Quadricuspid Aortic Valve: Echocardiographic Diagnosis

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Resumo

This article is a case report of a quadricuspid (or quadrivalve) aortic valve, type A of Hurwitz and Roberts, with mild insufficiency and dilatation of the ascending aorta, in an asymptomatic adult diagnosed during a routine echocardiography exam, confirmed by a transesophageal echocardiography. The article, with images, is followed by a brief literature revision. A quadricuspid aortic valve (QAV) is rare, and the diagnosis occur by an echocardiography exam, usually in an asymptomatic group. The importance of diagnosing this pathology is justified by the risk of progressive aortic valve disease, by the potential increase of endocarditis and possible association with anomalous origin of the coronary ostium or other associated malformations.

Keywords: **Aortic Valve/abnormalities; Echocardiography Transesophageal; Heart defects Congenital.**

Case Report

Female patient, 37 years old, asymptomatic, went to the echocardiography laboratory for her first routine examination. On transthoracic examination, it was identified mild dilatation of the ascending aorta (4.0 cm), in addition to mild aortic regurgitation, and probable anatomical abnormality of the aortic valve, on parasternal transverse view (Figures 1 and 2). It was suggested complementing the study with transesophageal examination, subsequently requested by the cardiologist.

On transesophageal echocardiographic examination, changes were confirmed, revealing the typical image of the quadricuspid aortic valve, described by Herman in 1984. In the parasternal cross-sectional view, in diastole, the typical valve closure in cross or X (Figure 3), unlike the closure in Y characteristic of normal tricuspid aortic valve. In systole, there is a rectangular opening, unlike the triangular opening of the normal valve with four equal valves; type A Hurwitz and Roberts (Figure 4). It was checked a mild dilatation of the ascending aorta and mild regurgitation (Figure 5).

Review of Literature

The quadricuspid or quadrivalvular aortic valve (QAV) is a rare congenital malformation, with an incidence between 0.008 and 0.043%[1,2]. The quadricuspid pulmonary valve has an incidence nine times higher than in aortic valve and generally course without dysfunction[3-5]. The QAV affects more male individuals at a rate of 1.6:1[1,2]. The dysfunction of the majority of patients is valvular regurgitation, followed by double dysfunction (with regurgitation greater than stenosis), and a minority of pure stenosis. The valve is normofunctioning in a few patients[1,2]. The real embryological change that originates the QAV[2] remains unknown.

Earlier, the diagnosis was performed by necropsy; later in aortic valve surgery, and currently by transthoracic echocardiography, particularly by transesophageal echocardiography[2]. Three-dimensional echocardiography has no additional information for the diagnosis of quadricuspid aortic valve[6].

The classic image of the quadricuspid aortic valve on echocardiography was described by Herman in 1984 and became pathognomonic. It is obtained in parasternal cross-sectional view, where during diastole shows the typical valve closure in cross or X, unlike the closure in Y characteristic of the normal tricuspid aortic valve, and in systole the rectangular opening, unlike the typical triangular opening of the normal valve[7].

There are two morphological classifications for QAV: The first (most frequent) is the Hurwitz and Roberts which classifies the valve with respect to accessory valve size: Type A: 4 equal valves; Type B: 3 equal valves and 1 small; Type C: 2 equal large valves and 2 equal small valves; Type D: 1 large valve, 2 intermediate, and 1 small; Type E: 3 equal valves and 1 large valve; Type F: 2 large equal valves and 2 unequal small valves; Type G: 4 unequal valves. The most common types were A and B[5].

The second is from Nakamura et al.[8], based on the position of the supernumerary non-dominant valve: I - Accessory valve between the left coronary and the right coronary valve; II - Accessory valve between the right coronary and

