

Congenital Quadricuspid Aortic Valve: Report of Nine Surgical Cases

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We report the clinicopathologic characteristics of the congenital quadricuspid aortic valve necessitating surgery. Among 616 patients for whom we performed an aortic valve operation over the past 20 years, nine patients (1.46%) (five men and four women, mean age 60 years) with quadricuspid aortic valve were encountered. All had aortic regurgitation (AR) except one with aortic stenosis and mild regurgitation (ASr). All were free of cardiac anomaly including that of the coronary arterial system. Macroscopically, severe calcification of the valve was seen in the one case of ASr. Fenestration of the cusp was seen in five cases of AR. Infective endocarditis was not seen. Histological study disclosed fibrous thickening and myxoid degeneration in the AR cases. In accordance with the Hurwitz and Roberts classification, four valves were type b (three equal-sized cusps and one smaller cusp), two valves were type a (four equal-sized cusps), two valves were type d (one large, two intermediate, and one small cusp), and one valve was type g (four unequal-sized cusps). Valve repair failed in one patient and was converted to valve replacement during the operation. All patients underwent successful aortic valve replacement (AVR). (Ann Thorac Cardiovasc Surg 2003; 9: 134–7)

Key words: quadricuspid aortic valve, hemodynamics, surgery, aortic valve replacement (AVR), aortic valve repair

Introduction

Congenital quadricuspid aortic valves are rare, whereas bicuspid valves are relatively common.¹⁾ The quadricuspid aortic valve frequently functions abnormally; the most common abnormality is valvular insufficiency. We report on nine cases of congenital quadricuspid aortic valve treated surgically in the past 20 years.

Cases

We encountered nine cases of congenital quadricuspid aortic valve among 616 cases of aortic valve surgery at Kagoshima University Hospital between January 1982 and December 2001. The patients were comprised of five

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men and four women with a mean age at surgery of 60 years (range, 47 to 70 years) (Table 1).

All patients had aortic regurgitation (AR) except for one with aortic stenosis and mild regurgitation (ASr). All patients were free of other cardiac anomalies including those of the coronary arterial system. Four patients were diagnosed preoperatively with quadricuspid aortic valve, one by aortography and three by two-dimensional echocardiography. The remaining four diagnoses were made at the time of surgery; however, the four cusps of aortic valve were apparent upon further detailed review of these patients' aortograms.

Macroscopically, marked calcification of the valve was seen in the patient with ASr, whereas no calcification was observed in the eight patients with AR. Small cusp fenestration was seen in five of the AR patients; however, this was probably not the cause of the AR. Histological study of the excised valves disclosed fibrous thickening and myxoid degeneration in the AR patients. Infective endocarditis was not seen in any of the valves.

In accordance with the classification system devised

Table 1. Surgical cases of quadricuspid aortic valve

| No. | Age (years) | Sex | Functional status | Diagnostic procedure | Type (Hurwitz and Roberts classification) | Associated condition | Operation |
|-----|-------------|--------|-------------------|----------------------|---|----------------------|-----------|
| 1 | 68 | Female | AR | UCG | b | AP | AVR+CABG |
| 2 | 49 | Male | AR | Operation | b | MR | AVR+MVR |
| 3 | 61 | Male | AR | Operation | b | None | AVR |
| 4 | 66 | Female | ASr | Operation | a | None | AVR |
| 5 | 57 | Male | AR | Operation | d | None | AVR |
| 6 | 59 | Male | AR | Operation | b | None | AVR |
| 7 | 65 | Female | AR | Aortography | g | None | AVR |
| 8 | 70 | Male | AR | UCG | a | None | AVP→AVR |
| 9 | 47 | Female | AR | UCG | d | None | AVR |

AR: aortic regurgitation, ASr: aortic stenosis and mild regurgitation, UCG: ultrasonic echocardiography, AP: angina pectoris, MR: mitral regurgitation, AVR: aortic valve replacement, CABG: coronary artery bypass grafting, MVR: mitral valve replacement, AVP: aortic valvuloplasty

by Hurwitz and Roberts,²⁾ four valves were type b (three equal-sized cusps and one smaller cusp), two valves were type a (four equal-sized cusps), two valves were type d (one large, two intermediate, and one small cusp), and one valve was type g (four unequal-sized cusps). A supporting fibrous band stretching from the commissure between the right and left coronary cusps to the aortic wall was seen in one of the four type b patients. The one patient with ASr was given a type a classification (Table 1).

