

**Case
Report**

Quadricuspid Aortic Valve: A Report on a 10-Year Case Series and Literature Review

Masato Hayakawa, MD, Tohru Asai, MD, PhD, Takeshi Kinoshita, MD, PhD,
and Tomoaki Suzuki, MD, PhD

Quadricuspid aortic valve is rare, with an estimated incidence of 0.008% to 1.46%. Most cases are encountered incidentally during aortic valve surgery or autopsy. The condition frequently progresses to aortic regurgitation, which can manifest in adulthood and may require surgical treatment. This anomaly may sometimes be associated with other malformations, the most common being coronary artery anomalies. We performed a retrospective chart review of patients diagnosed with quadricuspid aortic valve between January 2002 and July 2012 and report here on cases treated surgically. We encountered three cases of quadricuspid aortic valve among 627 patients undergoing aortic valve surgery at our institution (an incidence of 0.48%). All three had aortic regurgitation and two were free of cardiac anomaly; the other had ascending aortic aneurysm and coronary malformation. According to Hurwitz's classification, two of the valves were of type b and one of type d. Under Nakamura's classification, meanwhile, two of the valves were type II and the other type III. All patients underwent successful aortic valve replacement and had uneventful postoperative courses.

Keywords: quadricuspid aortic valve, aortic valve replacement, aortic regurgitation

Introduction

Congenital quadricuspid aortic valve is a rare malformation with an estimated incidence of 0.008%¹⁾ to 1.46%.²⁾ Most cases are discovered as an incidental finding during aortic valve surgery or autopsy. The condition frequently progresses to aortic regurgitation, which can manifest in adulthood and may require surgical treatment. This anomaly may sometimes be associated with other malformations, the most common being coronary

artery anomalies. We report on three cases of quadricuspid aortic valve treated surgically in the past 10 years.

Case Report

We performed a retrospective chart review of patients diagnosed with quadricuspid aortic valve between January 2002 and July 2012, during which period we encountered three cases among 627 patients undergoing aortic valve surgery at our institution. The incidence was thus 0.48%, which is a little lower than in other reports.

Case 1

The first case involved a 70-year-old man with complaints of chest pain. The patient had received a diagnosis of aortic regurgitation at a clinic 5 years before admission. Two months before admission, he experienced occasional chest pain and visited another hospital, where chest computed tomography and echocardiography were performed, revealing an ascending aortic aneurysm

Division of Cardiovascular Surgery, Department of Surgery, Shiga University of Medical Science, Otsu, Shiga, Japan

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Corresponding author: Masato Hayakawa, MD. Division of Cardiovascular Surgery, Department of Surgery, Shiga University of Medical Science, Setatsukinowa, Otsu, Shiga 520-2192, Japan
Email: yunta_sp@mail.goo.ne.jp
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(maximum diameter 62 mm) and moderate aortic regurgitation. The ascending aorta and aortic valve were replaced, but the quadricuspid aortic valve was not detected until directly visualized in the operating theater during the aortic valve replacement. The quadricuspid aortic valve had three equal cusps and one small cusp (type b in Hurwitz's classification). The accessory cusp was located between the right and non-coronary cusps. The right coronary artery originated from near the commissure between the left and right coronary cusps. Aortic valve replacement was carried out with a 23 mm Carpentier-Edwards Perimount valve and the ascending aorta was replaced with a 26 mm J Graft Shield Neo. The patient was discharged on postoperative day 15 after an uneventful postoperative course and was doing well nine years after surgery.

Case 2

The second case involved a 62-year-old woman with a known history of aortic regurgitation of more than 20 years. Two months before admission, severe aortic regurgitation was diagnosed via echocardiogram at another hospital. Aortic valve replacement was carried out but the quadricuspid aortic valve was again not discovered until directly visualized during surgery. The valve had three equal cusps and one small cusp (type b in the Hurwitz classification). The accessory cusp was located between the left and non-coronary cusps. There was no abnormality in the coronary arteries. A Carpentier-Edwards Perimount valve of 21 mm in diameter was implanted after excision of the native valve. The patient was discharged on postoperative day 12 after an uneventful postoperative course and was doing well five years after surgery.

