

## A Quadricuspid Aortic Valve: Case Report and Review of Literature

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### Abstract

Quadricuspid aortic valve is a rare congenital heart defect. The functional status is predominantly aortic regurgitation that usually remains asymptomatic and needs surgery at the fifth to sixth decade of life. Commonly it occurs in isolation, however, it may be associated with other defects in 30% of cases. About one-fifth require surgery and tricuspidalization is the preferred technique. We report a rare case where a 47 year old lady was incidentally found to have quadricuspid aortic valve. We present the case and review the related literature.

**Keywords:** *Quadricuspid Aortic Valve; Aortic Regurgitation; Tricuspidalization*

### Abbreviations

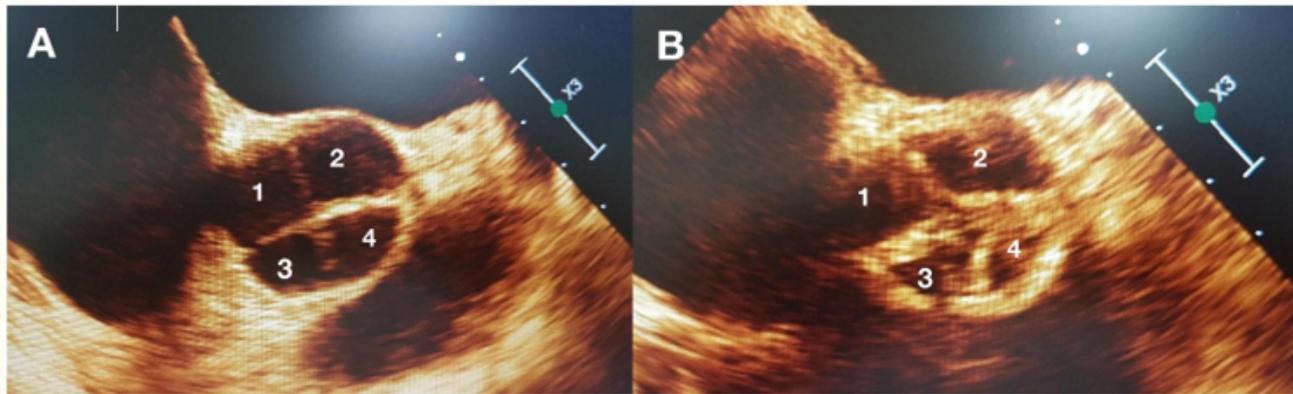
QAV: Quadricuspid Aortic Valve; TEE: Transesophageal Echocardiography; AR: Aortic Regurgitation; AS: Aortic Stenosis.

### Introduction

Quadricuspid aortic valve (QAV) indicates the presence of four valve cusps instead of the normal three. It is much rarer than its companion bicuspid aortic valve. The reported incidence is extremely low. However, there has been an increase in the reported cases since the use of echocardiography and other imaging techniques has become common. Still data is limited on its characteristics, natural history and long term outcomes. We report a case of QAV detected rather incidentally and review the related literature.

### Case Report

A 47 year old lady was referred from the medical clinic for echocardiography as she had some non-specific ECG changes. She had no known illness and was not taking any medications at home. She denied symptoms of chest pain, shortness of breath, palpitations, presyncope and syncope. Cardiovascular examination was unremarkable. BP was 104/72 mmHg and pulse 74/min, regular. She had no evidence of heart failure. ECG showed normal sinus rhythm, normal axis, nonspecific mild anterior T inversions. She had normal laboratory tests. Transthoracic echocardiography showed normal left ventricle and right ventricle. However, it showed mild aortic regurgitation with suspicion of quadricuspid aortic valve. A subsequent transesophageal echocardiogram confirmed a quadricuspid aortic valve with four symmetrical cusps (Figure 1). The patient was counselled and explained about the congenital heart defect. She is advised annual echocardiogram.



**Figure 1:** Transesophageal echocardiogram (at 350): aortic valve with 4 cusps closed in diastole (A) and the 'star' shaped opening in mid-systole (B).

