal without effusion.

Coronary angiography revealed severe single-vessel coronary artery disease involving the left anterior descending artery, with a 70% proximal stenotic lesion.

Therapeutic intervention

The patient was scheduled for mechanical aortic valve replacement combined with myocardial revascularization of the left anterior descending artery. During surgery, a quadricuspid aortic valve and proximal atherosclerotic disease of the left anterior descending artery were incidentally identified (*Figures 1* and *2*).

A median sternotomy was performed, followed by skeletonization of the left internal mammary artery. An inverted "T" pericardiotomy with suspension sutures was carried out. Double purse-string sutures with 3-0 Prolene were placed in the ascending aorta, and a single purse-string suture with 4-0 Prolene was placed in the right atrial appendage and right superior pulmonary vein.

After systemic heparinization at 3 mg/kg, arterial and bicaval cannulation were performed, and cardiopulmonary bypass was initiated. Following aortic cross-clamping, left heart decompression was achieved using a vent cannula. Aortotomy was performed, and Custodiol™ cardioplegic solution was administered through the coronary ostia until cardiac arrest was achieved.

The native aortic valve was resected, and a N.° 19 Sorin™ mechanical prosthetic valve was implanted. Exploration of the left anterior descending artery revealed an approximately 1.5 mm distal vessel bed. Arteriotomy and termino-lateral anastomosis with the left internal mammary artery were performed, confirming adequate distal flow.

Heparin reversal was achieved with protamine, followed by decannulation and hemostasis. Two mediastinal drainage tubes were placed. The pleurae were closed, and a temporary epicardial pacemaker lead was positioned in the right ventricle. Sternal closure was performed with steel wires, followed by routine layered closure using Vicryl and Monocryl sutures (*Figure 3*).

The postoperative course was uneventful, with no signs of cardiac tamponade and adequate urinary output.

