Aortic root replacement with a valve-sparing technique for quadricuspid aortic valve†

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Abstract

A 67-year old man with ascending aortic aneurysm was referred because of a quadricuspid aortic valve. He underwent aortic root replacement with a valve-sparing technique. Under deep hypothermic circulatory arrest, replacement of the ascending aorta was successfully performed. The postoperative course was uneventful without recurrence of aortic regurgitation.

Keywords: Aortic root replacement • Valve sparing • Quadricuspid aortic valve • Annuloaortic ectasia

INTRODUCTION

Quadricuspid aortic valve is a rare congenital cardiac anomaly, and most patients with this malformation require prosthetic valve replacement. We present a patient who successfully underwent aortic valve-sparing root replacement.

CASE

A 67-year old man was referred because of thoracic aortic aneurysms. He was asymptomatic. Chest radiography showed a cardiothoracic ratio of 54%. Chest computed tomography demonstrated an ascending aortic aneurysm with a diameter of 55 mm. Transthoracic echocardiography showed severe aortic regurgitation (regurgitation volume, 91 ml; regurgitant fraction, 53%) with annuloaortic ectasia and the enlarged left ventricle (left ventricular end-diastolic dimension 64 mm, end-systolic dimension 48 mm and ejection fraction 45%) (Fig.1A). Transoesophageal echocardiography revealed the aortic valve was quadricuspid. Diameter of aortoventricular junction was 27 mm, sinus of Valsalva was 46 mm and sinotubular junction was 37 mm. Coronary angiography was normal.

He underwent elective aortic root replacement with a valve-sparing technique and replacement of the ascending aorta. Cardiopulmonary bypass was established with ascending aorta and bicaval cannulations. The aortic valve consisted of two equal-sized larger cusps (right coronary and left coronary cusp) and two equal-sized smaller cusps [non-coronary cusp 1 (NCC1) and non-coronary cusp 2 (NCC2)]. Dimensions of the right coronary cusp were 36 mm in free margin, 22 mm in geometric height and 10 mm in effective height. Dimensions of the left coronary cusp were 36 mm in free margin, 23 mm in geometric height and 12 mm in effective height. Dimensions of the NCC1 were 24 mm in free margin, 25 mm in geometric height and 12 mm in effective height. Dimensions of the NCC2 were 20 mm in free margin, 25 mm in geometric height and 12 mm in effective height.

The rudimentary commissure between NCC1 and NCC2 was found. The left coronary cusp contained a large fenestration. A 0.6% glutaraldehyde-treated autologous pericardial patch (10 × 5 mm) was applied to the fenestration (Fig.2A). The quadricuspid aortic valve was converted into a tricuspid valve by obliterating the rudimentary commissure between NCC1 and NCC2. The aortic root was replaced with a 30-mm Valsalva graft (Terumo Medical, Somerset, NJ, USA) with 12 reinforced 3–0 polyester (Nespoflene®) mattress sutures in the first row and continuous 5–0 polypropylene (Nespolene®) sutures as the second row anastomosis (Fig.2B). After completion of the right and left coronary anastomoses, replacement of the ascending aorta was performed using a 30-mm woven Dacron Gelatin impregnated graft under deep hypothermic circulatory arrest. The postoperative course was uneventful, and the patient is doing well 6 months after surgery without recurrence of aortic regurgitation (Fig.1B).

DISCUSSION

Quadricuspid aortic valve is a rare congenital anomaly. Aortic regurgitation is often observed in adulthood as a result of fibrous thickening with incomplete coaptation. Only a few cases of quadricuspid aortic valve with annuloaortic ectasia have been reported.

Embryologically, the quadricuspid aortic valve results from the aberrant fusion of the aortopulmonary septum or to abnormal development of the aortic sinuses.
Figure 1: Transoesophageal echocardiography. (A) Preoperative transoesophageal echocardiography. (B) Postoperative transoesophageal echocardiography trivial aortic regurgitation. LCC: left coronary cusp; RCC: right coronary cusp; NCC1: non-coronary cusp 1; NCC2: non-coronary cusp 2.

Figure 2: Operative schema. (A) Closure of the fenestration with a 0.6% glutaraldehyde-treated autologous pericardial patch. (B) Aortic root replacement with a valve-sparing technique and tricuspidization.
proliferations in the common trunk during the fourth week of gestation. Hurwitz and Roberts classified the quadricuspid valve into seven types (Type A to Type G) based on the relative size of the four cusps [2]. They reported that the most common type consisted of three equal large and one smaller cusps; which was Type B, and the next most common was Type C [2]. The patient in our case was classified as Type C, which was two equally large and two equally small cusps.

The gold standard surgical procedure for annuloaortic ectasia is aortic root replacement using a composite valve graft with a mechanical or bioprosthetic valve. Valve-sparing root replacement with the reimplantation technique provides excellent haemodynamics and freedom from anticoagulation therapy. This surgical procedure has been an attractive option particularly for young patients. The indication of this procedure for annuloaortic ectasia was initially limited to patients with morphologically unimpaired valve cusps. More recently, the addition of repairing the abnormal cusps has provided the expansion of indication for valve-sparing aortic root replacement for annuloaortic ectasia.

Patients with a quadricuspid aortic valve generally have a mechanical or biological valve implanted. Ross procedure [3], tricuspidization [4, 5] and bicuspidization [6] without root replacement for patients with a quadricuspid aortic valve have been reported as alternative surgical procedures. Considering the age, Bentall operation with a bioprosthetic valve may have been an appropriate option in our case. However, we selected aortic root replacement with a valve-sparing technique because prosthetic valve degeneration may occur and transcatheter aortic valve replacement is now available. Long-term follow-up is required because durability of our surgical procedure is unknown.

**CONCLUSIONS**

We presented a case of severe aortic regurgitation with a quadricuspid aortic valve and annuloaortic ectasia who successfully underwent aortic root replacement with a valve-sparing technique.

**Conflict of interest:** none declared.

**REFERENCES**