SCIENTIFIC LETTER

QUADRICUSPID AORTIC VALVE WITH SEVERE TYPE II AORTIC REGURGITATION

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ABSTRACT

Heart valve diseases are multifactorial conditions with a significant genetic and/or hereditary component. In such cases, structural alterations of the aortic valve are often observed, including a supernumerary cusp that forms a quadricuspid valve. This anomaly is closely associated with the onset and progression of aortic regurgitation, even under optimal medical management. In a small proportion of patients, the quadricuspid valve may also be associated with aortic stenosis.

A quadricuspid aortic valve is usually discovered incidentally, most often in the sixth decade of life.

We report the case of a female patient with a quadricuspid valve associated with severe type II aortic regurgitation who underwent successful surgical replacement with a biological aortic prosthesis.

Keywords: *quadricuspid aortic valve, aortic regurgitation, congenital cardiac malformation.*

Received: 04/21/2025 ***Accepted: 05/19/2025*** ***https://doi.org/10.55200/raccv.v23.n3.0111***

INTRODUCTION

Quadricuspid aortic valve (QAV) is a rare congenital and hereditary anatomical defect characterized by the presence of a supernumerary cusp. It is strongly associated with the onset and progression of aortic regurgitation, even with optimal medical management, and may occur in various positions relative to the coronary ostia.[1]

It is an uncommon anomaly, with a prevalence of less than 0.005%, according to a study conducted between 1982 and 1988 that identified only 8 cases in a cohort of 60,000 patients.[1] In 2001, a prevalence of 0.0059% was reported in a population of 357,228 participants, and in 2014, a prevalence of 0.0065% was found in a sample of 431,505 individuals. The mean age of patients with this condition is 43.5 years.[1] Cadaveric studies have reported incidences of 0.008%, and among patients undergoing aortic valve surgery, a prevalence ranging from 0.55% to 1.46% has been observed.[2]

The diagnosis of QAV is usually made as an incidental finding, generally by transthoracic echocardiography. However, when this anomaly is identified, transesophageal echocardiography may be indicated for further evaluation, as it provides a more accurate characterization of valve morphology and allows for a more detailed classification of the type of quadricuspid valve.[3,4] This condition is commonly associated with moderate to severe regurgitation, although a small group of patients may present with stenotic valves.[5] Other diagnostic methods, such as computed tomography and magnetic resonance imaging, are also used in preoperative planning.[6]

This anomaly presents in different variants depending on cusp size and position. Two central classification systems are used: Hurwitz and Roberts (*Table 1*) and Nakamura (*Table 2*).[3,6] Several studies report a higher prevalence of types A and B in the Hurwitz and Roberts' classification, which account for up to 32% of cases.[1]

Type	Description
A	Four equal cusps.
B	Three equal cusps and one smaller cusp.
C	Two larger equal cusps and two smaller equal cusps.
D	One large cusp, two intermediate cusps, and one small cusp.
E	Three equal cusps and one larger cusp.
F	Two larger equal cusps and two unequal smaller cusps.
G	Four unequal cusps.

TABLE 1. Hurwitz and Roberts' classification

Type	Description
I	Accessory cusp between the left and right coronary cusps.
II	Accessory cusp between the right coronary and non-coronary cusps.
III	Accessory cusp between the left coronary and non-coronary cusps.
IV	Indistinguishable accessory cusp due to the non-coronary cusp being divided into two equal parts.

TABLAE 2. Nakamura classification

Although the association between QAV and aortic dilatation is not strong, up to 29% of patients may present some degree of aneurysmal dilatation. Of these, 36% showed dilatation of the aortic root, another 36% of the ascending aorta, and 29% of both segments.[7]

The predominant problem in QAV is aortic regurgitation, which often requires surgery during the fifth or sixth decade of life.[8] Pure stenotic QAV is very rare, occurring in approximately 0.7% of cases.[9] Surgical intervention is indicated in severe regurgitation or stenosis, especially when left ventricular function is compromised (LVEF <50%). While surgical treatment is typically necessary in symptomatic patients, some remain asymptomatic until later stages of life, underscoring the importance of careful follow-up and individualized treatment plans.

QAV can coexist with other congenital defects, such as coronary anomalies or septal defects, complicating surgical decision-making. Reviews report associated congenital heart disease in 18-32% of patients.[10]

Although QAV usually presents as an isolated condition, in one non-surgical cohort, 19% had mitral valve prolapse; 2%, tricuspid valve prolapse; 2%, atrial septal defect (ASD); and 5%, ventricular septal defect (VSD). In the surgical cohort, 13% presented myxomatous mitral valve changes and 10% had coronary anomalies due to ostial malformations and dysplasia, more frequently with left ostial obstruction by the accessory cusp.[8,9] Reports also mention the possible presence of hypertrophic cardiomyopathy, bundle branch blocks, and complete atrioventricular blocks.[10, 11]

Decision-making regarding patient selection and surgical strategy is crucial, as not all patients with QAV necessarily require surgery.[12] Approximately one-fifth of patients ultimately undergo surgical intervention, highlighting the importance of a multidisciplinary approach.