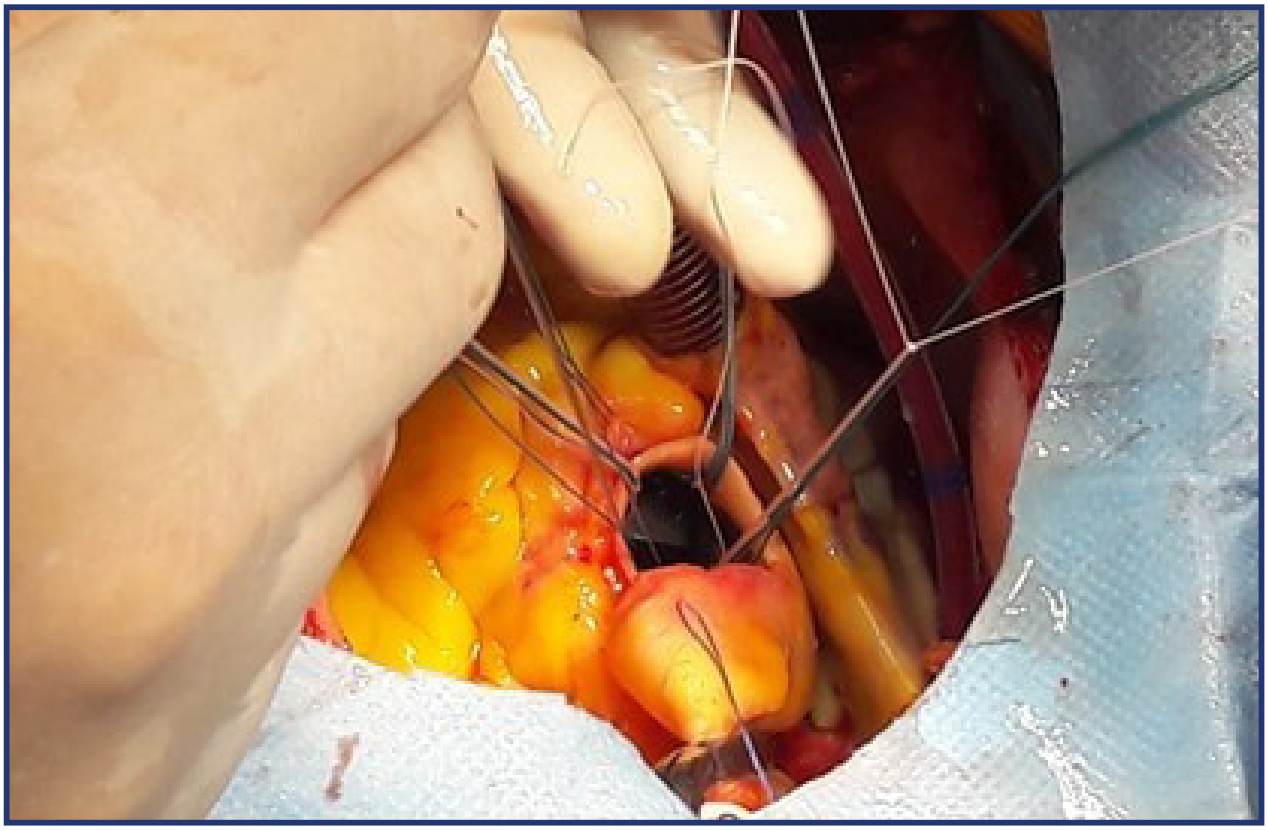

FIGURE 1. Surgical view of the quadricuspid aortic valve.

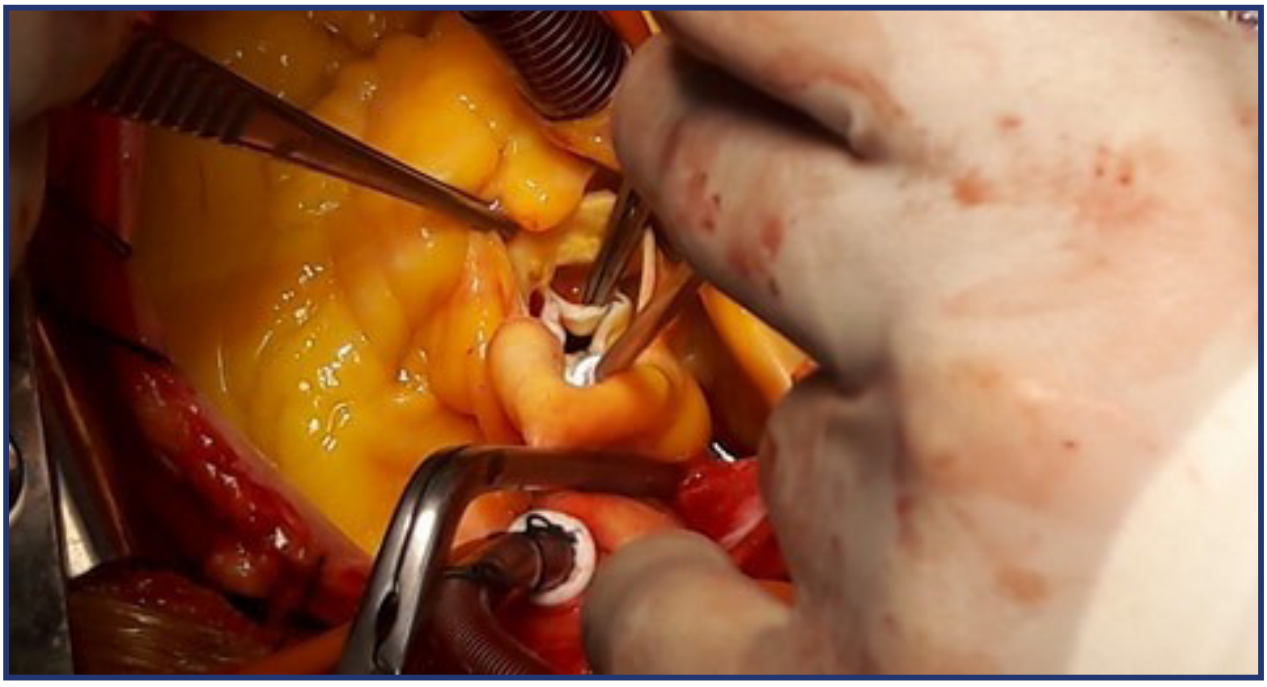

FIGURE 2. Quadricuspid aortic valve.

