

# Reoperation for ascending aorta aneurysm after double valve replacement in a Takayasu's aortitis patient: a case report

Xiaoping Fan<sup>1\*†</sup>, Jie He<sup>1†</sup>, Jihai Peng<sup>2†</sup>, and Jian Zhuang<sup>1\*</sup>

<sup>1</sup>Department of Cardiovascular Surgery, Guangdong Cardiovascular Institute, Guangdong Provincial Key Laboratory of South China Structural Heart Disease, Guangdong Provincial Key Laboratory of Coronary Heart Disease Prevention, Guangdong General Hospital, Guangdong Academy of Medical Sciences, 106th Zhongshan Er Road, Yuexiu District, 510080 Guangzhou, China; and <sup>2</sup>Department of Rehabilitation, Guangdong General Hospital, Guangdong Academy of Medical Sciences, 106th Zhongshan Er Road, Yuexiu District, 510080 Guangzhou, China

Received 26 July 2017; accepted 2 November 2017; online publish-ahead-of-print 21 December 2017

## Abstract

Takayasu's aortitis (TA) is a complicated disease. Surgical treatment, especially reoperation, can be difficult. Here, we report a case of reoperation for TA, which presented with three major complications (aortic aneurysm, valve detachment, and fistula) 3 years after surgical treatment. During the surgery, the aortic valve was reconsolidated and fixed to the mitral valve, the fistula was then repaired, and the aortic root was replaced with woven graft. Following an uncomplicated postoperative event, the patient was discharged at 24 days postoperatively. Follow-up echocardiography at 2 years showed no perivalvular leakage. In such reoperative cases, when double valve replacement is required, it may be better to proactively reconstruct the structure of the central fibrous body.

## Keywords

Reoperation • Aorta aneurysm • Takayasu's aortitis • Case report

## Learning point

- Reconstructing the central fibrous body and reducing the relative movement of the aortic valve may benefit Takayasu's aortitis patients requiring valve replacement.

## Introduction

Takayasu's aortitis (TA) is a chronic systemic vasculitis of unknown origin. It often involves the aorta, its major branches, and the aortic valve. Diagnosis of the disease is mainly based on pathological findings.<sup>1</sup> Surgical treatment such as valve replacement is occasionally required when cardiovascular symptoms occur. The most serious complications after valve replacement in TA are prosthetic valve detachment<sup>2</sup> and aortic root aneurysm.<sup>3</sup> Here, we present a surgical case of TA with valve detachment, aortic root aneurysm, and aorta–

left ventricular and aorta–left atrium fistula after previous double valve replacement (DVR).

## Case

This article was approved by the ethics committee of Guangdong General Hospital, and written informed consent was obtained from the patient.

A 29-year-old male patient was admitted for tachypnoea and exertional palpitations on 21 October 2014. He had previously undergone DVR with an ATS aortic mechanical valve of size 25 mm and an ATS mitral mechanical valve for aortic and mitral regurgitation of size 29 mm on 4 January 2011. At that time, the aortic root was slightly dilated to a diameter of about 35 mm. After operation, the patient was diagnosed with TA based on intraoperative observation and postoperative pathologic examination (*Figure 1*). The postoperative course was uneventful, and the patient was asymptomatic for 3 years. Before the initial operation, the patient had no previous hospitalizations and

\* Corresponding authors. Xiaoping Fan, Tel: +86 138 2503 2279, Fax: 020-83827812, Email: [fukui-hanson@hotmail.com](mailto:fukui-hanson@hotmail.com); Jian Zhuang, Tel: +8613602884548, Email: [jianzhuang@yahoo.com](mailto:jianzhuang@yahoo.com). This case report was reviewed by Dejan Milasinovic and Philipp Sommer.

† The first three authors contributed equally to the study.

© The Author 2017. Published by Oxford University Press on behalf of the European Society of Cardiology.

