

Aortic Valve Replacement for Quadricuspid Aortic Valve Associated With Right Coronary Ostium Anomaly: a Case Report

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Case report

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Abstract

Background: Quadricuspid aortic valve is a rare congenital heart disease that may be associated with coronary ostium anomalies. Care should be taken to avoid occluding or compressing the coronary ostium while performing aortic valve replacement.

Case presentation: Herein, we report a case of a 59-year-old woman who underwent aortic valve replacement for a quadricuspid aortic valve with severe aortic regurgitation. Intraoperatively, the aortic valve had four cusps of almost equal size and the right coronary artery arose adjacent to one of the commissures. The annular stitches were placed in a non-everting mattress fashion with pledgets on the ventricular side, and stitches near the right coronary ostium were transitioned to the subannular ventricular myocardium to maintain the distance from the ostium. Further, we selected a small prosthesis because oversized prosthetic valve could potentially compress the right coronary ostium.

Conclusions: While performing aortic valve replacement for a quadricuspid aortic valve associated with a right coronary ostium anomaly, careful selection of the size of the prosthesis and modification of the annular stitches are essential to prevent obstruction of the coronary ostium.

Background

Quadricuspid aortic valve (QAV) is a rare congenital heart disease that is less common than bicuspid and unicuspid aortic valve [1–5]. Coronary artery and coronary ostium anomalies are present in 2–10% of patients with QAV [2, 3, 6–8]. The functional status of QAV is predominantly pure aortic regurgitation (AR) [3, 6]. Clinical manifestations, such as palpitations, dyspnea, fatigue, and chest pain, depend on the functional status of QAV and usually present in the fifth or sixth decade of life [2]. Standard surgical repair is through aortic valve replacement (AVR), although aortic valve plasty, such as bicuspidalization and tricuspidalization, is also performed [6]. In the present case, we performed AVR for QAV associated with a right coronary ostium anomaly and described a modification of the annular stitches to prevent obstruction of the right coronary ostium.

Case Presentation

A 59-year-old female patient with asymptomatic severe AR was referred to our institution for surgical treatment. She was being followed up at an outpatient clinic for QAV and moderate AR for 7 years. AVR was indicated for the progressively worsening AR and left ventricular function.

On admission, the patient's blood pressure was 122/62 mmHg, and heart rate was 102 bpm with an irregular rhythm. Chest radiography showed a cardiothoracic ratio of 56%. Electrocardiography (ECG) revealed a heart rate of 100 bpm with atrial fibrillation. Transthoracic echocardiography revealed a QAV with a severe central AR jet due to incomplete coaptation. The left ventricular ejection fraction was 50%, without local asynergy. The left ventricular end-systolic and end-diastolic diameters were 46 mm and 62 mm, respectively, and the diameter of the aortic valve annulus was 23 mm. Coronary angiography

revealed no significant coronary artery stenosis or anomalies. ECG-gated enhanced computed tomography was not performed, and no coronary ostium anomalies were detected preoperatively.

The patient underwent median sternotomy. The aortic valve had four cusps of almost equal size (Hurwitz and Roberts type A [1], Fig. 1). Macroscopically, partial calcification and thickening of the cusps were observed. The left coronary ostium was located in the middle of the left aortic sinus, whereas the right coronary ostium was located near the commissure between the right coronary cusp and one of the two non-coronary cusps. After excision of all cusps, the annular stitches were placed in a non-everting mattress fashion with pledgets on the ventricular side, and three stitches near the right coronary ostium were transitioned to the subannular ventricular myocardium to maintain the distance from the ostium (Fig. 2). We decided to use a biologic prosthesis, according to the patients' desire. Although a 25-mm prosthetic sizer could pass through the annulus, we selected a 23-mm Inspiris Resillia biologic prosthesis (Edwards Lifesciences, Irvine, California, United States of America), because an oversized prosthetic valve could potentially compress the right coronary ostium. In addition to AVR, pulmonary vein isolation using AtriCure (AtriCure, Mason, OH, USA) and left atrial appendage closure using AtriClip (AtriCure) was performed for atrial fibrillation.

