# Quadricuspid aortic valve: a rare cause of aortic insufficiency diagnosed by Doppler echocardiography. Report of two cases and review of the literature

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**Quadricuspid aortic valve is an uncommon congenital valve disease mostly occurring as isolated lesion or sometimes in association with truncal anomalies. Approximately 50% of patients with quadricuspid aortic valve have aortic regurgitation. Before the advent of echocardiography most cases were diagnosed at the time of surgery or at *post-mortem* examination. We describe 2 cases of patients with quadricuspid aortic valve diagnosed by echocardiography. The first case, a quadricuspid aortic valve with four equal-sized cusps (type A, according to the classification of Hurwitz and Roberts), was identified in a 26-year-old man undergoing echocardiography because of a heart murmur. The second case, a quadricuspid aortic valve with three relatively equal cusps and one smaller cusp (type B, according to the classification of Hurwitz and Roberts), was identified in a 47-year-old man with a history of murmur. The identification and periodical non-invasive evaluation of a quadricuspid aortic valve is important, because such valves are more vulnerable to infection and need adequate prophylaxis against endocarditis.**

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## Introduction

Quadricuspid aortic valve is a rare cardiovascular anomaly and an uncommon cause of significant aortic regurgitation. The reported incidence of quadricuspid aortic valve in the literature is variable. A total of 186 cases have been reported. The mean age of patients was 50.7 years, and there was a slight male predominance (male:female ratio 1.61:1)[1].

Most cases have been identified as an incidental finding at autopsy[2-4] with a prevalence between 0.008% (2 in 25 666 necropsies)[2] and 0.033% (2 in 6000 necropsies)[3], or at aortic valve replacement with a prevalence between 0.55% (2 in 363 aortic valve procedures)[5] and 1.46% (9 in 616 patients)[6]. Fewer cases have been detected by aortic root angiography for the assessment of valvular regurgitation[7,8] or by echocardiography[9,10]. The first diagnosis by two-dimensional echocardiography was reported by Herman et al.[9] in 1984.

We report 2 cases of quadricuspid aortic valve diagnosed by Doppler echocardiography.

## Description of cases

**Case 1.** A 26-year-old man presented with a history of murmur (New York Heart Association functional class I). No past history of rheumatic disease or endocarditis could be found. Pulses were symmetrical and of normal contour, blood pressure was 120/80 mmHg, and heart rate was 65 b/min. A systolic ejection murmur grade II/VI could be heard along the left sternal board and a diastolic murmur grade I-II/IV in the aortic valve area was present. The ECG and chest X-ray were normal.

Two-dimensional echocardiography in parasternal short-axis view of the great arteries showed a quadricuspid aortic valve with a cross shape in diastole (the so-called "X" configuration). The four cusps were of normal texture and of approximately equal size (Fig. 1). The parasternal long-axis view did not show any eccentricity of the valve in diastole and its opening was complete in systole. In short-axis view, a squared image of the open four cusps could be detected (Fig. 2). Mild aortic regurgitation was detected by continuous Doppler

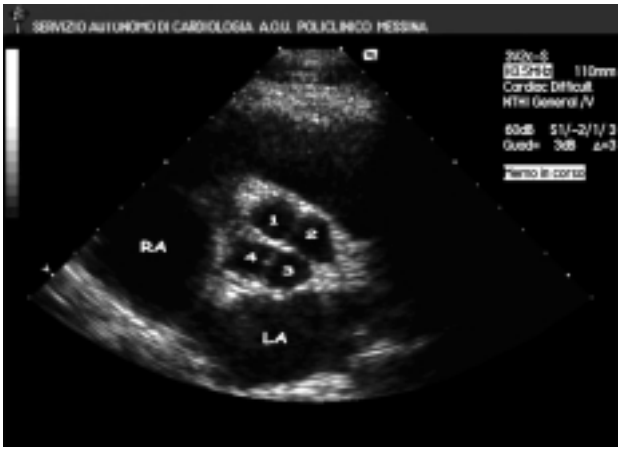


**Figure 1.** *Parasternal short-axis view of the quadricuspid aortic valve in diastole showing four equal-sized cusps with the so-called "X" configuration. LA = left atrium; RA = right atrium.*