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Case Report

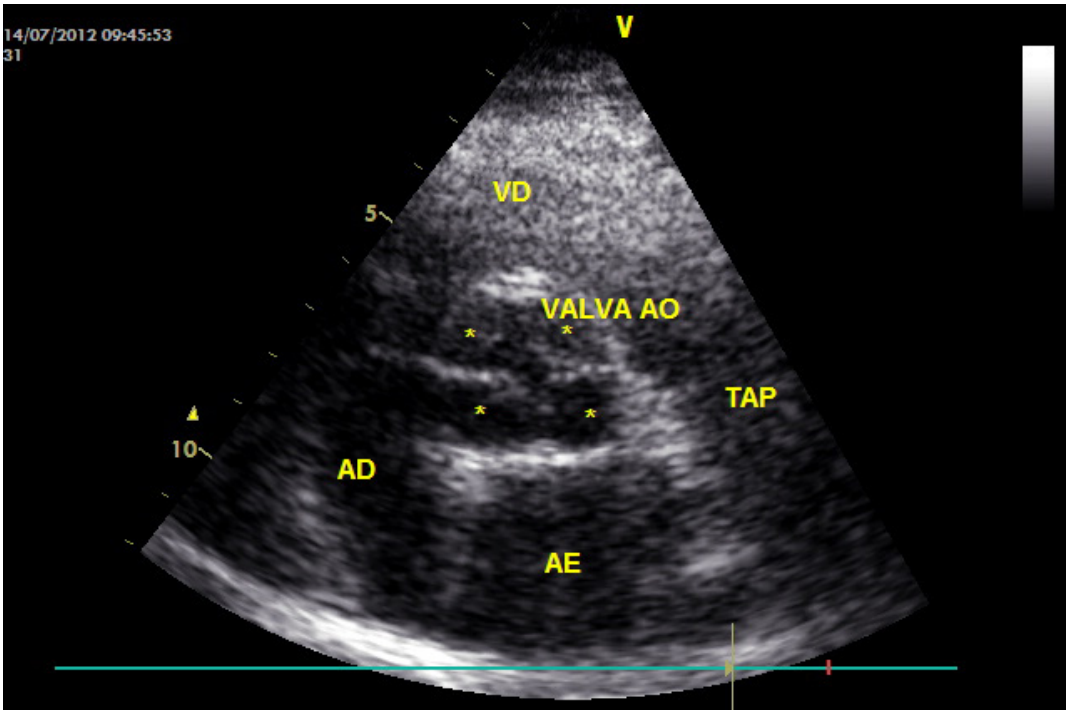

Figure 1 - *Quadricuspid aortic valve - cross-sectional parasternal view on transthoracic echocardiography.*

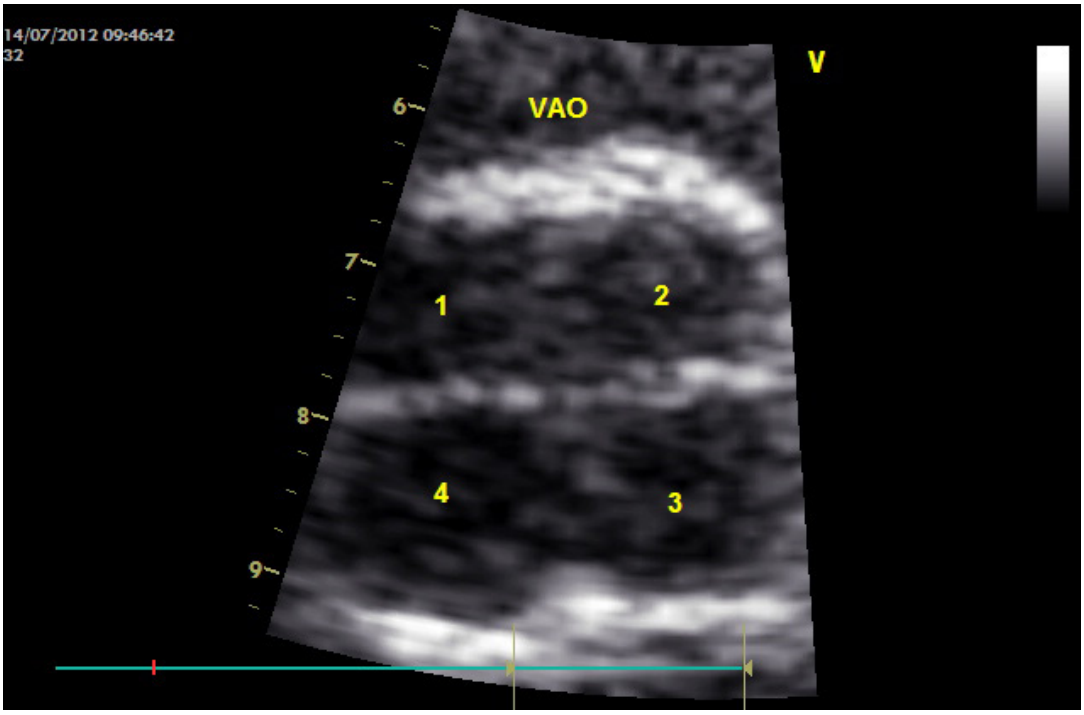

Figure 2 - *Quadrivalvular aortic valve - Zoom of the aortic valve from the image of Figure 1.*