The patient tolerated the procedure adequately, and postoperative echocardiography revealed normal prosthetic valve function without paravalvular leakage. Except for recurrent atrial fibrillation, the postoperative course was uneventful. The patient was discharged on postoperative day 17.

Discussion

Coronary artery and coronary ostium anomalies, such as a single coronary artery and displacement of the left and right coronary ostia, are present in 2–10% of patients with QAV [2, 3, 6–8]. While performing AVR, care should be taken to avoid occluding or compressing the coronary ostium. Withana et al. reported AVR in a patient with a low origin of both the main coronary arteries and hypoplastic aortic annulus. Implantation of an oversized prosthetic valve resulted in severe stretching of the aortic annulus and the two coronary orifices, ultimately causing coronary ostial stenosis and occlusion [9]. Alsaddique et al. reported AVR in a patient with the left coronary ostium located close to the annulus. They placed everting mattress sutures with the pledgets on the aortic side; thus, the pledgets themselves partially obstructed the left coronary ostium and eventually required coronary artery bypass grafting [10]. In the case of an anomalous left circumflex artery originating from the right coronary artery and coursing behind the aortic annulus, the risk of ligation of this vessel and compression by the prosthetic valve must be considered during AVR [11]. In the present case, we prevented obstructing the right coronary artery, which arose adjacent to the aortic annulus, by using a small prosthesis, placing the annular sutures with the pledgets on the ventricular side, and utilizing sutures that transitioned to the subannular ventricular myocardium at the area closest to the right coronary ostium. We did not select the annular stitches placed from outside the aortic valve annulus because dissecting the Valsalva sinus and placing stitches under the origin of the right coronary artery was associated with an increased risk of injury of the right coronary ostium.

Suturing to the subannular ventricular myocardium is associated with a risk of causing a complete atrioventricular block, particularly in a patient with abnormal aortic valve development, as the anatomical relationship between the aortic annulus and the membranous septum (MS) is different. In fact, Pirundini et al. reported that complete atrioventricular block occurred after AVR for QAV, which had a small accessory cusp between the right and non-coronary cusp [12]. They also reported another case in which they avoided injury to the conduction system by suturing to a supra-annular position in the accessory cusp area. In these cases, the MS was meant to be located beneath the area of the small accessory cusp. Unlike their cases, however, the present case showed an almost equal-sized QAV with a normal course of the coronary system. The MS is generally connected to the right fibrous trigone; therefore, it was assumed that the MS was located on the right of the anterior mitral leaflet and beneath the area close to the nadir of one of the two non-coronary cusps. Thus, the subannular valve sutures around the commissure between the right coronary cusp and one of the two non-coronary cusps were effective in protecting the right coronary ostium, while avoiding complete atrioventricular block.

When coronary ostial occlusion is inevitable, even if every possible measure is taken, the addition of coronary artery bypass grafting should be considered.

Conclusions

We encountered a case of AVR performed for QAV associated with a right coronary ostium anomaly. Careful selection of the size of the prosthesis and modification of the annular stitches are essential to prevent obstruction of the coronary ostium.

Abbreviations

QAV: Quadricuspid aortic valve; AR: aortic regurgitation; AVR: aortic valve replacement; ECG: Electrocardiography; MS: membranous septum

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Informed consent to publish was obtained from the patient presented in this article.

Availability of data and materials

The data are not available for public access due to patient privacy concerns but are available from the corresponding author upon reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

ST, SS, YI and HY performed the surgery. ST and SS were major contributors in writing the manuscript. All authors read and approved the final manuscript.