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact [journals.permissions@oup.com](mailto:journals.permissions@oup.com)

history of medical complications. He did not take any oral corticosteroids, and C-reactive protein (CRP) was not monitored. Echocardiography was undertaken once every 6 months. The patient unexpectedly experienced tachypnoea and exertional palpitations 3 years after the procedure. Physical examination revealed systolic murmurs at the cardiac apex. Heart rate was 80 b.p.m., respiratory rate was 20 breaths/min, and blood pressure was 121/51 mmHg. A crisp metallic A<sub>2</sub> and loud P<sub>2</sub> was noted during auscultation. Breaths sounded clear, and no rale was noted. Echocardiography showed large movement of the mechanical aortic valve (Figure 2A). Thus, the patient was readmitted to the hospital for further examination. Laboratory examination showed a white blood cell count of  $7.8 \times 10^9/L$  (with standard range of  $4\text{--}10 \times 10^9/L$ ), erythrocyte sedimentation rate of 28 mm/h, and CRP of 23.3 mg/L (with standard range of 0–15 mm/h and 0.068–8.2 mg/L respectively). Other laboratory results were within normal limits. Transoesophageal echocardiography showed severe detachment of the aortic valve and fistula from the aortic root to both the left ventricle and the left atrium (Figure 2B). Computed tomography showed aneurysmal dilation of the aortic root to a diameter of 64 mm (Figure 3A and B). After evaluation, we diagnosed this patient with (i) TA, (ii) aortic aneurysm, (iii) aortic valve detachment, (iv) aortic fistula, and (v) previous DVR.

## Timeline

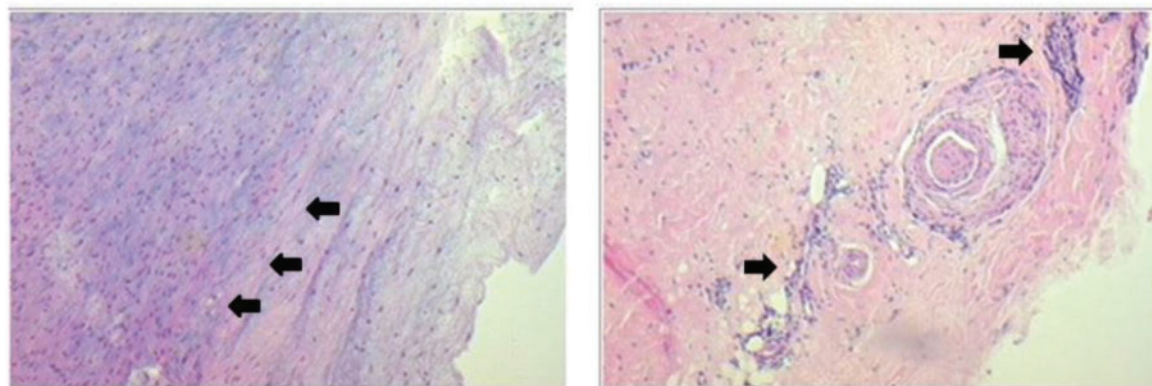
| Time       | Event  |
|------------|--|
| 4/1/2011   | First surgery-DVR, and TA was diagnosed                  |
| 1/5/2014   | Polypnea and exertional palpitations occurred            |
| 9/6/2014   | Echocardiography detected detachment of the aortic valve |
| 28/8/2014  | CT found aortic root dilation                            |
| 5/11/2014  | Reoperation  |
| 29/11/2014 | Discharge  |
| 25/5/2017  | Lastest follow-up echocardiography                       |

On 5 November 2014, we reoperated through a median sternotomy. Cardiopulmonary bypass (CPB) was established through the femoral artery and axillary artery. The aneurysm was resected close to the annulus. The mechanical aortic valve was almost completely detached; however, the valve leaflet was intact. Thus, we reconsolidated the aortic valve to the annulus using continuous 4-0 Prolene suture and repaired the fistula of the aorta–left ventricular and aorta–left atrium with a patch of artificial pericardium. A woven graft of size 28 mm (MAQUET) was implanted and the coronary arteries were anastomosed to the graft. When we tried to take the patient off CPB, unexpected and severe bleeding occurred in the margin of the repaired fistula. After restarting CPB, we re-exposed the fistula by opening the patch that was placed over the fistula of the aorta–left atrium. We then fixed the aortic valve to the mitral valve using 3-0 Prolene through the fistula followed by repair and reinforcement of the patches using 4-0 Prolene. The aortic root–right atrial shunt was performed by covering the woven graft with the remaining aortic wall and making a shunt to the right atrium. Finally, the patient was weaned from CPB. Aortic cross-clamp time was 373 min, and CPB time was 581 min.

