



# Quadricuspid aortic valve: A rare etiology of aortic regurgitation

O. Godefroid<sup>a</sup>, P. Colles<sup>b</sup>, S. Vercauteren<sup>a</sup>, Y. Louagie<sup>c</sup>,  
B. Marchandise<sup>a,\*</sup>

<sup>a</sup> Cardiology Unit, Mont-Godinne University Hospital, Avenue G. Therasse 1, B-5530 Yvoir, Belgium

<sup>b</sup> Centre de Santé des Fagnes, Boulevard Louise 18, B-6460 Chimay, Belgium

<sup>c</sup> Cardiovascular Surgery Unit, Mont-Godinne University Hospital, Avenue G. Therasse 1, B-5530 Yvoir, Belgium

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## KEYWORDS

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**Abstract** Quadricuspid aortic valve is a rare cause of aortic insufficiency. We report two unusual cases of this valvular pathology associated with a dilatation of the aortic root. The mechanism leading to this valve incompetence is incompletely understood and is discussed in regard to these cases.

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## Introduction

We report two unusual cases of quadricuspid aortic valve revealed by the presence of a diastolic murmur of aortic valvular regurgitation.

In regard to other congenital malformations of the aortic valve, the incidence and the pathophysiology of this aortic valve abnormality are discussed.

## Case report 1

A 51-year-old man was referred with a six-months history of dyspnea (NYHA class II), he had no history of endocarditis or rheumatic disease and presented no coronary risk factor.

On physical examination, his blood pressure was 150/50 mmHg and cardiac auscultation revealed a 3/6 diastolic murmur along the left sternal border.

Twelve-leads electrocardiogram showed a sinus rhythm, an incomplete right bundle branch block and signs of left ventricular hypertrophy.

Transthoracic echocardiography confirmed left ventricular hypertrophy (LV mass index: 120 g/m<sup>2</sup>) with an enlarged cavity (left ventricular end

\* Corresponding author. Tel.: +32 81 42 36 00; fax: +32 81 42 36 15.

E-mail address: [marchandise@card.ucl.ac.be](mailto:marchandise@card.ucl.ac.be) (B. Marchandise).

diastolic dimension: 73 mm), a normal left ventricular ejection fraction (59%) and a grade 3 aortic regurgitation; an abnormality of the aortic valve was suspected. Transoesophageal echocardiography showed, in parasternal short axis view of the great vessels, a quadricuspid aortic valve with the presence of four thin cusps of equal size with a "X-shaped" commissural aspect in diastole (Fig. 1). There was a severe (grade 3) central aortic regurgitation, resulting from an incomplete diastolic coaptation of the cusps. The aortic root was enlarged (diameter: 46 mm).

Coronary angiography was normal, except for an abnormal cranial displacement of the ostium of the right coronary artery, from the right anterior coronary sinus.

Aortography confirmed a grade 3 valvar aortic regurgitation. The left ventricular end diastolic pressure was 23 mmHg in basal conditions.

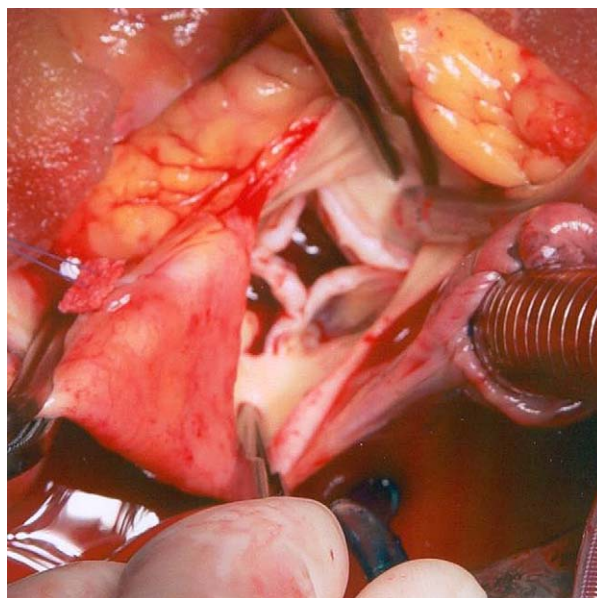
The patient underwent elective aortic valve replacement with a mechanical prosthesis (Carbomedics number 25); at surgery, the valve was noted to be quadricuspid (Fig. 2) and the abnormal take-off of the right coronary artery was confirmed. Moreover, a small surnumary coronary artery was also present near the ostium of the main trunk. The diameter of the ascending aorta was reduced at the closure of the aortotomy (aortoplasty). The patient was reviewed 6 months later and was asymptomatic.

