

Aortic regurgitation due to quadricuspid aortic valve - case report and literature review

C. Ginghină^{1,2}, D. Așchie¹, A. Călin^{1,2}, B.A. Popescu^{1,2}, C. Călin^{1,2}, F. Stoian¹, A.D. Nanu^{2,3}, L. Dorobantu¹, O. Stiru¹, C. Bulescu¹, S. Bubenek^{1,2}, V.A. Iliescu^{1,2}

¹Institute of Emergency of Cardiovascular Disease "Prof. C.C. Iliescu", Bucharest, Romania

²University of Medicine and Pharmacy "Carol Davila", Bucharest, Romania

³Colentina Clinic Hospital, Bucharest, Romania

Rezumat

Regurgitare aortică datorată unei valve aortice cvadricuspide: prezentare de caz și revederea datelor din literatură

Cvadricuspidia aortică este o variantă anatomică rară, de multe ori fiind o descoperire întâmplătoare în cursul intervențiilor chirurgicale cardiace. Prezentăm cazul unei paciente în vârstă de 59 de ani, internată în clinică pentru dispnee la eforturi mici și palpitații. Ecocardiografia transtoracică a relevat prezența unei regurgități aortice severe în condițiile unei cvadricuspidii aortice, confirmată ulterior prin examen ecografic transesofagian. Pacienta avea indicație de clasă I de înlocuire valvulară aortică, intervenția chirurgicală efectuându-se fără complicații. Tehnica chirurgicală și particularitățile cazului sunt discutate în lumina datelor existente pe această temă în literatură.

Cuvinte cheie: valvă aortică cvadricuspidă, ecocardiografia transtoracică, înlocuire valvulară aortică

Abstract

Quadricuspid aortic valve is a rare variant of aortic semilunar valve, often being an unexpected discovery during cardiac surgery. We present the case of a 59 years old patient, who was

admitted for dyspnea on light exertion and palpitations. The transthoracic echocardiography revealed severe aortic valve regurgitation due to a quadricuspid aortic valve, the result being confirmed by the transesophageal echo examination. The patient had a first class indication for aortic valve replacement and the surgical intervention was uneventful. The operative technique and case particularities are discussed in view of the literature published so far regarding this uncommon condition.

Key words: quadricuspid aortic valva, transthoracic echocardiography, aortic valve replacement

Introduction

Quadricuspid aortic valve is a rare variant of aortic semilunar valve, often being an unexpected discovery during cardiac surgery. The first case was described by Balington in 1862. (1)

Case presentation

We present the case of a 59 years old patient, who was admitted for episodes of fast irregular palpitations and dyspnea on light exertion, symptoms that appeared four months before admission. We point out the lack of any significant familial or personal history of cardiovascular diseases.

The physical exam reveals a hyperdynamic diffuse cardiac impulse, best felt in the VIth intercostal space on medioclavicular line. The heart sounds were arrhythmic and a diastolic murmur was heard at the left sternal border. The electrocardiogram showed atrial fibrillation with a rapid

Corresponding author:

Carmen Ginghina, M.D.
Institute of Emergency of Cardiovascular
Disease "Prof. C.C. Iliescu", Bucharest
Romania
E-mail: carmenghina2010@gmail.com

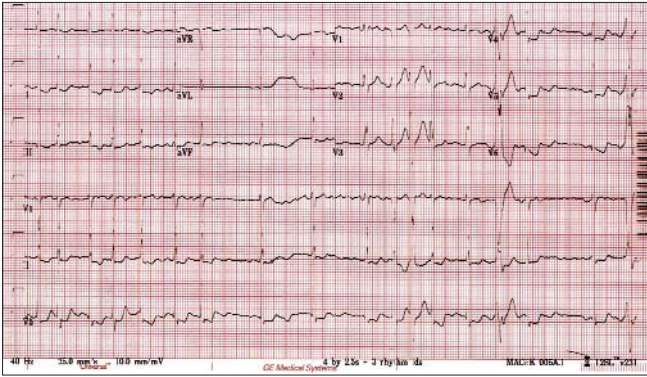


Figure 1. Electrocardiogram on admission: atrial fibrillation with a rapid ventricular response, left ventricular hypertrophy with secondary ST-T changes and premature aberrantly conducted complexes

ventricular rate of 110 beats per minute, left ventricular hypertrophy with secondary ST-T changes. (Fig. 1)

Chest X-ray revealed a cardiothoracic index greater than 0.5, enlargement of the left ventricle and a prominent aortic knob. (Fig. 2, 3)

