Quadricuspid aortic valve as a cause of severe aortic regurgitation

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Key words:
Aortic valve; Echocardiography; Valve disease.

Quadricuspid aortic valves (QAVs) constitute a rare congenital malformation, with an incidence ranging from 0.008 to 0.048%. We report a case of severe aortic regurgitation associated with a QAV, which was diagnosed intraoperatively using transesophageal echocardiography.

Since the first case described in 1862, 186 QAVs have been reported. In most cases, QAVs are associated with valve regurgitation, with a concurrent stenosis in some patients, while only a small number of QAVs are functionally normal. Once the diagnosis has been made, echocardiographic follow-up is recommended, as progression to severe valve regurgitation is common. Antibiotic prophylaxis is advisable for dental, and "dirty" surgical procedures, to minimize the risk of infective endocarditis.

(Ital Heart J 2005; 6 (2): 157-159)

Received September 8, 2004; revision received December 17, 2004; accepted December 20, 2004.

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Introduction

Congenital malformations of the aortic valve are not uncommon, with the incidence of bicuspid aortic valve approximating 2%[1]. However, quadricuspid aortic valves (QAVs) are rare, with the reported incidences ranging from 0.008%[2] to 0.048%[3]. QAVs were traditionally reported as an incidental finding at necropsy, surgery, or aortography. This condition is now being increasingly detected by means of echocardiography.

We report a case of QAV leading to severe aortic insufficiency, detected at intraoperative transesophageal echocardiography.

Case report

A 77-year-old male with a history of aortic valve regurgitation, depressed left ventricular function and chronic atrial fibrillation, was referred to our cardiac surgery department for evaluation.

His preoperative transthoracic echocardiography showed severe aortic valve regurgitation, with a mild increase in the transvalvular gradient (ΔP_{max} 22 mmHg), mild mitral regurgitation, and left ventricular dilation (left ventricular end-diastolic diameter 67 mm, left ventricular end-diastolic volume 190 ml) with a depressed systolic function (left ventricular ejection fraction 28%).

The patient was scheduled for elective aortic valve surgery and mitral valve repair.

Transesophageal echocardiography was performed intraoperatively to assess the mechanism of aortic regurgitation, and the feasibility of valve-sparing surgery.

A mid-esophageal short-axis view on the aortic plane revealed a QAV, with a small accessory leaflet located between the non-coronary and the left coronary leaflets (Fig. 1). The diagnosis was confirmed at surgical inspection (Fig. 2), and is well documented in the excised valve (Fig. 3).

The anatomy of the valve suggested its replacement, and a tissue valve was implanted (SJM Supra Tissue Valve #23, St. Jude Medical, St. Paul, MN, USA). Mitral annuloplasty was also performed using a 28 mm annuloplasty ring (Future Band #28, Medtronic, Minneapolis, MN, USA).

The intraoperative course was regular, and the surgical result was excellent, with normal function of the prosthetic valve, and no residual mitral regurgitation. The patient had an uneventful recovery, and was discharged home in good conditions.

Discussion

QAV constitutes a rare condition. The first known case was reported by Balington[4] in 1862, and according to a recent review by Tutarel[5], 186 cases have since been identified.

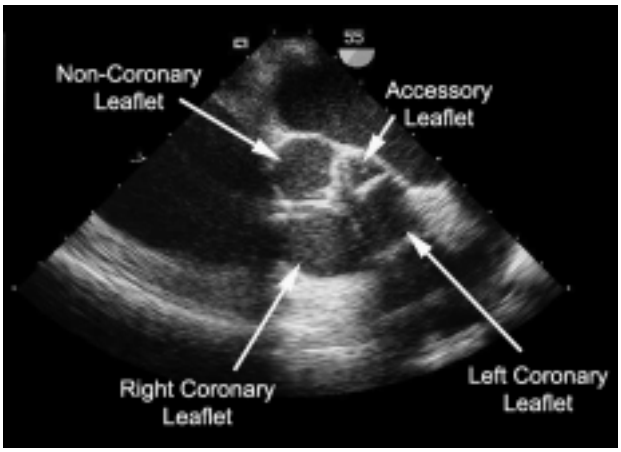

Figure 1. *Intraoperative transesophageal echocardiography: a mid-esophageal short-axis view of the aortic valve shows the presence of a small accessory leaflet between the non-coronary and left coronary leaflets.*