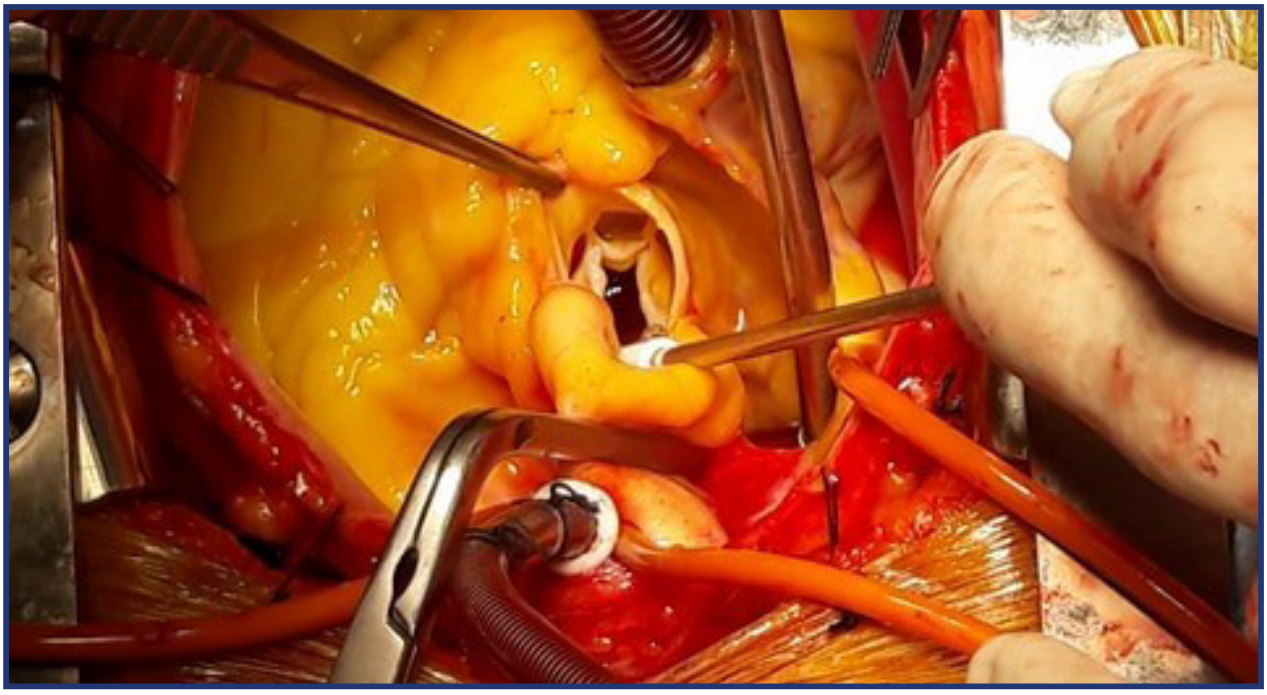

FIGURE 3. Aortic valve replacement.

DISCUSSION

The aortic valve has an average diameter of 23.3 mm in adult men and 21.6 mm in adult women. It is located at the outlet of the left ventricle, between the tricuspid/mitral valves and the pulmonary valve. It has a swallow's nest-like configuration and consists of three sinuses: the right coronary sinus, from which the right coronary artery arises; the left coronary sinus, from which the left coronary artery originates; and the non-coronary sinus.[3] The opening of the aortic valve marks the onset of ventricular systole. During this phase, the aortic cusps move in response to pressure changes, aligning parallel to the ascending aorta and allowing ventricular ejection. As systole progresses, the outflow decreases, promoting the formation of vortices between the aortic cusps and the sinus walls, thereby maintaining coronary perfusion and facilitating valve closure.[4]

Congenital anomalies of the aortic valve account for approximately 3-6% of congenital heart diseases

in adults. Among these, the bicuspid aortic valve is the most common, affecting nearly 2% of the population, followed by unicuspid and quadricuspid aortic valves.[5] This is the second documented case of a quadricuspid aortic valve at our hospital. The first case has already been published,[6] highlighting the importance of recognizing this rare congenital anomaly, its clinical manifestations, and the associated diagnostic and therapeutic implications.