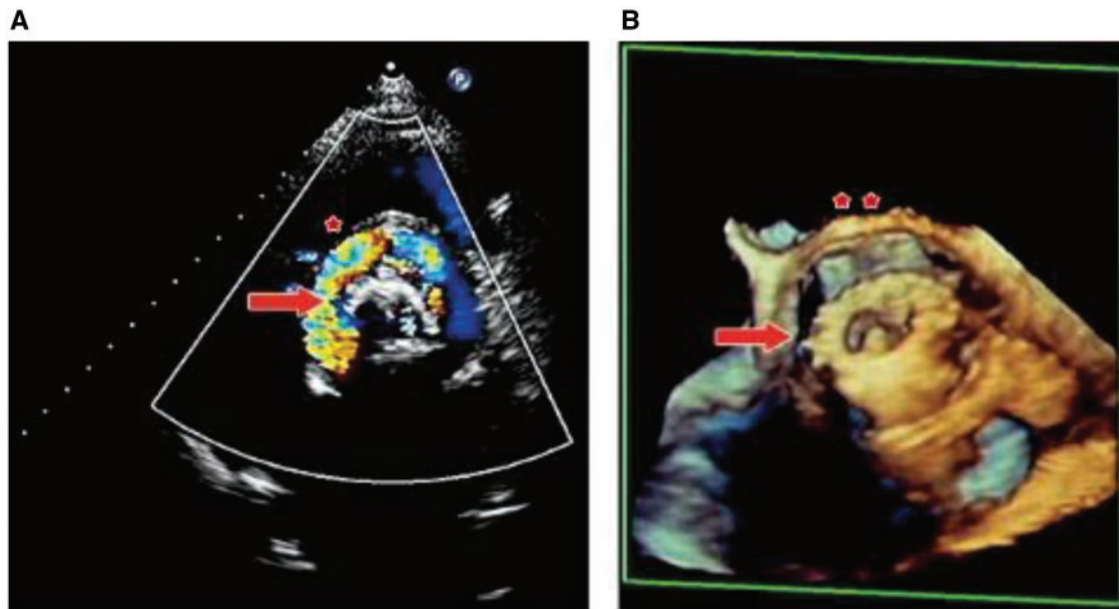
The postoperative course was uneventful. Postoperative haemostatics, vasoactive agent (dopamine, epinephrine, etc.), steroids, and antibiotics were routinely used. After a hospital stay of 24 days, the patient was discharged. Echocardiography before discharge showed only slight malfunction of the medial leaflet of the mitral valve and no perivalvular leakage. The patient took oral corticosteroids (from 30 mg/day down to 10 mg/day) for 6 months postoperatively. Follow-up echocardiography at 3 months, 1 year, and 2 years showed only moderate tricuspid regurgitation and the medial leaflet malfunction (Figure 4).

