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Isolated Quadricuspid Aortic Valve

Mehrnoush Toufan,[1,*] and Seyed Sajjad Mahmoudi[1]

[1]Cardiovascular Research Center, Tabriz University of Medical Sciences, Tabriz, IR Iran

[*]Corresponding author: Mehrnoush Toufan, Cardiovascular Research Center, Tabriz University of Medical Sciences, Tabriz, IR Iran. Tel: +98-4133357767, Fax: +98-41-33344021, E-mail: mtoufan@gmail.com

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Isolated quadricuspid aortic valve (QAV) is a very rare cardiac anomaly, with an estimated incidence of 0.031 to 0.043% among all congenital heart diseases (1). Although association with other anomalies has been reported, QAV very often presents as an isolated congenital anomaly (2).

According to Hurwitz and Roberts' classification, QAV is divided into 7 types (A to G) with respect to the size of the AV cusps (Table 1). Types A and B are the more common types. Aortic regurgitation as a usual complication of QAV occurs in type B with high eventuality because of its specific anatomy and unbalanced stress on the cusps.

Table 1. Hurwitz and Roberts' Anatomical Classification of Quadricuspid Aortic Valve

Type	Description
A	Four equal cusps
B	Three equal cusps and one smaller cusp
C	Two equal larger cusps and two equal smaller cusps
D	One large cusp, two intermediate cusps, and one small cusp
E	Three equal cusps and one larger cusp
F	Two equal larger cusps and two unequal smaller cusps
G	Four unequal cusps

Echocardiographic study in the AV plane of the short-axis view will manifest an X-shaped commissure pattern and a rectangular feature during diastole and systole, respectively (3).

QAV may occur in association with other deformities-including coronary artery abnormalities, ventricular septal defect, cardiomyopathy, pulmonary valve stenosis, Valsalva aneurysm, supravalvular aortic stenosis, and fibromuscular subaortic stenosis (4). Various coronary anomalies as common relevant malformations accompany QAV in about 10% of cases. These anomalies may include single coronary ostium, anomalous position of the coronary ostia, coronary-pulmonary artery fistula, and a mortal occlusion of a coronary artery ostium by the valve tissue (5).

The clinical course of QAV, especially that of the uncommon types, is not well known. No correlation has been found between valve morphology and function. In a recent review of all published cases, 66% presented severe, 8% moderate, and 8% mild aortic regurgitation. Among all cases, 13% had regurgitation and aortic stenosis and 10% had a normal valve function (6). There have been some cases of bacterial endocarditis affecting QAV; nevertheless, it is not fully established whether or not QAV is more vulnerable to endocarditis. Endocarditis prophylaxis is no longer recommended in the management of patients with QAV.

The surgical indication for patients with QAV depends on the extent of aortic regurgitation and its associated lesions. For aortic regurgitation, the indication is almost the same as regurgitation caused by a tricuspid AV. However, surgeons should pay attention to the origin of the coronary artery to prevent injury during surgery. Valve replacement is the most widely used operation in such patients. Some surgeons have tried tri-cuspidized or bi-cuspidized repair techniques and obtained good short-term results, but the long-term outcome has yet to be defined (7).

Early diagnosis of QAV in life is important because about 75% of these individuals will require valve replacement surgery in the fifth or sixth decade of life, due to worsening aortic insufficiency. Patients with QAV should have regular long-term follow-up on account of significant aortic valve insufficiency and need for valve replacement before left ventricular impairment.

Footnote

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