Case 3

The third case was a 66-year-old-man who had complained of shortness of breath about five years earlier. The patient was admitted repeatedly to another hospital due to heart failure. The chest radiograph showed moderate cardiomegaly, with a cardiothoracic ratio of 0.64, while echocardiography demonstrated moderate to severe aortic regurgitation, mild mitral regurgitation, and moderate tricuspid regurgitation. Coronary angiography was normal. Aortic valve replacement, mitral annuloplasty, and tricuspid annuloplasty were performed. The quadricuspid aortic valve was discovered on direct inspection of the valve, with the accessory cusp found again between the right and non-coronary cusps (**Figs. 1 and 2**). The

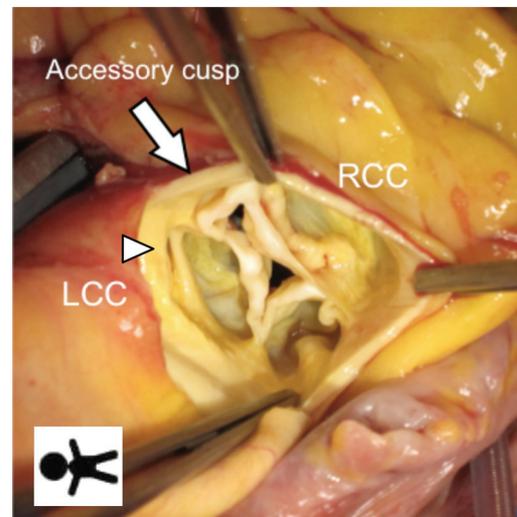


Fig. 1 Intraoperative findings. We discovered quadricuspid aortic valve in case no. 3 during surgery. The accessory cusp was located between the right and left coronary cusps. RCC: right coronary cusp; LCC: left coronary cusp; arrow head: left coronary ostium.

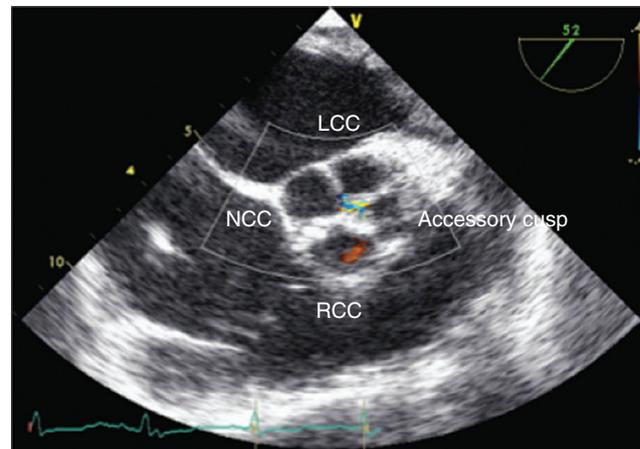


Fig. 2 Trans-esophageal echocardiographic image (case no. 3). Intraoperative trans-esophageal echocardiogram showed this patient to have quadricuspid aortic valve, a finding not revealed by trans-thoracic echocardiography before surgery. RCC: right coronary cusp; LCC: left coronary cusp; NCC: non-coronary cusp.

aortic valve showed one large, two intermediate, and one smaller cusp, which corresponded to type d under Hurwitz's classification (**Fig. 3**). The patient was discharged on postoperative day 20 after an uneventful postoperative course and was doing well six months after surgery.

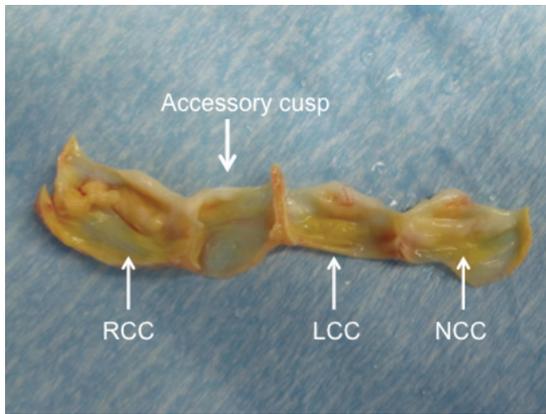


Fig. 3 Excised specimen. In case no. 3, the valve had one large, two intermediate, and one small leaflet, corresponding to type d under Hurwitz's classification. RCC: right coronary cusp; LCC: left coronary cusp; NCC: non-coronary cusp.

Discussion and Conclusion

Quadricuspid aortic valve is a very rare congenital abnormality. Systematic autopsy studies have estimated its incidence at between 0.008%¹⁾ and 0.033%,³⁾ while a more recent echocardiographic review reported an incidence of 0.043%.⁴⁾ Among patients undergoing aortic valve replacement, the incidence ranges from 0.55% to 1.46%.²⁾ In our series, we found three quadricuspid aortic valves among 627 aortic valve procedures (an incidence of 0.48%).