The other type a patient (with AR) (Figs. 1, 2) underwent annuloplasty reinforced with a 2-mm-wide Gore-Tex strip (Gore-Tex EPTFE patch II/pericardial membrane, W.L. Gore & Associates, Inc., Flagstaff, AZ, USA); however, the

valve was then excised and replaced with a mechanical valve during the operation because residual regurgitation was detected by transesophageal echocardiography. All patients, including this one, underwent successful aortic valve replacement (AVR) with a mechanical valve prosthesis. Concomitant coronary artery bypass grafting was performed in one patient with angina pectoris due to coronary arterial sclerosis,³⁾ and the mitral valve was replaced in another patient with mitral regurgitation.

Discussion

Quadricuspid aortic valve is a very rare congenital ab-



A



B

Fig. 1. Echocardiographic images from patient 8 showing the quadricuspid aortic valve with poor coaptation of the four equally sized cusps during diastole (A) and with all four cusps open (B).

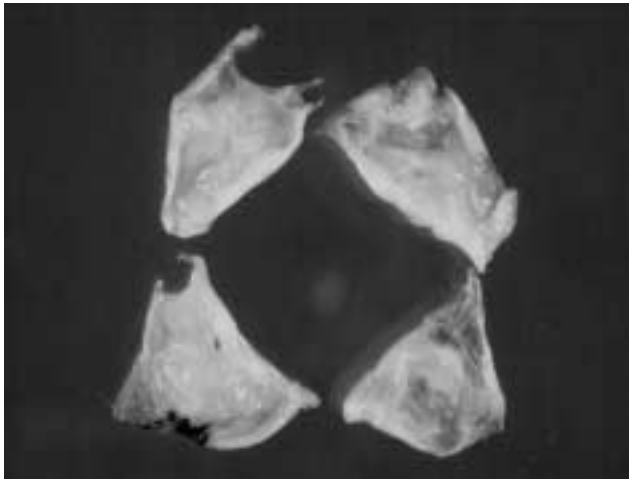


Fig. 2. The four cusps excised at surgery from patient 8.

normality. Most cases have been discovered incidentally at autopsy^{2,4,5)} or during aortic valve surgery,⁶⁾ and a lesser number of cases have been detected preoperatively by aortography⁷⁾ or two-dimensional echocardiography.^{8,9)} Two reported autopsy series had an incidence of 0.008% (2 in 25,666 necropsies)⁴⁾ and 0.033% (2 in 6,000 necropsies).²⁾ In a series of 60,446 echocardiograms, 8 aortic valves were found to be quadricuspid (an incidence of 0.013%).¹⁰⁾ Among 225 patients who underwent aortic valve surgery for pure AR, 2 were found to have quadricuspid aortic valve (an incidence of 0.88%).¹¹⁾ Mathison et al.¹²⁾ reported 2 quadricuspid aortic valves seen during 363 aortic valve procedures (an incidence of 0.55%). In our series, the incidence was 1.46%, which was considerably higher than that of these other reports.