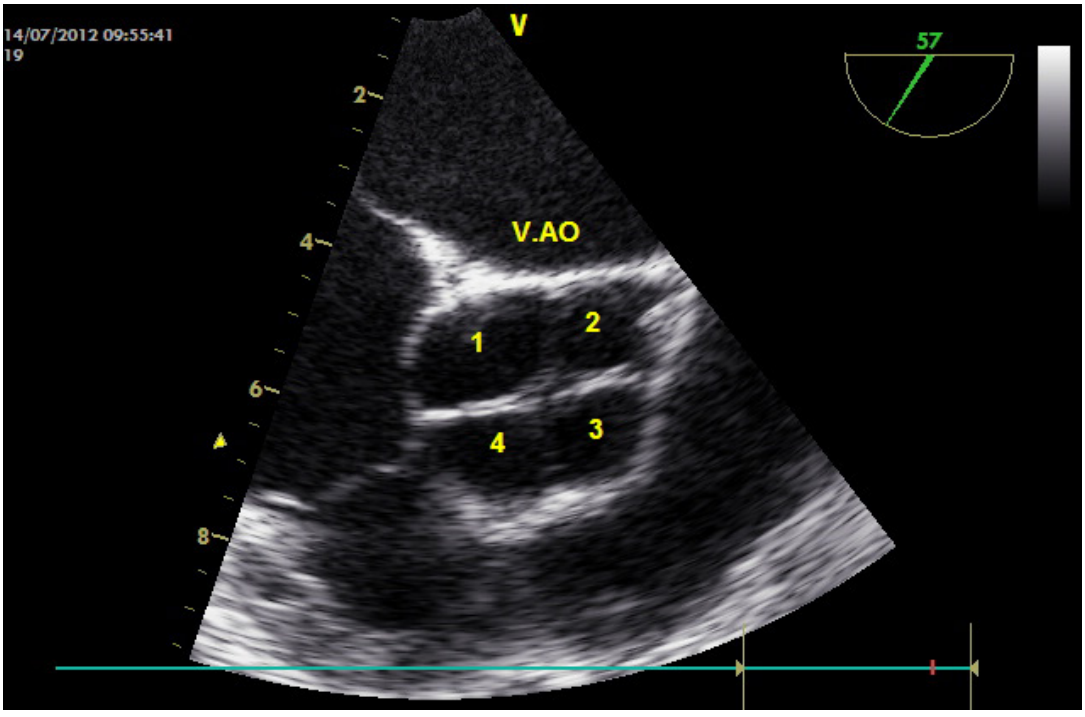

Figure 3 - *Imaging of quadricuspid aortic valve by transesophageal echocardiography - valve closure in cross or X.*

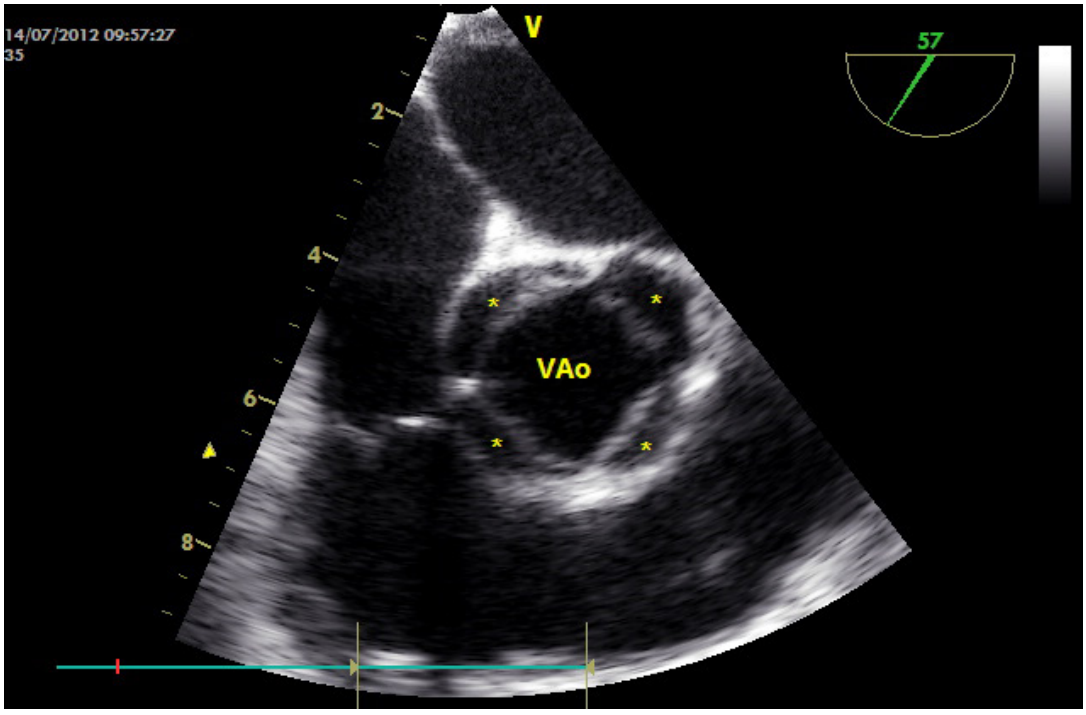

Figure 4 - *In systole, the rectangular opening of the quadricuspid aortic valve, type A Hurwitz and Roberts, seen by transesophageal echocardiography.*