### Case report 2

A 70-year-old man with a long history of atrial fibrillation is referred for assessment of cardiac



**Figure 1** Short axis transoesophageal view of the aortic valve.



**Figure 2** Per-operative view of the aortic valve.

murmur .The patient is asymptomatic except for palpitations.

The physical examination shows a blood pressure of 130/80 mmHg, an irregular cardiac rhythm; at the cardiac auscultation, a 3/6 diastolic murmur is heard along the left sternal border.

The ECG shows an atrial fibrillation and a complete right bundle branch block.

A transthoracic echocardiogram shows an enlarged left ventricle (left ventricular end diastolic dimension: 60 mm), a reduced left ventricular ejection fraction (40%) and a grade 3 aortic regurgitation. The aortic root is enlarged (41 mm) and the aortic valve appears abnormal.

Transoesophageal echocardiogram confirms the severity of aortic regurgitation which is central (failure of the coaptation) and shows four thin aortic cups with a cruciate shape in diastole. The aortogram confirms a grade 3 aortic regurgitation, an enlarged aortic root and the coronary angiogram is normal showing no abnormality in the take-off of the coronary arteries.

The patient underwent elective aortic valve replacement with a mechanical prosthesis (Carbomedics number 25), associated with a Maze operation. At the surgery, the quadricuspid morphology of the aortic valve was confirmed with a very thin and symmetrical aspect of the cusps. An aortoplasty was realized to reduce the enlarged ascending aorta.

### Discussion

The quadricuspid aortic valve is a rare manifestation of congenital aortic valve abnormalities.

Necropsy series have shown an incidence of only two cases in 6000 autopsies<sup>1</sup> but the Mayo Clinic has noted an incidence of 1% in a review of patients undergoing surgery for pure aortic regurgitation.<sup>2</sup> The diagnosis is commonly made between the 5th and the 6th decade and is more frequent in male patients.<sup>3</sup>

The anatomy of the quadricuspid aortic valve is variable, according to the size of each individual aortic valve cusp: 4 equal cusps, 3 equal cusps with one minor and 2 large cusps with 2 small cusps represent the more frequent anatomic variations.<sup>4</sup> Different embryologic mechanisms have been suggested including excavation of one of the valve cushions and septation of a normal valve cushion as a result of inflammatory episode.<sup>4</sup>

The functional aspect of the quadricuspid valve is mainly represented by pure insufficiency.<sup>4</sup>

The physiopathology of the valve dysfunction is poorly understood: anatomical abnormalities of the cusps could induce unequal shear stress leading to fibrosis and incomplete coaptation.<sup>5</sup> However, insufficiency is also observed in cases of quadricuspid valve with four equal cusps.<sup>5</sup> As in bicuspid valve, regurgitation of an abnormal aortic valve may also occur as a result of a prolapse of the layer of the cusps or be associated with aortic root dilatation.<sup>6</sup>

Aortic root dilatation in patients with quadricuspid aortic valve was reported in only one case.<sup>7</sup> Interestingly, in our two patients, severe aortic regurgitation was associated with aortic root dilatation and a central defect of coaptation of the

cusps. Regurgitation of an abnormal aortic valve may induce diffuse dilatation of the ascending aorta. The reverse – aortic regurgitation secondary to aortic root dilatation – may also occur as a result from disruption or dissolution of elastic tissue within the aortic ring, as this structure provides the main support for the valve cusps.<sup>6</sup>

*In conclusion*, quadricuspid aortic valve is a rare congenital malformation which can be diagnosed by two-dimensional echocardiography mainly transoesophageal, and which can frequently be associated with aortic regurgitation.

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