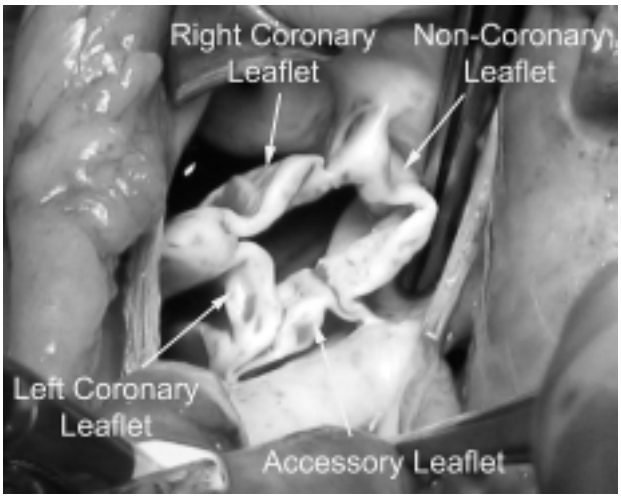

Figure 2. *Surgical view of the aortic valve; the accessory leaflet is documented.*

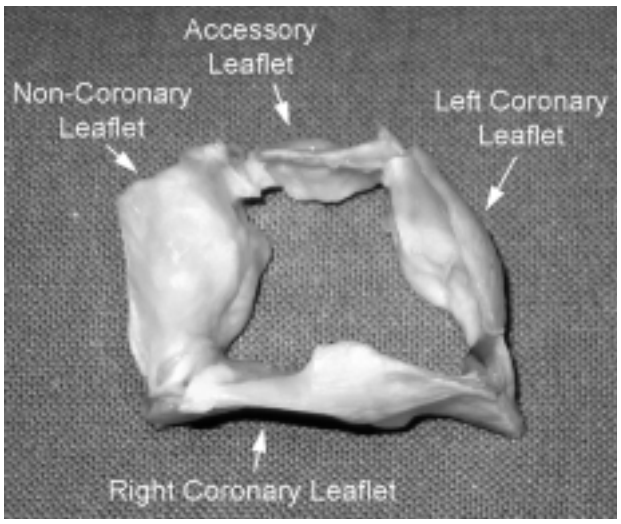

Figure 3. *Anatomic preparation of the excised aortic valve; the valve is fixed in the systolic position.*

QAVs were classified by Hurwitz and Roberts[6] in 1973 according to the morphology of the valve leaflets. They described seven variations: A = four equal cusps; B = three equal cusps and one smaller cusp; C = two equal larger and two equal smaller cusps; D = one large, two intermediate and one small cusp; E = three equal cusps and one larger cusp; F = two equal larger and two unequal smaller cusps; and G = four unequal cusps.

In the present case the QAV was characterized by three equal cusps and a smaller cusp between the left coronary and the non-coronary leaflets (type B). This has been reported as the most common variation. However, analysis of all published cases by Tutarel[5] revealed a higher frequency of type A (51 cases among the 124 QAVs in which any comment was made on the morphology of the leaflets, while type B was detected in only 43 cases).

QAV was detected in 95 (51.1%) cases at echocardiography, in 42 (22.6%) at surgery, while 29 (15.6%) were found at *post-mortem* examination, and 12 (6.5%) at aortography; in 7 cases the diagnostic method was not reported, while one QAV was identified at cine-magnetic resonance imaging.

In our patient, the diagnosis of QAV was made at intraoperative transesophageal echocardiography, while repeated transthoracic examinations had revealed severe regurgitation but had failed to identify the accessory leaflet despite an adequate acoustic window. This may be partly due to the posterior position of the accessory leaflet; moreover, Timperley et al.[7] stated that identification appears easier in the presence of four equal cusps (type A), while it is more challenging in type B QAVs.

Aortic regurgitation is the most common functional abnormality associated with QAV, pure regurgitation accounting for 74.7% of the reported cases, and combined stenosis and regurgitation for another 8.4%. Only 16.2% of QAVs presented with a normal function. No cases were characterized by pure aortic stenosis. Although QAV is a congenital anomaly, regurgitation does not occur until adulthood. Therefore, if a QAV is found at echocardiography, follow-up assessment is required as progress to severe regurgitation is to be expected.

Even though the risk of endocarditis is unclear, at least three cases have been reported[7], and therefore endocarditis prophylaxis is advised.

In conclusion, QAVs are a cause of significant aortic regurgitation, often requiring surgery in the fifth and sixth decades; transthoracic echocardiography should be able to clarify the valve morphology in a vast majority of patients presenting with aortic regurgitation. However, in young patients investigated for aortic regurgitation, in whom transthoracic echocardiography has failed to identify the number of valve leaflets, it may be advisable to perform a transesophageal echocardiography. In all cases of QAV planned echocardiographic follow-up is recommend-

ed. There is a risk of infective endocarditis associated with QAVs, and patients should thus undergo antibiotic prophylaxis for dental and "dirty" surgical procedures.

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