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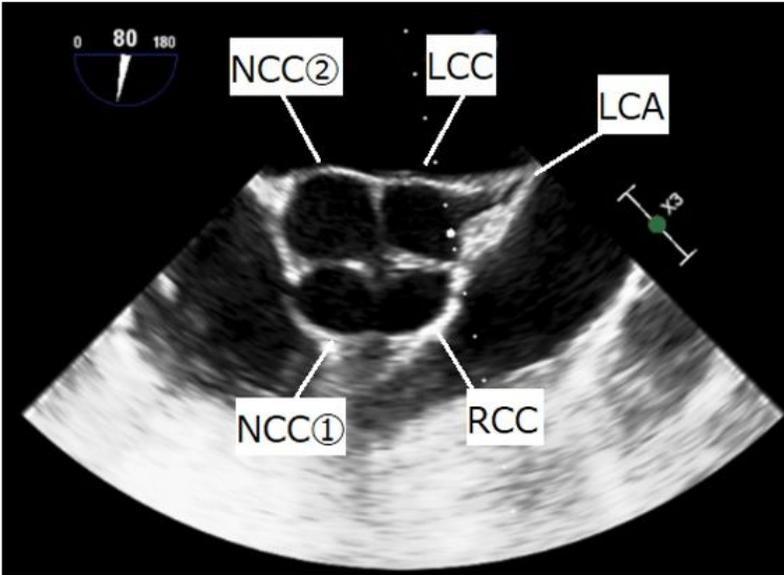
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Figures

a.



b.

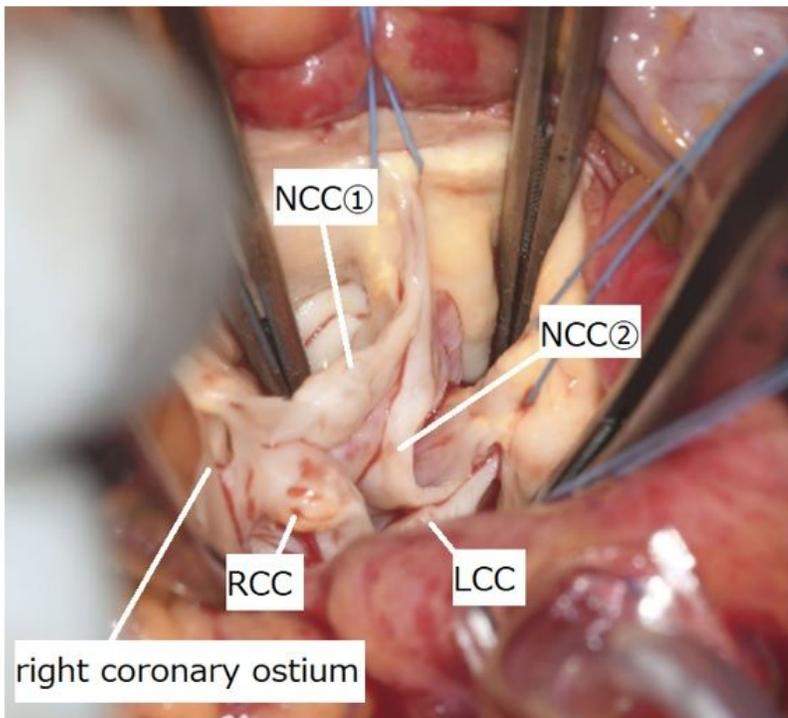


Figure 1

a. Intraoperative transesophageal echocardiography showing almost equal-sized QAV (Hurwitz and Roberts classification type A). b. Intraoperative photograph showing the QAV. The right coronary ostium is located adjacent to the commissure between the right coronary cusp and one of the two non-coronary cusps. LCA, left coronary artery; LCC, left coronary cusp; RCC, right coronary cusp; NCC, non-coronary cusp; QAV, quadricuspid aortic valve.

