Case Series of Patients with Echocardiographic Diagnosis of Quadricuspid Aortic Valve

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Introduction

Quadricuspid aortic valve (QAV) is a rare congenital malformation. Most patients are asymptomatic and have valves without any functional disorders. The presentation of this pathology generally occurs without any clinical manifestation, with an incidence ranging from 0.008 to 0.043%. It usually occurs in males at a ratio of 1.6:1. It may be found occasionally in asymptomatic patients, but it is often associated with aortic regurgitation. Echocardiography is the ideal method for determining and monitoring this anomaly and its repercussions.

Clinical cases

Case 1

Male patient, 77 years old, hypertensive and smoker. Admitted to the coronary intensive care unit (ICU) of Hospital de Base do Distrito Federal, with ST-segment elevation acute myocardial infarction. Echocardiography evidenced the presence of QAV with discrete sclerosis and regurgitation and segmental abnormality. Coronary artery ostia could be seen (Figure 1).

Case 2

Female patient, 40 years old, referring fleeting paroxysmal palpitations. The patient did not refer any cardiovascular symptoms. On a routine outpatient echocardiography scan, QAV revealed mild regurgitation with no other abnormalities (Figure 2).

Case 3

Female patient, 44 years old, under murmur investigation to be clarified in a preoperative cholecystectomy appointment. The patient did not refer any cardiovascular symptoms and diseases. A routine outpatient echocardiography scan revealed the presence of QAV with turbulent flow, with peak gradient of 20 mmHg and mild aortic regurgitation with no other abnormalities (Figure 3).

Keywords

Aortic Valve Insufficiency/physiopathology; Congenital Abnormalities; Aortic Valve Insufficiency/surgery; Echocardiography/methods; Stroke Volume.

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Discussion

Diagnosis of QAV is rare and is most often delivered during autopsy or as an incidental intraoperative finding. Studies estimate that its incidence is between 0.008%[1] and 0.33%.[2] The latest revision, based on echocardiography scans, evaluates an incidence of 0.043%.[3] When it comes to patients undergoing valve replacement surgery, the incidence increases and ranges from 0.55% to 1.46%.[4]

Considering the pediatric age group, this diagnosis becomes even rarer, since most patients are asymptomatic and have valves without any functional disorders. In the literature, male predominance is observed at a ratio of 1.6:1. This diagnosis is more common after the fifth decade of life, when some individuals may manifest symptoms, predominantly of aortic regurgitation.

The embryological origin of QAV remains unknown. It is known that both semilunar valves derive from the mesenchyme folds, which form in the roots of the aortic and pulmonary trunks, after the common trunk has been split, usually arising three protrusions that develop into the vascular lumen between the fifth and the ninth week of gestation. Several pathophysiological mechanisms have been suggested to lead to changes in the number of valvular leaflets, namely anomalous septation of the conotruncal junction, with asymmetries in the distribution of the number of protrusions in each of the large vessels; abnormal proliferation of one or more mesenchymal folds; or splitting of one of the valvular protoleaflets during their formation.

The main congenital malformation of the aortic valve is the aortic valve with only two leaflets, with an incidence of 2%, being the most common congenital aortic valve anomaly and one of the most common cardiac malformations. The second most frequent one is the aortic valve with one leaflet, more commonly diagnosed in childhood, being related to the valve stenosis.

There are two QAV classifications. Hurwitz and Roberts[2] developed a classification system according to the size of the valve leaflets, dividing them into eight types, from A to H (Figure 4). Nakamura simplified and divided them according to the position of the accessory cusp[5] (Figure 5). According to an investigation conducted by Hurwitz and Roberts[2], about 85% of the cases belong to types A, B or C.

Initially, the diagnosis of this anomaly was made by necropsy; then, during aortic valve surgeries. These days, much greater accuracy has been achieved through transthoracic echocardiography and, mainly, transesophageal echocardiography.