## Discussion

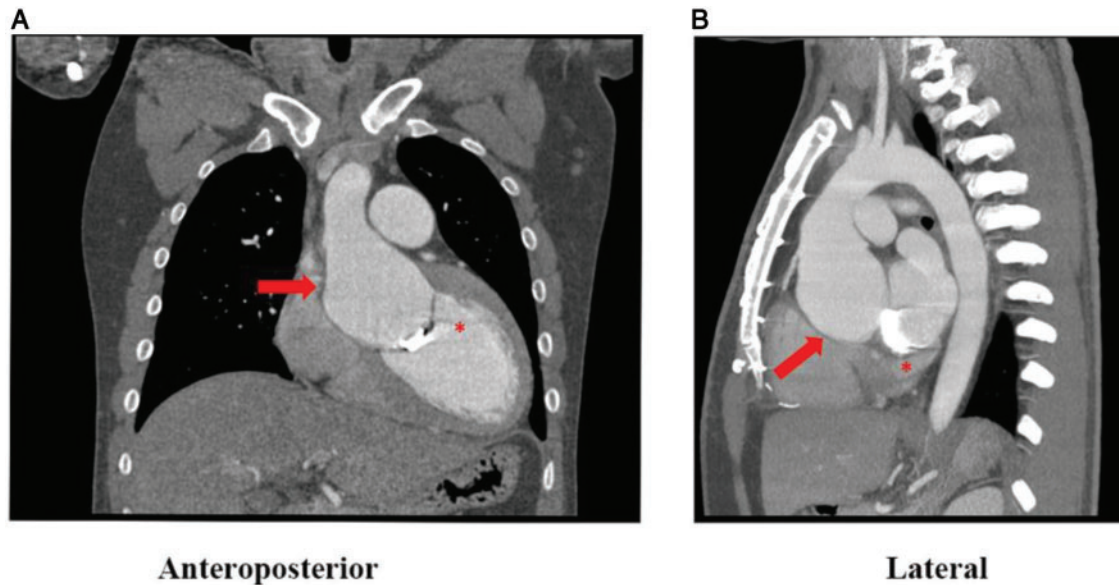
Takayasu's aortitis is an autoimmune vasculitis affecting medium and large arteries. Surgical intervention is required in some cases,<sup>4</sup> and good surgical outcome has been obtained.<sup>5</sup> However, postoperative complications such as perivalvular leakage, valve



**Figure 1** Pathological sections taken from the aortic wall showed irregular collagen proliferation, hyaline and mucinous degeneration, and inflammatory cell infiltration (arrows).



**Figure 2** Perioperative echocardiography (A) and transoesophageal echocardiography (B). (A) Echocardiography detected large movement of the aortic valve (arrow) and perivalvular leakage (\*). (B) Transoesophageal echocardiography indicated loose aortic valve (arrow) movement and aortic annulus (\*\*).

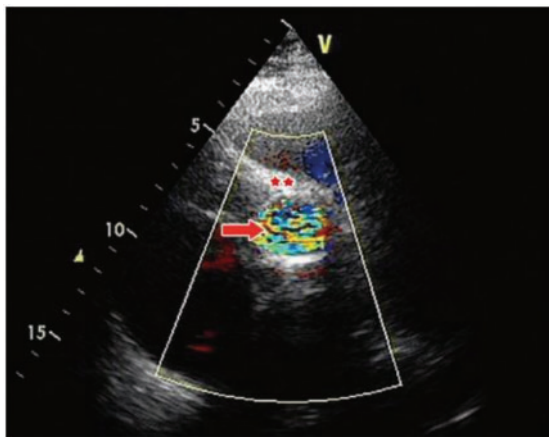


**Figure 3** Computed tomography scan showing enlarged aortic root (arrow) and sign of avulsion of the aortic valve (\*).

detachment, and aneurysm formation are relatively common because of extensive and severe lesions as well as fragile and inflamed tissue encountered during the procedure.<sup>6,7</sup> Reoperations for valve detachments and aneurysm formation have been reported.<sup>8</sup> In addition, fistulation,<sup>9</sup> a rare complication, has also been documented in TA. We have presented a case of reoperation

for TA that presented with all of these complications, making it a notable and complex procedure.

In this case, the patient refused to take steroid after the first surgery, which might be a factor contributing to the disease progression. Thus, we instructed the patient to take steroid for 6 months after he was discharged (the second time) just for safety. However, there is a



**Figure 4** Postoperative echocardiography showed no movement of the aortic valve (arrow) and no perivalvular leakage around the aortic annulus (\*\*).

report<sup>10</sup> saying the benefit of the use of steroid in TA patient is uncertain and that the follow-up examination should be taken rigorously.