## Discussion

A normal aortic valve comprises of three symmetrical cusps. However, congenital abnormalities may occur in the form of the common bicuspid valve (1-2% of population) or the much rarer quadricuspid variety, where the aortic valve has four cusps. The incidence of QAV varies depending on the method of investigation. Before echocardiography became common, QAV was mostly diagnosed either on autopsy or while undergoing aortic valve replacement. Balington [1] first described a case of QAV in 1862 when he found it incidentally during autopsy. The reported incidence is 0.00028 - 0.00033% in autopsy series, 0.0059 - 0.0065% for patients undergoing transthoracic echocardiography and 0.05-1% for those receiving aortic valve replacements for aortic regurgitation [2,3]. Although the mechanism of developing QAV is unclear, the proposed mechanisms include abnormal septation of the conotruncus, excavation of one of the valve cushions, and septation of a normal valve cushion due to inflammation [4].

The presence of varied morphology of QAV has led to different classifications, of which two are commonly used. Hurwitz and Roberts [4] have provided a comprehensive classification where QAV is divided into 7 types, A to G, based on the size of the accessory cusp. Vali, *et al.* then added type H (Figure 2). More than 85% of the cases belong to types A, B and C. Nakamura, *et al.* [5] provided another classification and divided QAV into four types that was based on the position of the accessory cusp (Figure 3).

Echocardiography is the most common modality used to diagnose QAV. Although transthoracic echocardiography can diagnose QAV, transesophageal echocardiography has become the standard investigation to confirm the diagnosis. A study by Godefroid, *et al.* [6] has reported that echocardiography most frequently diagnosed QAV (51%). Other modalities included surgery (22.6%), autopsy (15.6%) and aortography (6.5%). Not much is known about the natural history of QAV as not many cases have been reported in the literature. The anomaly is typically associated with aortic regurgitation (AR), although stenosis can develop at a later stage. Tutarel and Westhoff-Bleck [7] found that 74.7% had regurgitation, 8.4% had combined stenosis and regurgitation, 0.7% were only stenotic and the valve was normal functioning in 16.2%. Yotsumoto, *et al.* [8] studied 616 patients who underwent aortic valve surgery. They found QAV in 9 (1.46%) patients. Most of them had significant aortic regurgitation except one with combined stenosis and AR. Cusp fenestration was found in 55.6% of the AR patients. The AR usually progresses with time. Tsang, *et al.* [2] have provided some data on the follow up of QAV. During a mean follow up of  $5.5 \pm 3.7$  years, 23% of patients had progression of aortic regurgitation. Unequal shear stress may lead to leaflet fibrosis and incomplete coaptation.