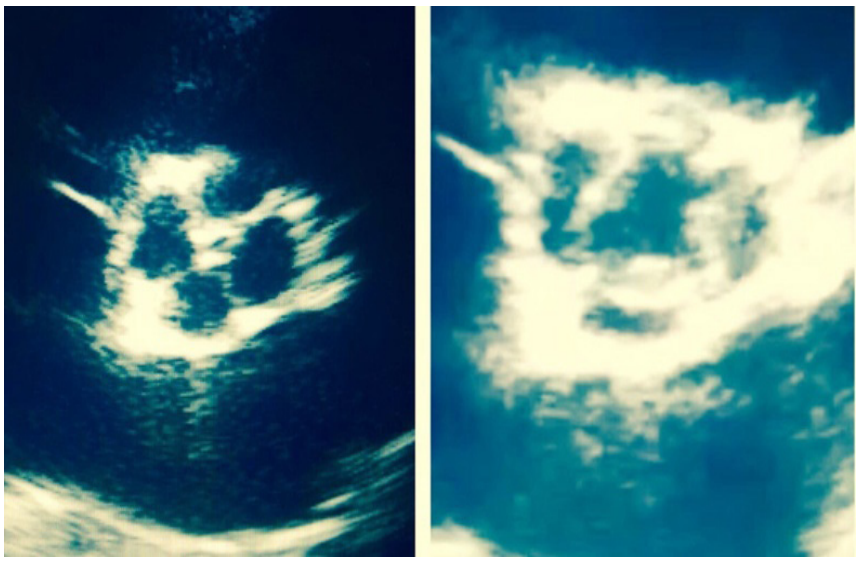

Figure 1 – *Quadricuspid aortic valve (patient 1).*

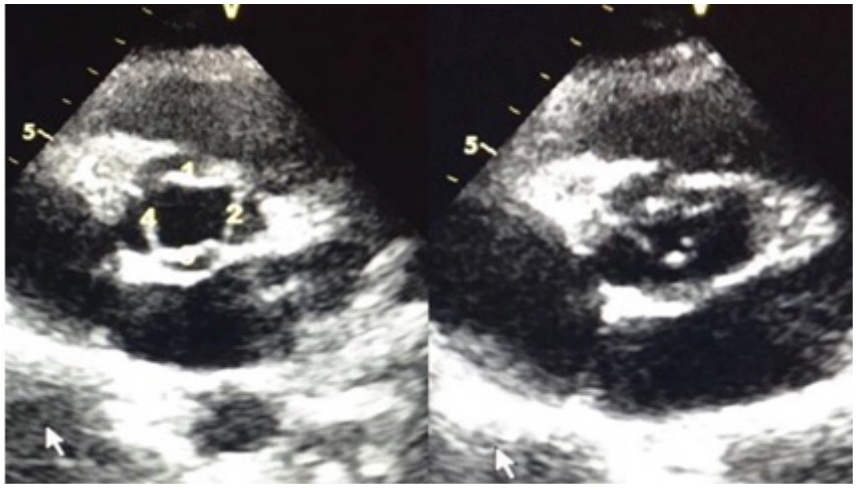

Figure 2 – *Quadricuspid aortic valve (patient 2).*

Most often, QAV is an isolated anomaly; however, it may sometimes be associated with other cardiac malformations. There are reports of cases of QAV in association with hypertrophic cardiomyopathy, interatrial and interventricular septal defect, pulmonary valve stenosis, aortic ring hypoplasia, mitral valve abnormalities, transposition of the great arteries and tetralogy of Fallot. Among them, the most common association is with the anomalous origin of the coronary artery ostia, found in 10% of the cases of QAV described in the literature. The main anomalies described were single coronary ostium and anomalous positions of the coronary ostia. There are also reported cases of association with Valsalva sinus fistula, Valsalva sinus aneurysm and ascending aortic aneurysm.