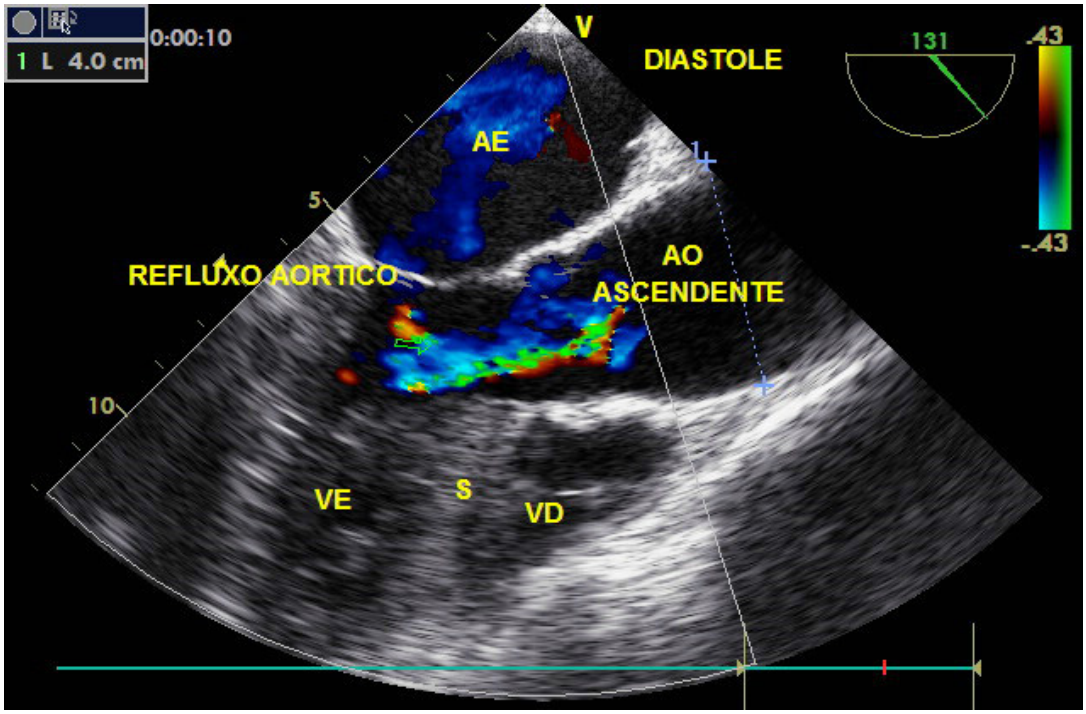

Figure 5 - *Mild aortic regurgitation and mild dilatation of the ascending aorta, demonstrated by transesophageal echocardiography.*

non-coronary valve; III - Accessory valve between the left coronary and non-coronary valve; IV - Accessory aortic valve indistinguishable because the non-coronary valve is divided into 2 equal parts[8].

The development of stenosis or regurgitation in QAV occurs by valvular thickening and calcification[9] or by dilatation of the ascending aorta[2], or more rarely by valve prolapse[10].

The QAV may be associated with other cardiac malformations in a minority of cases. The most frequent association is with anomalous origin of the coronary artery ostium (in 10% of cases of QAV described in the literature). Major anomalies described in the coronaries were single coronary ostium and anomalous position of the coronary ostium[1,3,11-13].

In most cases, the VAQ carrier is asymptomatic and diagnosis occurs in routine checkups[14]. The literature report a case of postmortem diagnosis in a young man of 16 years who died suddenly[14] and another of infarction in a child of 10 years by acute occlusion of the left coronary ostium by tissue adhered from the leaflet[12]. Thus, it is advisable to search for ischemia by non-invasive methods in children with QAV[14].

Some authors agree that the risk of endocarditis is higher in patients with QAV, especially those with unequal leaflets, and there may be benefits in the prophylaxis for procedures with bacteremia[1,9,14-16].

Surgical treatment (usually valve replacement) is indicated in symptomatic patients and recommended for asymptomatic patients with dysfunction or left ventricular dilatation (diastolic diameter > 75 mm and systolic diameter > 55 mm) and which did not progress to cardiac decompensation[17].

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