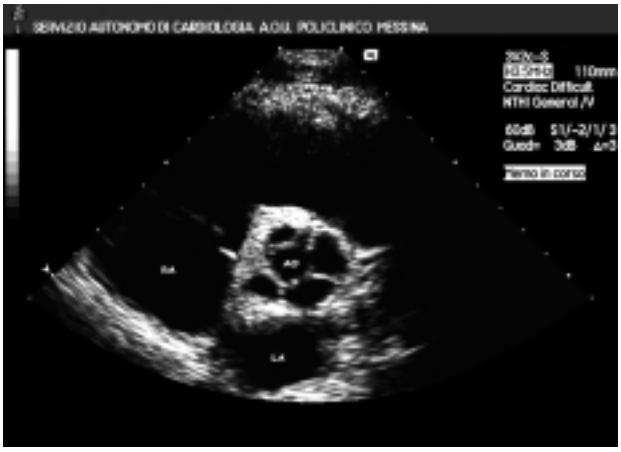

**Figure 2.** *Parasternal short-axis view showing a squared image of the open four cusps in systole. LA = left atrium; AO = aorta; RA = right atrium.*

echocardiography. Left ventricular dimensions and systolic function were normal.

**Case 2.** A 47-year-old man presented with a history of murmur (New York Heart Association functional class II). No past history of rheumatic fever or endocarditis could be found. His medical history was remarkable for hypertension. Physical examination revealed the presence of a blood pressure of 140/80 mmHg, and pulse rate of 80 b/min. A grade III/VI systolic ejection murmur could be heard along the left sternal board and a diastolic murmur grade II/IV in the aortic valve area was present. ECG revealed normal sinus rhythm, left ventricular hypertrophy with a systolic overload pattern. Chest X-ray was unremarkable.

Two-dimensional echocardiography revealed a quadricuspid aortic valve with three relatively equal cusps and one smaller cusp in the parasternal short-axis view of the great arteries (Fig. 3). Left atrial enlargement, left ventricular hypertrophy with normal left ventricular function was present, and no other cardiac defect was found. In the parasternal long-axis view the opening of the valve was complete in systole. The short-axis view showed a squared image of the opened four cusps in systole. Mild aortic regurgitation and moderate mitral regurgitation were detected by Doppler echocardiography.

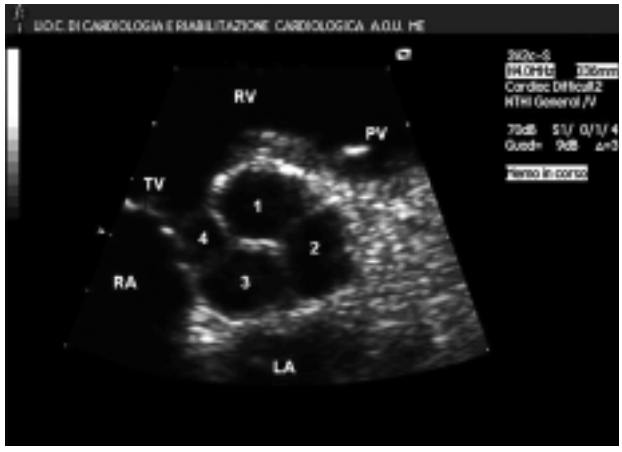


**Figure 3.** *Parasternal short-axis view of the quadricuspid aortic valve in diastole showing four unequal-sized cusps. LA = left atrium; PV = pulmonary valve; RA = right atrium; RV = right ventricle; TV = tricuspid valve.*

## Discussion

The semilunar valves derive from mesenchymal swellings in the aortic and pulmonary trunks following partitioning of the truncus arteriosus. Normally, three of such swellings eventually develop into three semilunar cusps that coapt in the center of the valve orifice. It is thought that a supernumerary aortic cusp may derive during the early stages of truncal septation from either an abnormal aortopulmonary septation or an asymmetry in the number of primordial valve cusps or abnormal proliferation or fusion of the mesenchymal buds[4]. Animal experiments have suggested that a quadricuspid valve occurs in the early stages of valvulogenesis and results from the division of one of the three mesenchymal swellings that originate a normal aortic valve[11].