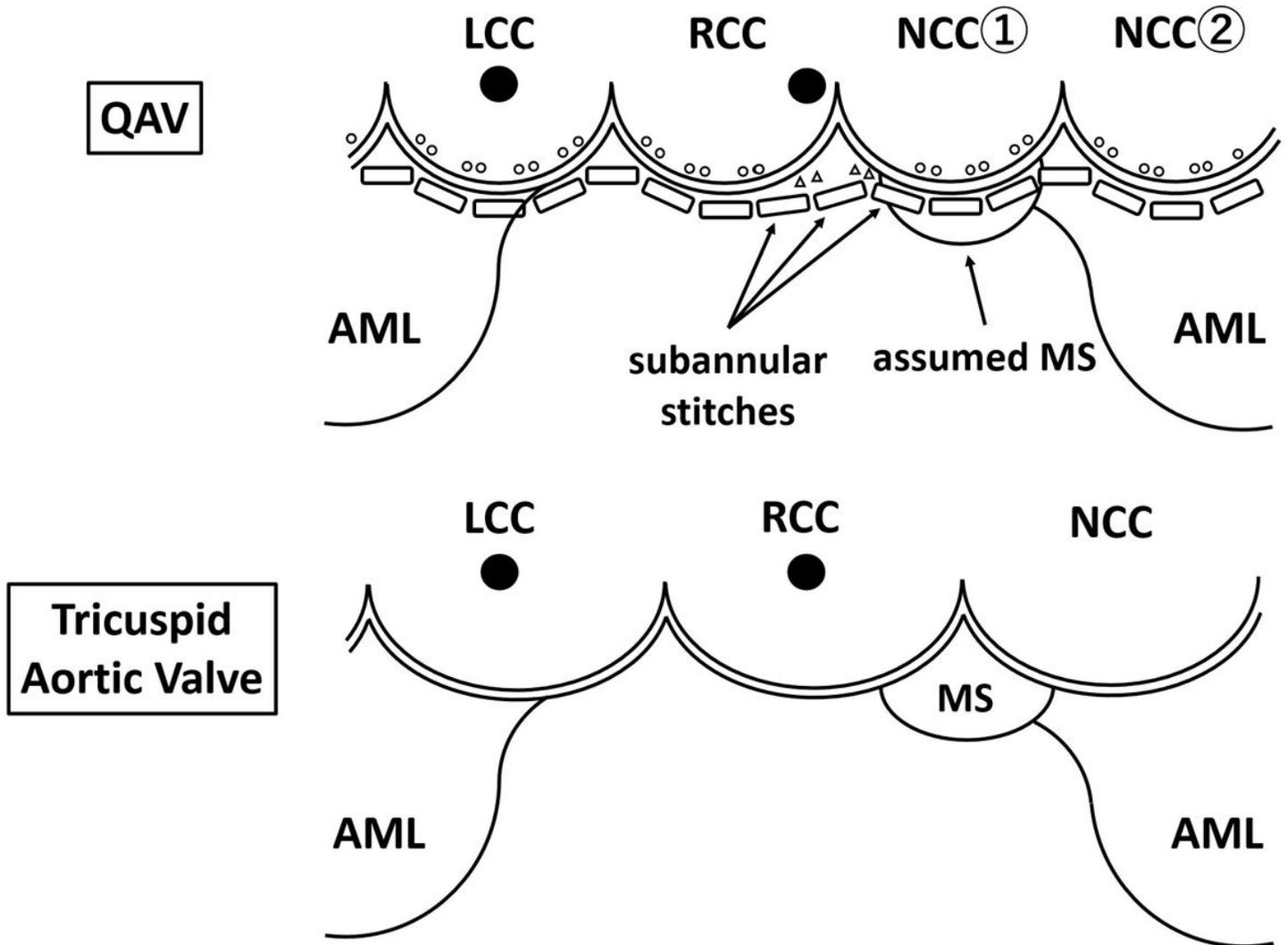


Figure 2

Operative schema of QAV in comparison with tricuspid aortic valve. The annular sutures are placed with pledgets on the ventricular side (circular symbols show exit points of stitches in a non-everting mattress fashion) and three stitches near the right coronary ostium are transitioned to the subannular ventricular myocardium (triangular symbols show exit points of stitches near the right coronary ostium). QAV, quadricuspid aortic valve; LCC, left coronary cusp; RCC, right coronary cusp; NCC, non-coronary cusp; AML, anterior mitral leaflet; MS, membranous septum

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