In patients with QAV, the development of aortic regurgitation is more common than stenosis. Patients with this disorder may require valve replacement surgery when they reach the fifth or sixth decade of life.[4] According to Hurwitz and Roberts,[2] the QAV reported in this case falls into category A (with four cusps of the same size), which is the most frequently found.

Treatment is based on the abnormality and degree of valve involvement, i.e., regurgitation and/or stenosis, and thickening and calcification degree. These anomalies define the choice of treatment (Figure 6). Valve replacement is the primary choice in most cases, and the procedure is performed around the fifth decade of life. In younger patients, other procedures are usually

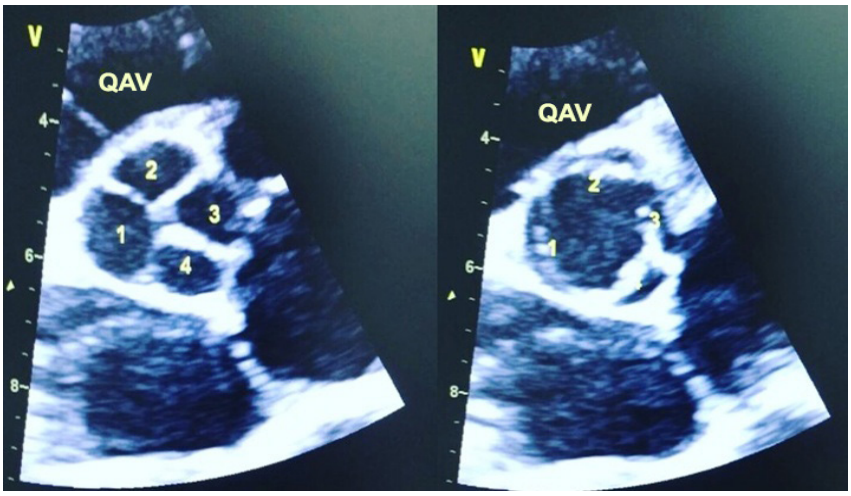

Figure 3 – *Quadricuspid aortic valve (patient 3).*

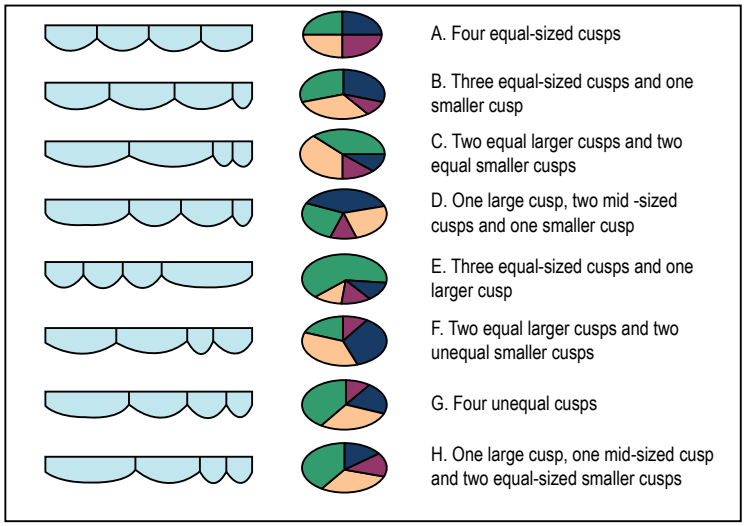

Figure 4 – *Classification according to the size of the valve leaflets (2).*

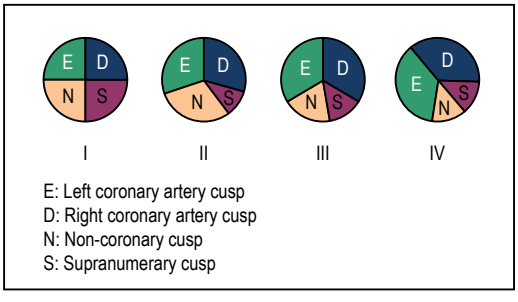

Figure 5 – *Classification according to the position of the accessory cusp.*[5]