The first case of quadricuspid aortic valve (QAV) was described by Balington in 1862 as an incidental autopsy finding. Later, in 1923, Simonds reported five cases of QAV among 25,666 autopsies. More recently, in 2004, Tutarel published studies based on autopsy findings and echocardiography-diagnosed cases.[7] Currently, QAV is recognized as an uncommon congenital cardiac anomaly, with a reported incidence of 0.008% in autopsy studies, 0.043% on two-dimensional transthoracic echocardiography, and approximately 1% among patients undergoing aortic valve surgery.[8]

QAV usually occurs as an isolated anomaly; however, it may also be associated with other congenital malformations, including ventricular septal defect, atrial septal defect, tetralogy of Fallot, mitral valve insufficiency, transposition of the great arteries, hypertrophic cardiomyopathy, subaortic fibromuscular stenosis, patent ductus arteriosus, and coronary ostial obstruction, the latter reported in nearly 10% of cases.[9]

The mechanism underlying this malformation remains poorly understood. Nevertheless, abnormalities during early truncal septation have been proposed, including aberrant fusion of the aorticopulmonary septum, abnormal mesenchymal proliferation, and asymmetric division of one of the three cusps, resulting in an unequal number of valvular primordia.[10]

Two morphological classification systems have been described for quadricuspid aortic valve (QAV): the Hurwitz and Roberts classification and the Nakamura classification (*Figures 4* and *5*). The Hurwitz and Roberts system, originally described in 1973, classifies QAV according to the relative size and configuration of the aortic cusps and remains the most widely used classification worldwide. Among the different anatomical variants described, types A, B, and C represent the most reported cases, whereas other subtypes are considerably less frequent. In the present case, the valve morphology appeared to correspond to type C, characterized by two larger and two smaller cusps.