To complete the previous data, a transthoracic echocardiography was performed, which identified severe aortic valve regurgitation due to a quadricuspid aortic valve, moderate dilatation of the left atrium, mild pulmonary hypertension (estimated systolic pulmonary artery pressure of 40 mmHg). The ejection fraction of the left ventricle was 45% and the ascending aorta had a normal size. (Fig. 4)

The transoesophageal echocardiography confirms data from the transthoracic exam, additionally revealing a patent foramen ovale with a small interatrial shunt, without a surgical indication. (Fig. 5, 6)

Based on clinical and paraclinical data, the final diagnosis is:

- Severe aortic regurgitation due to a quadricuspid aortic valve;
- Permanent atrial fibrillation;
- Mild mitral regurgitation;
- Mild pulmonary hypertension;
- Patent foramen ovale with a small interatrial shunt;
- Chronic heart failure NYHA 2 class.

This patient with severe symptomatic aortic regurgitation had a first class indication for surgical aortic valve replacement according to the European Society of Cardiology guidelines. Preoperative coronary artery angiography revealed normal epicardial arteries.

The intervention was accepted by the patient. After sternotomy, the normothermic cardiopulmonary bypass was started using an arterial canula in the distal ascending aorta and a single atrio-caval canula in the right atrial appendage. We used a single dose of antegrade cold blood cardioplegia administered in the coronary ostia. The aortic valve inspection revealed quadricuspid aortic valve, with 2 normal cusps (the right and left coronary cusps) and two smaller cusps, nonequal, with a supplementary cusp situated between the

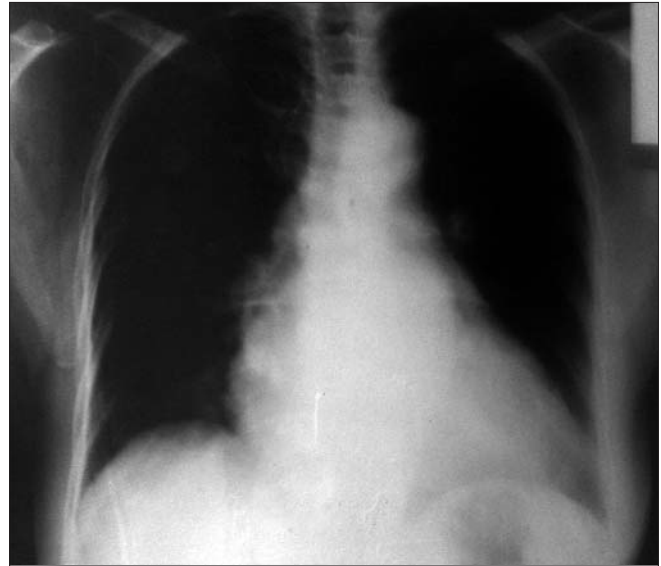


Figure 2. Posterior-anterior chest X ray reveals a cardiothoracic index greater than 0.5, lower left heart border elongation and prominent aortic knob

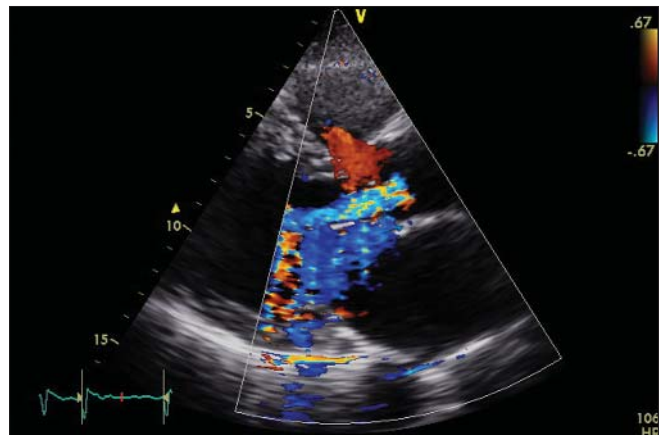


Figure 3. Transthoracic echocardiography, parasternal long axis view. Color Doppler flow showing the central jet of a significant aortic regurgitation