Two classifications are usually used to describe quadricuspid aortic valves. The first, established by Hurwitz and Roberts,³⁾ classifies the quadricuspid aortic valve into seven types on the basis of the relative size of the four cusps. The most common type consists of three equally sized cusps and one smaller cusp (type b). Nakamura and colleagues⁵⁾ designed a classification which focuses on the position of the accessory cusp. The most common is type II, in which the accessory cusp is located between the right coronary cusp and the non-coronary cusp. In the present series, two of the valves were of type b and one of type d according to Hurwitz's classification. Under Nakamura's classification, meanwhile, two of the valves were of type II and one of type III.

Embryologically, the semilunar valves are derived from mesenchymal swelling in the aortic and pulmonary trunk after partition. Abnormal cusp formation results from either aberrant fusion of the aorticopulmonary septum or from abnormal mesenchymal proliferation in the common trunk.^{1,3)} The development of the aortic valve

leaflets occurs immediately after the development of the coronary artery origins from the sinuses of Valsalva. It is therefore possible that anomalies in these two areas are related embryologically.

A quadricuspid aortic valve usually appears as an isolated congenital anomaly, but may also be associated with other abnormalities, including patent ductus arteriosus, ventricular septal defect, pulmonary valve stenosis, subaortic fibromuscular stenosis, and anomalies of the coronary arteries.^{1,3)} Malformation of the coronary artery origin and distribution occurs in around 30% of quadricuspid aortic valves.^{1,3,4)} In our series, there was coronary displacement only in the first case (an incidence of 33%). From the viewpoint of the surgeon, it is important to notice any displacement of the coronary ostium in order to prevent ostial obstruction at the time of valve replacement or repair. On the other hand, ascending aortic aneurysm coexisting with quadricuspid aortic valve, as in our first case, is quite rare.⁶⁾

Unlike quadricuspid pulmonary valve, quadricuspid aortic valve tends to progress to regurgitation, resulting from progressive leaflet fibrosis and progressive failure of leaflet coaptation with aging. With unequal distribution of stress and abnormal leaflet coaptation, aortic regurgitation may occur later in life, but not in younger patients. There are also some differences between bicuspid aortic valve and quadricuspid aortic valve in aortic regurgitation. Aortic regurgitation in bicuspid aortic valve may cause diffuse dilatation of the ascending aorta secondary to medial disarray. Approximately half of young adults with a bicuspid aortic valve have aortic root dilatation and are thus potential candidates for resultant aortic regurgitation.⁷⁾ On the other hand, in patients with quadricuspid aortic valve, the concurrent presence of an ascending aortic aneurysm has only rarely been reported. The relative absence of associated aneurysm of the aorta has clinical and management significance, and has not been addressed previously. Histologic and clinical evaluation of the ascending aorta in quadricuspid aortic valve is thus advisable.⁸⁾

In our series, all patients showed aortic regurgitation, but no stenosis. The location of aortic regurgitation was considered to be at the accessory cusp in all patients. Aortic valve replacement for a quadricuspid valve is generally the treatment of choice for patients with aortic regurgitation, with few cases of aortic valve repair reported. Naito and colleagues reported that, in a patient with quadricuspid aortic valve (type b in Hurwitz's classification), aortic valve repair was performed by suturing

together the commissure between the right coronary cusp and the accessory cusp, thus converting the quadricuspid valve into a tricuspid valve.⁹⁾ In our series, all patients underwent aortic valve replacement with a bioprosthetic valve. As previously noted, attention needs to be given to preventing coronary ostial obstructions or unintended injury at the time of valve replacement or repair in quadricuspid aortic valve. In all our patients, the alignment of the bioprosthetic valve against the commissure was performed as is usual in aortic valve replacement and there was no complication of the coronary arteries. The risk of complete heart block is also potentially increased after valve replacement because the accessory cusp is commonly located between the right coronary cusp and the non-coronary cusp, positioned over the membranous septum.¹⁰⁾ In our series, all patients underwent aortic valve replacement without complications.

Finally, in our series, no patient was diagnosed preoperatively with quadricuspid aortic valve. This was because all patients preoperatively underwent only trans-thoracic echocardiography, not trans-esophageal echocardiography, since quadricuspid aortic valve was not envisaged as a differential diagnosis in aortic regurgitation. In recent years, real-time three-dimensional trans-esophageal echocardiography has proved helpful in reaching an accurate diagnosis in cases of aortic regurgitation, especially in patients with quadricuspid aortic valve.

Disclosure Statement

All authors have no conflict of interest.

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