CLINICAL CASE REPORT

A 63-year-old woman with a history of hypothyroidism, hypertension, and type 2 diabetes mellitus was followed by the cardiovascular surgery service due to progressive functional deterioration (NYHA class III/IV). An external echocardiogram of a tricuspid aortic valve showed severe insufficiency and preserved LVEF as evidenced by transthoracic echocardiography; admission was decided for scheduled open aortic valve replacement surgery.

Intraoperative transesophageal echocardiography revealed a quadricuspid aortic valve (*Figures 1* and *2*). Coronary angiography showed normal epicardial arteries. The patient underwent aortic valve replacement with a Medtronic Hancock II™ biological prosthesis No. 23 (SN: J140961) and annular enlargement, without complications.

FIGURE 1. Intraoperative transesophageal echocardiogram. Mid-esophageal short-axis view of the aortic valve showing an "x"-shaped or cross-like appearance during diastole.

FIGURE 2. Intraoperative finding: native valve with four cusps.

At her one-month postoperative follow-up, the patient showed clear improvement in symptoms and functional class, with no wound complications or signs of infection.

Surgical techniques

The optimal surgical approach for patients with QAV and severe aortic stenosis or regurgitation may involve open surgical or transcatheter techniques,

depending on the patient´s specific condition and surgical risk. Recent studies have highlighted the efficacy of transcatheter aortic valve implantation (TAVI) and surgical repair techniques as alternative options to traditional valve replacement.[7,8]

Aortic valve replacement: Traditional surgical replacement remains a common approach, particularly in patients with significant comorbidities or those unsuitable for repair or TAVI, as in the present case.[13]

Tricuspidization and annular banding: The choice of technique depends on cusp flexibility and the presence of ascending aortic dilatation.[13] This approach involves reconstruction of the aortic root, correction of severe regurgitation, and replacement of the dilated ascending aorta.

Transcatheter aortic valve implantation (TAVI): This has shown promise for patients at high risk for traditional surgery. For example, an 83-year-old patient with severe aortic stenosis underwent successful TAVI with a self-expanding valve, demonstrating significant hemodynamic improvement.[14,15] Device choice (e.g., Evolut R™ or SAPIEN 3™) is critical, as specific designs minimize paravalvular leak and adapt to the unique anatomy of QAV.[16]

DISCUSSION

First described by Balinton in 1862, the quadricuspid aortic valve is a rare congenital anomaly with an incidence of less than 0.005% in the general population.[1,4] The mean age of presentation is 45-60 years, with a higher prevalence in men.[4] According to the Hurwitz and Roberts classification, type A is the most frequent, associated with aortic regurgitation in up to 75% of cases.[17] QAV generally presents as an isolated anomaly without associated congenital defects.[3,11]

We present the case of a female patient slightly older than the mean reported age. She had a type A QAV according to Hurwitz and Roberts, associated with severe type II aortic regurgitation, consistent with published data.[16,17] No other structural congenital heart anomalies were observed at diagnosis or during surgery. Her chronic symptoms mainly reflected functional limitation, an uncommon presentation given the absence of syncope or chest pain, as frequently described in the literature.[18-20]

Determining the optimal surgical approach for QAV requires careful consideration of valve function, associated anomalies, and the patient's overall health. Decision-making is a complex process that demands a comprehensive evaluation of these factors.

Although TAVI offers a less invasive alternative with favorable outcomes for high-risk patients, surgical repair may still be preferable when anatomy allows effective reconstruction. Ultimately, the choice depends on individual patient characteristics, the surgical team's expertise, and the use of advanced imaging modalities such as transesophageal echocardiography to assess valve morphology and associated anomalies.

In conclusion, quadricuspid aortic valve disease is a hereditary condition that, according to the current literature, typically manifests between the fifth and seventh decades of life and is strongly associated with mechanisms of aortic regurgitation. However, further statistical data are required to clarify its associations with sex, comorbidities, progression of regurgitation and/or stenosis, as well as morbidity and mortality in patients treated surgically versus those managed with optimal medical therapy.

ETHICAL CONSIDERATIONS

This study complies with the principles of respect for human dignity, confidentiality, beneficence, non-maleficence, and justice. The results will be used exclusively for scientific and academic purposes, in accordance with principles of good clinical research practice.

Declarations

The authors declare no conflict of interest.

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