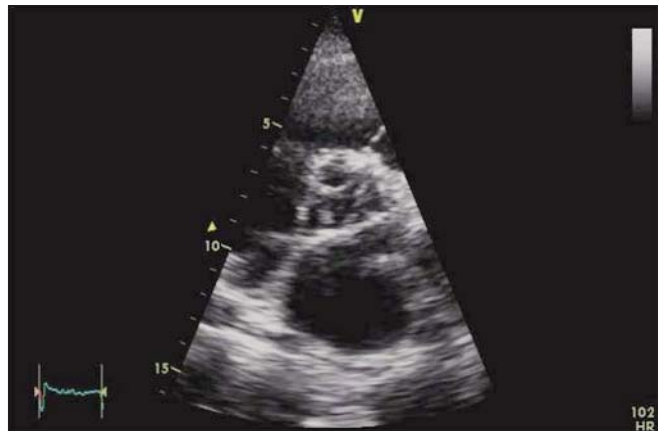


Figure 4. Transthoracic echocardiography, parasternal short axis view at the level of the great vessels, centered on the aortic valve: the four cusps can be seen in systole



Figure 5. Short axis transoesophageal view of the aortic valve in diastole, 2D and color Doppler flow, showing the incomplete closure of the quadricuspid aortic valve with a central jet of aortic regurgitation

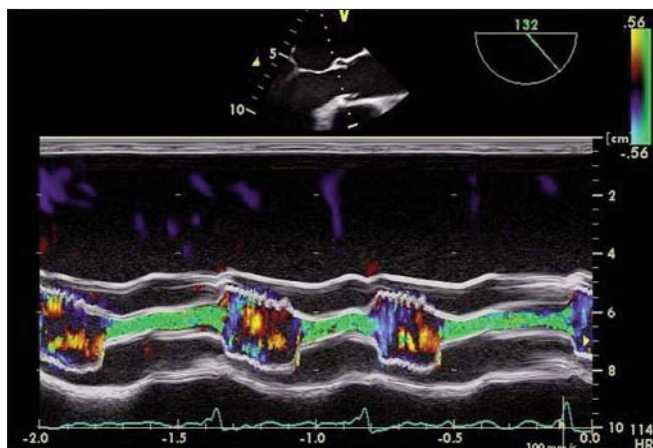


Figure 6. Transoesophageal echocardiography. Color M mode placed at the level of the aortic valve identifies the incomplete closure of the valve in diastole and a thick jet of severe holodiastolic aortic regurgitation

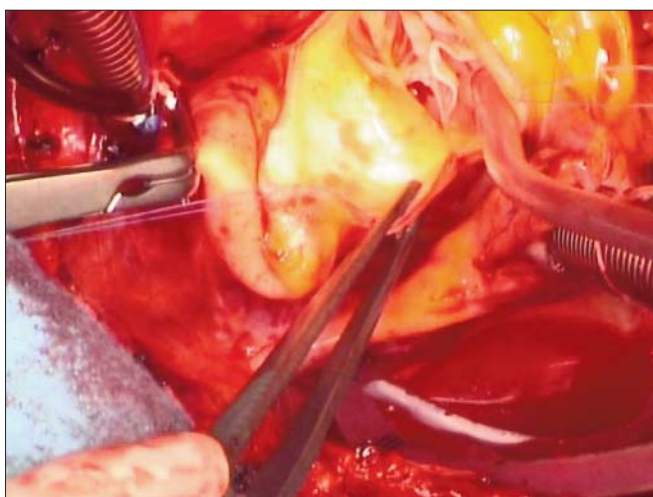


Figure 7. Intraoperative view of the quadricuspid aortic valve

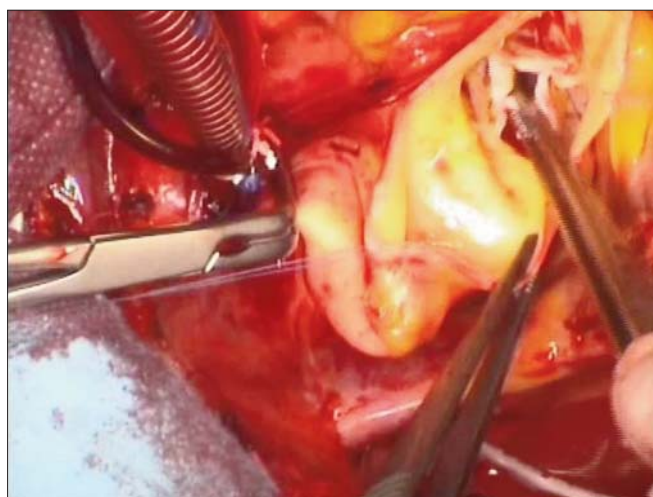


Figure 8. Intraoperative view of the quadricuspid aortic valve – the forceps indicates the supplementary cusp.

noncoronary and left coronary cusp (Fig. 8, 9, 10); this situation corresponds to type F according to Hurwitz classification.