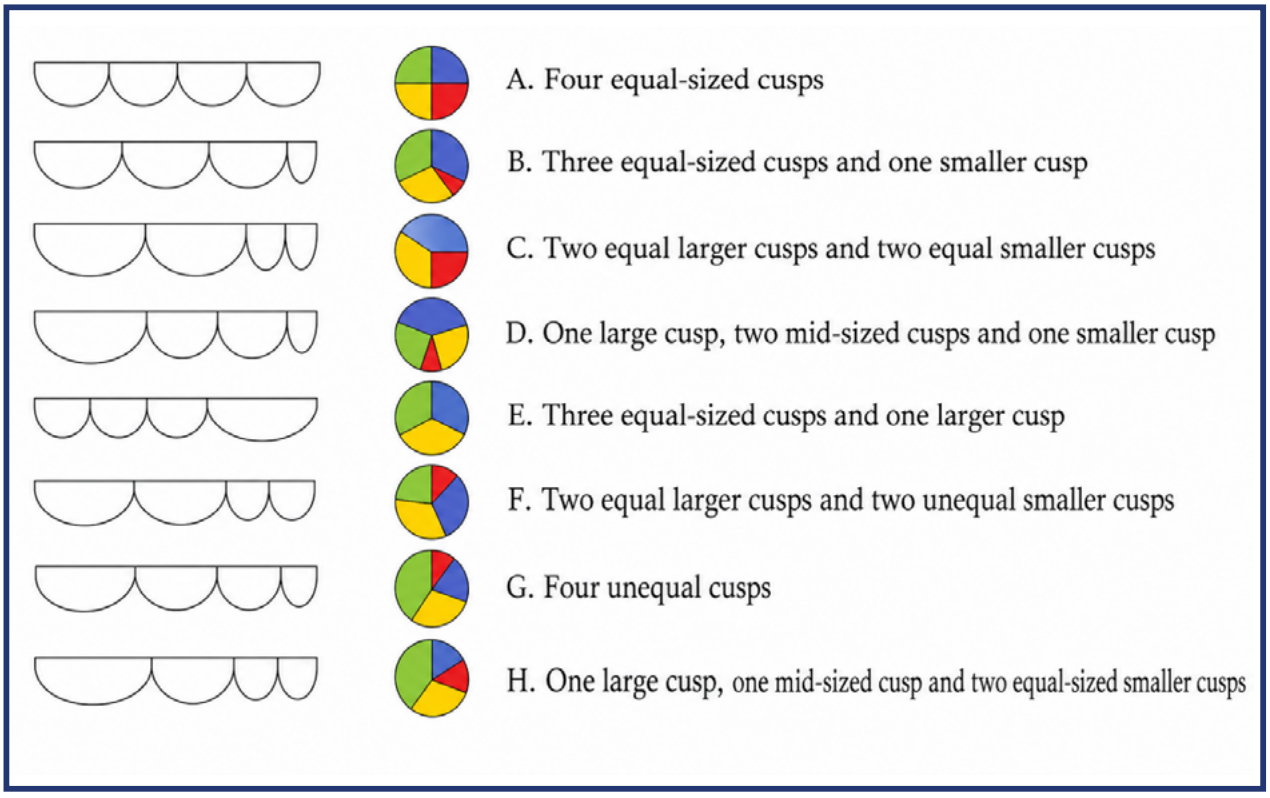

FIGURE 4. Modified Classification of Hurwitz and Roberts.
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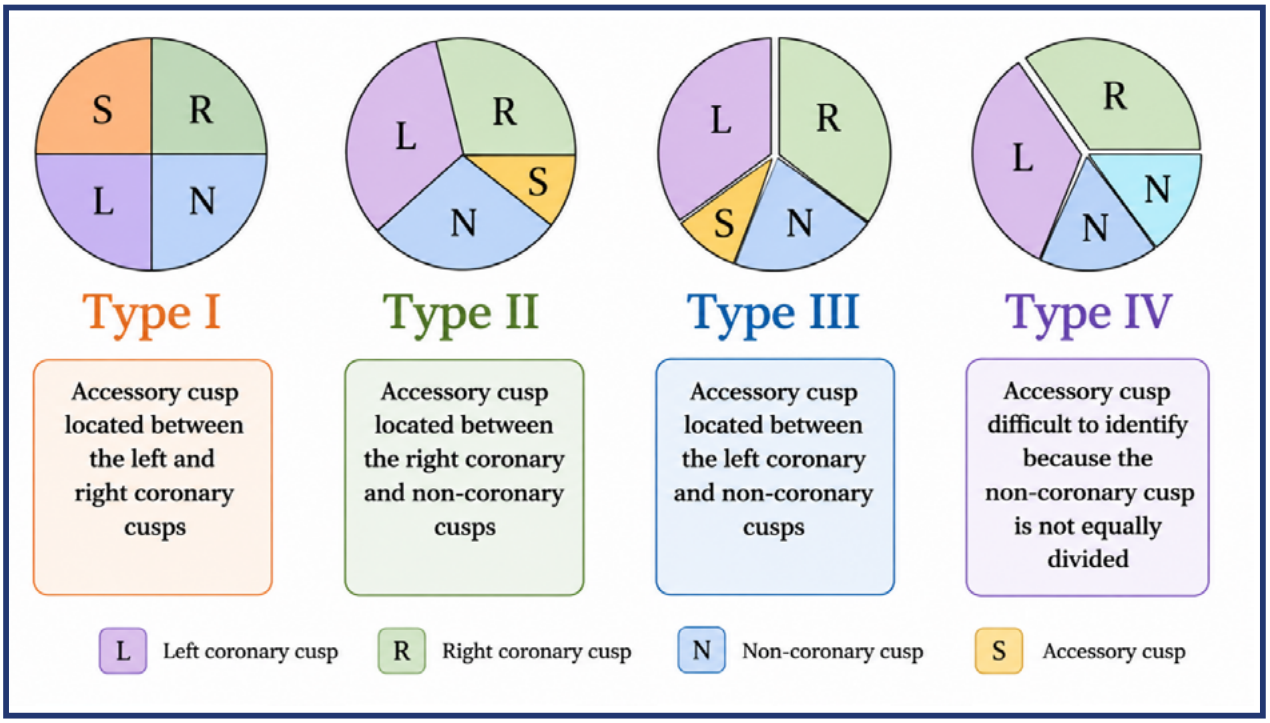

FIGURE 5. Nakamura classification.
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The Nakamura classification was later proposed as a complementary anatomical system based on the location of the accessory cusp in relation to the coronary and non-coronary cusps. This classification facilitates anatomical description and may contribute to surgical planning and echocardiographic interpretation. Together, the two classification systems provide a practical framework for describing the morphological variability of QAV and improving communication among clinicians, imaging specialists, and surgeons.