Hurwitz and Roberts³⁾ classified the quadricuspid semilunar valve into seven types on the basis of the relative size of the four cusps. In the present study, type b (three equal-sized cusps and one smaller cusp) was most prevalent (4 of 9), as it was in previously reported autopsy²⁾ and surgical series.¹³⁾ In the noted echocardiographic series, there was an equal incidence of type a and type b valves.¹⁰⁾ Two-dimensional echocardiography has become the diagnostic procedure of choice because, with it, the four cusps and their relative sizes can be easily recognized.^{10,14)} However, even when the echocardiographic images suggest cusps of similar size, cusps of unequal size may be found at surgery.¹⁴⁾

Unlike the quadricuspid pulmonic valve, the quadricuspid aortic valve frequently functions abnormally; stenosis is unusual and regurgitation is the most prevalent abnormality. Of 18 reported postmortem cases of quadricuspid aortic valve, 1 (6%) was stenotic, 7 (39%)

were incompetent, and 10 (56%) were hemodynamically normal.⁵⁾ Of 34 patients with quadricuspid aortic valve undergoing surgery, 4 (12%) had aortic stenosis.¹³⁾ In our series, only one of nine patients (11%) had aortic stenosis with mild regurgitation. Sievers et al.¹⁵⁾ suggested that disproportion in the size of the cusps may result in abnormal valve motion that may lead to fibrous thickening of the valve, thus aggravating incomplete coaptation of the cusps. Feldman et al.¹⁰⁾ found AR to be more common in patients with a smaller accessory cusp (type b) and suggested that there is an unequal distribution of stress and abnormal leaflet coaptation in this particular type that may result in progressive AR.

Congenital quadricuspid aortic valve is usually an isolated lesion, but several concomitant congenital abnormalities have been described, including anomalies of the coronary arteries,^{6,16-18)} ventricular septal defect,²⁾ patent ductus arteriosus,^{2,4)} and subaortic fibromuscular stenosis.¹⁹⁾ Although there were no coronary artery anomalies in the present series, such anomaly is frequently found.¹³⁾ Embryologically, the semilunar valves are derived from mesenchymal swelling in the aortic and pulmonary trunk after they have separated. Abnormal cusp formation results from either aberrant fusion of the aorticopulmonary septum or from abnormal mesenchymal proliferation in the common trunk.^{2,4)} Because septation of the embryonic arterial trunk and development of the aortic valve leaflets occur temporally just after development of the coronary artery origins from the sinuses of Valsalva, researchers have speculated that a developmental error might result in both an abnormal number of aortic cusps and abnormality of the coronary arteries.¹⁸⁾

The quadricuspid aortic valve is replaced in the majority of patients requiring surgery; only a few cases of in situ surgical repair have been reported. Iglesias and co-workers¹⁹⁾ reported on a patient with a type b quadricuspid aortic valve that was converted into a tricuspid valve by suturing together the common commissure between the right coronary and supernumerary cusps. Chandrasekaran et al.⁹⁾ and van Son et al.²⁰⁾ both reported three cases of successful surgical repair; however, neither group reported the Hurwitz and Roberts classifications or the operative details. Long-term results were also not described. In a different study, however, one patient with an aortic valve of unknown classification required AVR 2.5 months after the original valve repair that had consisted of annuloplasty with mid-leaflet excision of the prolapsed cusp.²¹⁾ In one patient with four unequal-sized cusps (type g), valvuloplasty with plication after excision of the supernumerary

cuspid failed and was converted to AVR during the operation because of poor coaptation due to the unequal sizes of the residual three cusps.²²⁾ In our series, annuloplasty was also attempted in one patient with four equal-sized cusps (type a). This failed, however, and the procedure was converted to that of AVR. In this case, transesophageal echocardiography clearly showed poor coaptation of the four equally sized leaflets in diastole and a central regurgitation jet during incomplete closure of the aortic valve. The annuloplasty already mentioned was performed to reduce the diameter of the aortic annulus to render the aortic cusps more coaptive; however, moderate central regurgitation was detected by immediate postoperative transesophageal echocardiography. Because we were able to reduce the regurgitation by digital compression of the sinotubular junction at the site of the noncoronary cusp, the regurgitation could have been controlled by further shortening of the noncoronary annulus. In general, quadricuspid valves seem to be less amenable than tricuspid or bicuspid valves to valvuloplasty techniques probably due to the longer coaptation line present in quadricuspid valves. However, it seems improper not to consider valve repair first because this may be the most acceptable procedure to treat heart failure and restore satisfactory aortic valve function without incurring thromboembolic or hemorrhagic complications of AVR.

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