The valve was resected (Fig. 9, 10) and replaced with an ATS no.23 mechanical valve.

Cross-clamp time was 25 minutes and the duration of the cardio-pulmonary by-pass was 41 minutes. The postoperative evolution was without any complications. The patient was discharged on the 7th postoperative day.

Case particularities

Severe aortic regurgitation due to a quadricuspid aortic valve discovered by transthoracic echocardiography

The association of a patent foramen ovale with a minimal interatrial shunt

Clinical expression of a congenital defect in a 59 yrs old patient without any personal medical history, who presented with an important hemodynamic impact



Figure 9. Quadricuspid aortic valve. The excised aortic valve presents two cusps of equal size and two smaller cusps with unequal size, which corresponds to type f in Hurwitz classification

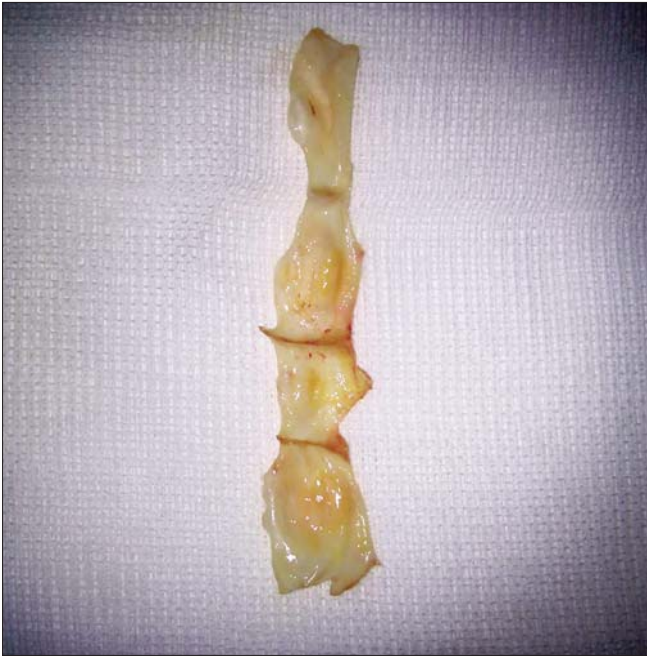


Figure 10. *Quadracuspid aortic valve with 2 cusps of equal size and 2 smaller, unequal sized cusps*

Discussion

Epidemiology

Quadracuspid aortic valve is a rare congenital heart disease with an incidence between 0.08% and 0.033%. So far, approximately 200 cases have been described. Pulmonary quadracuspid valve is nine times more frequently found than quadracuspid aortic valve. This valvular lesion has a light predominance in the male population (2).

Embriology and classification

The mechanism of this congenital malformation is not fully understood. One hypothesis is related to abnormal arterial trunk septation. Normally, after its septation, aortic and pulmonary valves develop from the three mesenchymal protrusions, so quadracuspid aortic valve represents a supranumerary variant of a primary aortic mesenchymal bud or an abnormal proliferation of the cusps (3).

Depending on the size of each aortic valve cusp, Hurwitz and Roberts (4) classified the malformation in 7 types:

- a. All four cusps of equal size;
- b. Three cusps with equal size, the fourth cusp smaller in size (most common type);
- c. Two larger cusps and two smaller cusps;
- d. One larger cusp, two cusps of intermediate size and one smaller cusp;
- e. Three cusps with equal size and one of bigger size.
- f. Two cusps of equal size and two cusps with unequal, smaller size;
- g. All four unequal size (the less frequent variant).

Functional expression

In 75% of all quadracuspid aortic valve cases aortic regurgitation is found (2). The regurgitation tends to be more frequent in cases with a smaller additional cusp and the risk is minimal for four cusps of equal size. Aortic stenosis is very rare (0.7%) (5), although in the series case described by Yatsumoto the reported prevalence is between 7-12% (6). The quadracuspid aortic valve dysfunction is minimal or absent in children and adolescents. The average age of diagnosis is 45 years. (2)