Clinical manifestations depend on the functional status of the quadricuspid valve and the presence of associated abnormalities. In most cases, diagnosis is incidental, since patients may remain asymptomatic until the sixth decade of life. The most frequently reported symptoms include palpitations, precordial pain, fatigue, asthenia, adynamia, lower extremity edema, syncope, and, in extreme cases, sudden cardiac death.[11]

Although QAV does not always produce hemodynamic compromise, approximately 44% of patients exhibit some degree of hemodynamic abnormality. Tutarel and Westhoff-Bleck demonstrated that the most common finding is aortic regurgitation, occurring in approximately 75% of cases, as observed in our patient. This is followed by regurgitation associated with aortic stenosis in 8.4% of cases, whereas isolated stenosis is extremely rare, with an incidence of 0.7%.[7]

Diagnosis is often made incidentally during cardiac surgery, as occurred in our patient.

Computed tomography angiography has demonstrated high accuracy compared with conventional coronary angiography. It allows precise assessment of QAV morphology, failed cusp coaptation, significant aortic regurgitation, coronary ostial location, aortic dimensions, and coronary artery status.[12]

Cardiac magnetic resonance imaging provides additional information regarding the severity of regurgitation. Patients with a regurgitant fraction greater than 33% generally require valve replacement, as well as left ventricular size, identification of aberrant coronary ostia, and associated anomalies.

Two-dimensional transthoracic echocardiography provides valuable information regarding aortic valve morphology, aortic root dimensions, and the left ventricule size.

Two-dimensional transesophageal echocardiography became available in the 1970s; however, it was not until 1984 that it was first used to diagnose QAV, when Herman described the classic parasternal short-axis image. In diastole, the valve exhibits the characteristic X-shaped closure pattern, unlike the Y-shaped closure of a normal tricuspid aortic valve. In systole, the valve

opening appears rectangular rather than triangular. Additionally, this modality provides further details on valve morphology, including cusp prolapse.[12]

Surgical indications include severe aortic regurgitation, severe aortic stenosis, or dysfunctional QAV associated with other lesions, such as left coronary ostial obstruction.[8] The goal of aortic valve repair is to restore adequate cusp coaptation and maintain a low transvalvular gradient without turbulent flow, thereby ensuring favorable long-term durability.

The most commonly performed repair technique is tricuspidization of the aortic valve. Song et al. reported successful tricuspidization in eight consecutive patients with at least moderate aortic regurgitation. Key surgical aspects included pericardial cusp reconstruction, reduction of the sinotubular junction, and commissure coaptation suturing.[13]

In our case, aortic valve replacement was selected due to severe structural abnormalities of the native valve cusps.

CONCLUSION

A quadricuspid aortic valve is an exceptionally rare congenital cardiac anomaly that is frequently identified incidentally during imaging studies or at the time of surgery. Despite its rarity, it carries important clinical implications because of its association with progressive valvular dysfunction, particularly aortic regurgitation, as well as other congenital cardiovascular abnormalities.

This case underscores the importance of maintaining a high index of suspicion in patients with valvular heart disease. It highlights the value of multimodal imaging techniques and intraoperative assessment in establishing an accurate diagnosis. Early recognition and timely surgical intervention are essential to achieve favorable clinical outcomes in patients with significant valvular dysfunction.

To the best of our knowledge, this report represents one of the few documented cases of a quadricuspid aortic valve from southern Colombia. It contributes to the limited body of literature regarding this uncommon condition.

Ethical considerations

Informed consent for treatment and open-access publication was obtained or waived from all participants in this study. Committee on Ethics, Bioethics, and Research issued approval 04-03.

Declaration

The authors declare no conflicts of interest.

Acknowledgements

The authors express their sincere appreciation to the Department of Cardiovascular Surgery, the anesthesiology team, the nursing staff, and the diagnostic imaging personnel of the Hospital Universitario Hernando Moncaleano Perdomo for their multidisciplinary support in the diagnosis, perioperative management, and postoperative care of the patient described in this report. The authors also acknowledge the institutional support provided by the Universidad Surcolombiana in promoting academic and scientific research activities. All individuals acknowledged in this section provided consent for their recognition and did not meet the criteria for authorship according to international recommendations for biomedical publications.

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