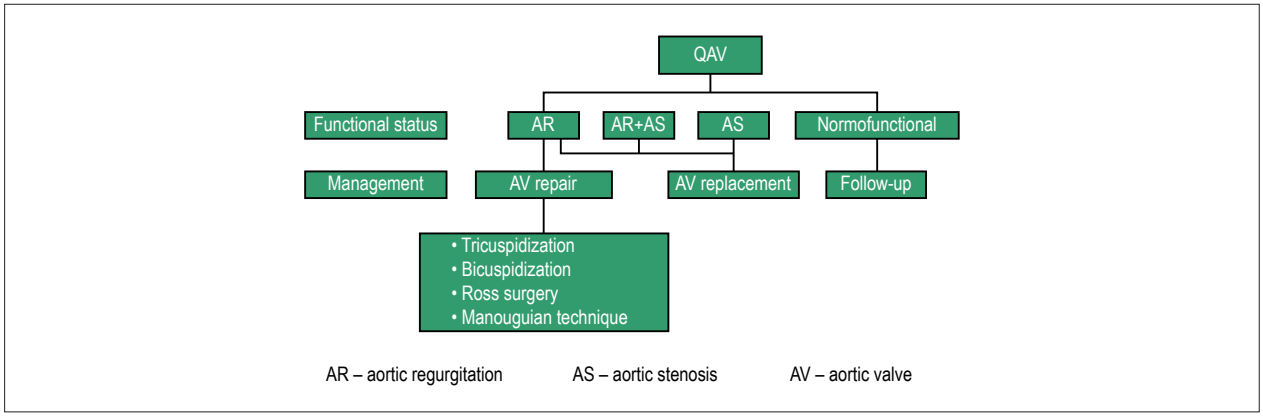

Figure 6 – *Definition of the treatment of patients with quadricuspid aortic valve (QAV).[6-8] AR: aortic regurgitation; AS: aortic stenosis; AV: aortic valve.*

chosen, such as valvuloplasty, which leads to tricuspidization of the valve, or Ross surgery, in which these valves do not present severe thickening and calcification. Ross surgery is performed with pulmonary autograft replacing the QAV, and with homograft, to replace the pulmonary valve.

Surgical treatment is recommended for patients presenting symptoms, with evidence of left ventricular dysfunction (reduced ejection fraction and systolic diameter greater than 55 mm) and who did not progress to the stage of cardiac decompensation.

Conclusion

Quadricuspid aortic valve is, in most cases, a rare and isolated anomaly diagnosed as an incidental finding or due to the investigation of heart failure in adulthood, with a high potential for presenting complications. Patients with this abnormality should be carefully evaluated be properly followed up.

Due to the risk of progressive aortic regurgitation and the potential evolution to endocarditis, it is important to consider the quadricuspid aortic valve as one of the probable diagnoses during the investigation of valve regurgitation. This anatomical disorder can sometimes be related to cardiac malformations, the most common being the association with the anomalous origin of coronary artery ostia. For this reason, the aortic leaflets should be evaluated during systole and diastole and, where possible, the origins of the coronary arteries must be identified.

Authors' contributions

Research creation and design: Cunha WC, Beal JR, Fontenele SMF, Rabelo AAK, Galdeano PA, Miguel GAS; Data acquisition: Cunha WC, Beal JR, Fontenele SMF, Rabelo AAK, Galdeano PA, Miguel GAS; Data analysis and interpretation: Cunha WC, Beal JR, Fontenele SMF, Rabelo AAK, Galdeano PA, Miguel GAS; Manuscript writing: Cunha WC, Beal JR, Fontenele SMF, Rabelo AAK, Galdeano PA, Miguel GAS; Critical revision of the manuscript for important intellectual content: Cunha WC, Beal JR, Fontenele SMF, Rabelo AAK, Galdeano PA, Miguel GAS.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

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Academic Association

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