Diagnostic methods and therapeutical approach

The clinical picture is dominated by aortic regurgitation, so, the first step in the clinical diagnosis is the physical exam, which reveals a typical diastolic high pitched murmur best heard along the left lower sternal border. (7) The first investigations carried out to complete the physical exam are the electrocardiogram and the chest X ray but echocardiography is the investigation that establishes the diagnosis of quadracuspid aortic valve and evaluates the severity of aortic regurgitation. Two-dimensional echocardiography provides information about the morphology of the aortic valve (number of cusps, degree of thickening, vegetations), aortic root size, left ventricular size, and color Doppler is the most sensitive method to diagnose aortic regurgitation, detecting even a mild aortic regurgitation by identifying a turbulent diastolic jet in the outflow tract of the left ventricle. (8,9)

Sometimes, the quadracuspid aortic valve is not discovered by transthoracic echocardiography and a transoesophageal examination is necessary (10). It provides additional data regarding the morphology of the aortic valve (number of cusps, presence of prolapse) and measurement of several parameters that are useful for valve surgery (aortic annulus diameter). Zhenghua et al describes a case of a quadracuspid aortic valve, undiagnosed by transthoracic ultrasound in which the transoesophageal echocardiography identified four cusps with typical "X" configuration during systole, different from the "Y" configuration of the normal, tricuspid valve.(11)

The prevalence of quadracuspid aortic valve is too low to study the diagnostic accuracy of transthoracic versus transoesophageal echocardiography in the diagnosis of this valvular lesion. Alegret et al investigated patients with bicuspid aortic valves and 15 out of 32 bicuspid valves were missed on transthoracic, but detected on transoesophageal echocardiography. (12)

Another diagnostic method is aortography, which can describe the aortic cusps in terms of mobility, number, calcification and assess the severity of the aortic regurgitation semi-quantitatively based on visual estimation of the amount of contrast substance that appears in left ventricle after injection of the aorta. The first case of quadracuspid aortic valve diagnosed by aortography was reported by Peretz, in 1969.

Often, the quadracuspid aortic valve is an intraoperative accidental discovery, in cardiac surgery, usually for severe aortic regurgitation. Yatsumoto et al reported four cases diagnosed intraoperatively, from the series of 9 cases with quadracuspid aortic valves. (6)

Finally, this malformation can be found at necropsy.

By integrating all the data presented above, we observe that most cases of quadricuspid aortic valve are diagnosed by echocardiography (51%), followed by surgery (22.6%), necropsy (15.6%) and aortography (6,5%) (2).

From a therapeutic point of view, surgical replacement of the quadricuspid aortic valve is the most frequent option. In few published cases, "in situ" surgical repair was performed. Iglesias et al reports such a case, in which a quadricuspid aortic valve was converted into a tricuspid valve by suturing the common commissure between the right coronary cup and the supranumerary cusp (13). Also, there are a few cases that were solved by valvuloplasty, but quadricuspid aortic valve is less suitable than tricuspid or bicuspid aortic valve for this procedure, probably due to the presence of a longer coaptation line (6). On the other hand, almost all the published cases reporting aortic valvuloplasty involved the B type of quadricuspid aortic valve and not the F type; also, there are no published long term results regarding this type of operation (13); thus, aortic valve replacement was the correct decision in this case.

The risk of infective endocarditis

Some cases of infective endocarditis associated with a quadricuspid aortic valve have been reported, although it is so far unclear whether quadricuspid aortic valve is more prone to infectious risk. Bacterial endocarditis prophylaxis is necessary in cases associated with a supranumerary smaller cusp, due to abnormal hemodynamic stress distribution and abnormal coaptation. In patients with four equally sized cusps the risk of bacterial endocarditis is lower. However, Matsukawa et al presented a case of bacterial endocarditis in a 75 years old patient, diagnosed with quadricuspid aortic valve with equally sized cusps (14).

Associated lesions. Quadricuspid aortic valve may be associated with other congenital lesions, such as atrial or ventricular septal defect, aneurysmal dilation of a Valsava sinus, Ehler-Danlos syndrome, hypertrophic cardiomyopathy, dilatation of ascending aorta, pulmonary stenosis but most frequently, it is associated with coronary arteries anomalies (15).

An interesting case of an adolescent who was diagnosed with rheumatic carditis involving the aortic, mitral and tricuspid valves was published, in whom the necropsy revealed the coexistence of quadricuspid aortic and pulmonary valves. (16)

Conclusions

Quadricuspid aortic valve is a rare congenital anomaly, associated in this case with severe aortic regurgitation.

Diagnosing this condition is difficult most of the times, being performed in the intraoperative setting in the majority of cases. In this particular case, both the transthoracic and the transoesophageal echocardiograms were the correct diagnostic tools.

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