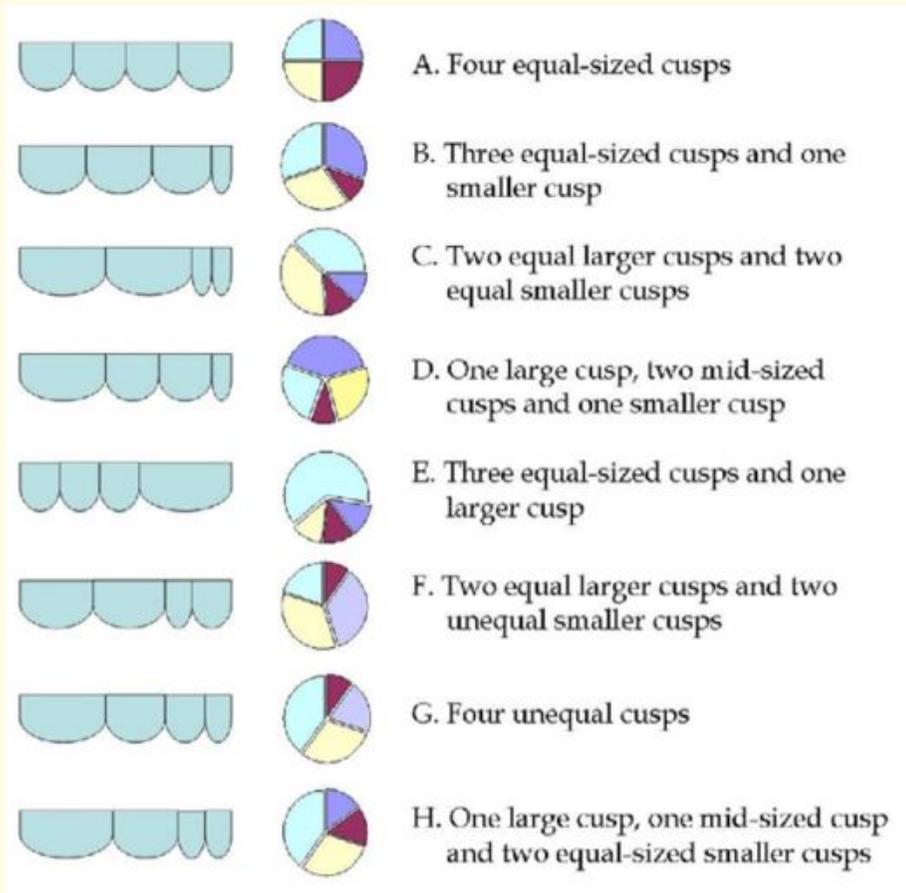


Figure 2: Hurwitz and Roberts classification of quadricuspid aortic valve.

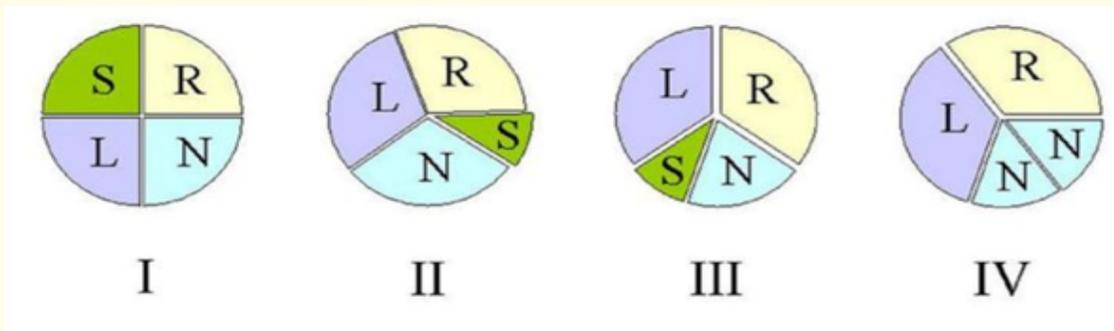
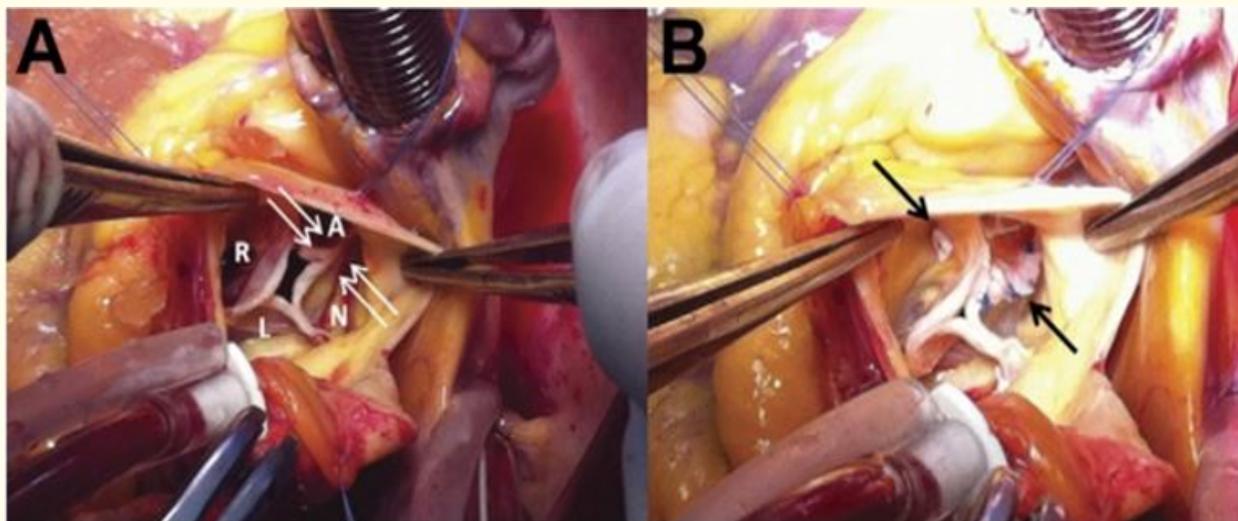