When not associated with truncal anomalies, quadricuspid aortic valve is usually an isolated lesion. Some associated cardiac defects have been reported; the most frequent are anomalies of the coronary ostium and coronary arteries[10,12-15]. Ventricular[3,16] and atrial septal defects[17], patent ductus arteriosus[2,3], pulmonary valve stenosis[18], subaortic fibromuscular stenosis[19], mitral valve abnormalities[16] and transposition of the great arteries[20] have also been reported.

Hurwitz and Roberts[3] classified the quadricuspid semilunar valves according to the relative size of the four cusps. Seven types were found; the types most frequently identified were type B (three equal cusps and one smaller cusp) and type A (four cusps of similar size) (Fig. 4).

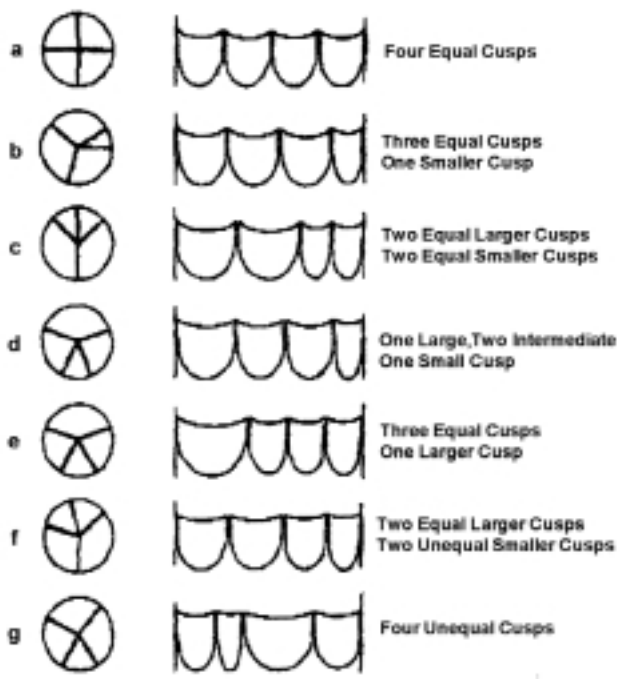


**Figure 4.** *Hurwitz and Roberts classification of the quadricuspid semilunar valves modified by Recupero et al. according to the relative size of the four cups. Seven types were found.*

The first case presented had a type A quadricuspid aortic valve, and the second had a type B quadricuspid aortic valve.

Two-dimensional transthoracic echocardiography has become the diagnostic test of choice, because the four cusps and their relative sizes can be easily assessed[10,21], as shown in the present cases. However, because of technical factors such as suboptimal echocardiographic windows or extensive aortic calcification, transthoracic echocardiography may occasionally be limited in visualizing the aortic valve. Transesophageal echocardiography overcomes many of the imaging limitations of transthoracic echocardiography and can clearly delineate the aortic valve morphology[22].

Quadricuspid aortic valve is often incompetent; normally functioning valves have been found in only 16% of cases[1]. Mechanisms of aortic regurgitation in quadricuspid valves have been proposed. Surgical and histologic findings have shown that fibrous thickening of valves, due to uneven distribution of mechanical stress, results in incomplete coaptation of the cusps, in aortic regurgitation and more vulnerability to infection resulting in endocarditis[10,23,24]. Fenestrations can also be a cause[8]. The findings reported in the literature suggest that the occurrence of aortic regurgitation is not necessarily associated with the type of quadricuspid aortic valve and Doppler echocardiography is a sensitive technique that allows its detection even when it is not clinically significant[21]. For these reasons the patients with quadricuspid aortic valve need periodical non-invasive evaluation and adequate prophylaxis against endocarditis[25]. In patients who require surgical treatment for aortic incompetence presenting with quadricuspid aortic valve, preoperative echocardiography should focus on the location and possible displacement of the coronary ostia, in order to avoid ostial obstruction when fixing the prosthetic ring at the level of the supernumerary commissure[26].

In conclusion, quadricuspid aortic valve is a rare congenital malformation that can be easily diagnosed by two-dimensional echocardiography because of the typical aspect of the four cusps in both diastole and systole. Mild aortic regurgitation is frequently associated with quadricuspid aortic valve; Doppler echocardiography is a sensitive method to detect it.

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