Figure 3: Nakamura et al simplified classification of quadricuspid aortic valve.

Other congenital heart defects may be associated with QAV in 18-32% of the patients. However, mostly it happens in isolation. These include coronary artery and coronary ostium anomalies, atrial septal defect, ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, sinus of Valsalva fistula, subaortic fibromuscular stenosis, mitral valve prolapses and regurgitation, hypertrophic cardiomyopathy and transposition of great arteries [2]. These patients do not have associated ascending aortic dilatation that is commonly seen with bicuspid aortic valve. QAV may be associated with infective endocarditis. Savino, *et al.* [9] detected infective endocarditis in 1.4% of the cases. The risk is lower in patients with four equally sized cusps. However, the risk of endocarditis is high in valves with unequal cusps due to uneven stress distribution and incomplete apposition during diastole [10]. The matter of endocarditis prophylaxis remains debatable. While some authors advise unconditional antibiotic prophylaxis, others recommend prophylaxis only in patients with AR with a small supranumerary cusp. Guidelines, however, have not recommended endocarditis antibiotic prophylaxis for patients with QAV [11].

Surgery offers the only definitive treatment to these patients. Indications include severe AR, severe AS or a dysfunctional QAV associated with lesions like occlusion of the left coronary ostium [2]. In the past, the procedure of choice was aortic valve replacement, especially in patients with severe valve calcification or endocarditis. Janssens, *et al.* [12] reported that 66.7% (26/39) patients with QAV with AR required aortic valve replacement. However, aortic valve repair now is the preferred technique. It avoids the risks associated with valve replacement that includes thromboembolism, bleeding and infective endocarditis. Tricuspidalization is the most commonly employed technique to repair QAV. This involves the conversion of a quadricuspid valve into a tricuspid valve by anastomosing the commissures of a coronary, or non-coronary, cusp and the supranumerary cusp, thereby restoring normal coaptation [13]. Several techniques have been described. Daprati, *et al.* [14] have described tricuspidalization of the quadricuspid valve by anastomosing the commissures of the non-coronary and the supranumerary small cusp with separated stitches with felts (Figure 4). Other options include bicuspidalization and Ross procedure. Post-operative complications are infrequent and include complete heart block, transient ischemic attack and progressive AR. Tsang, *et al.* [2] have described overall survival rates of QAV patients as 89.9% at 5-year and 84.9% at 10-year follow up.



**Figure 4:** Intraoperative view of the quadricuspid valve: (A) shows the four cusps pre-procedure (A: Accessory Cusp; L: Left Coronary Cusp; N: Non Coronary Cusp; R: Right Coronary Cusp) white arrows indicate the site of procedure; (B) shows the valve after the tricuspidalization procedure, anastomosis of the accessory and non-coronary cusps, black arrows indicate the improved leaflet coaptation. (Daprati A, Generali T, Arlati F, Roberto M. Quadricuspid aortic valve plasty: Is it worth it to repair as an alternative to substitution? *Ann Thorac Surg.* 2013;95:e7-8).

### Conclusion

Quadricuspid aortic valve (occurrence of four cusps) is an extremely rare congenital heart disease. The functional status is predominantly aortic regurgitation that usually remains asymptomatic and needs surgery at the fifth to sixth decade of life. Commonly it occurs in isolation, however, it may be associated with other defects in 30% of cases. About one-fifth require surgery and tricuspidalization is the preferred technique.

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