Initially, we planned to reconsolidate the detached aortic valve, repair the fistula, and replace the aortic root. However, during the operation, we observed the valve leaflet was intact. Under these circumstances, replacing the detached prosthetic valve with a new one might increase the chance of recurrence detachment.<sup>11</sup> Thus, the decision was made to reconsolidate the detached aortic valve. After placement of the patches, we tried to take the patient off CPB, but severe bleeding occurred in the previous aorta–left ventricle and aorta–left atrium fistula. We believe that the cause of this bleeding might be attributed by the relative movement of the mitral valve and the aortic valve during heartbeats damaging the tissue. Thus, CPB was restarted, and 3-0 Prolene suture was used to fix the aortic valve to the mitral valve, reconstructing the stability of the central fibrous body followed by reinforcement of the patches. Because of the extended operation and CPB time, the coagulation system was badly compromised, and there was continuous haemorrhage on the anastomotic sites. An aortic root–right atrial shunt was performed to drain the blood and reduce the pressure of the perigraft space. The patient was then successfully weaned off CPB. In a TA patient, cardiac tissue, especially the central fibrous body, is already compromised by the disease. Therefore, it is very important to reconstruct the central fibrous body. Kim *et al.*<sup>12</sup> also reported their experience of reconstructing the fibrous body with promising results, but with some differences. In Kim *et al.*'s series, bovine pericardial patch was used to reconstruct the fibrous body, which helps lower the mortality and prevent recurrence of the disease, such as infectious endocarditis; in this case, we directly fix the aortic valve to the mitral valve, including the central fibrous body, to reconstruct the fibrous body, with the purpose of reducing the relative movement of the aortic valve and preventing avulsion and preavalvular leakage. We used a slightly larger patch during this procedure to reduce the suture line tension when repairing the fistulation.

Postoperative treatment was uneventful. The echocardiography before discharge showed only slight malfunction of the mitral valve medial leaflet, which could be caused by the suture between the aortic valve and the mitral valve. Echocardiography 2 years after the surgery showed only moderate tricuspid regurgitation and the medial leaflet malfunction, which is the result of the aortic root–right atrial shunt and reconstruction of the central fibrous body. Although a report by Rosero *et al.*<sup>13</sup> described a case of long-term shunt patency with detrimental consequences. In our case, the tricuspid regurgitation is moderate (about 1.2 cm<sup>2</sup>), and the patient has not developed any symptoms. We believe that thorough haemostasis in the surgery and precise management of the size of the shunt is very important. Takayasu's aortitis is a progressive disease, and the diagnosis of disease activity is based on clinical and imaging features. In this case, the patient developed symptoms and echocardiography detected an abnormality just 6 months after the follow-up. Therefore, more rigorous follow-up should be employed in TA patients. Surgical intervention should be performed in a timely manner when clinical symptoms develop or severe imaging abnormality is detected.

Similar reoperations in TA patients have been reported.<sup>8,14,15</sup> However, to the best of our knowledge, this is the first report of an aorta–left ventricle and aorta–left atrium fistula combined aneurysm and prosthetic valve detachment in a TA patient.

Because of the retrospective nature of this case report and, in particular, the long follow-up of nearly 3 years, it is difficult to obtain valuable patient perspective. In addition, we believe the most valuable information from this case lies in the treatment itself, particularly in the emergency treatment and planning that occurred intraoperatively.

## Conclusion

We successfully treated a case of aortic aneurysm combined with aortic valve detachment and aortic fistula after previous DVR in TA patient. The teaching points of this case were: (i) the two key procedures in this operation, i.e. reconstruction of the central fibrous body and the shunt of aortic root–right atrial shunt; (ii) because of the fragility of the tissue in TA patients, the surgical operation must be meticulous to ensure as little tension on the suture line as possible; (iii) for DVR operations in TA patients, proactively fixing the aortic valve to the mitral valve might be beneficial; (iv) imaging follow-up should be performed on a more regular basis in TA patients; and (v) even in such a challenging and complex case, surgery is still the feasible and the preferred option.

## Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

## Acknowledgements

We thank Guangdong Cardiovascular Institute and its wonderful surgical team for the support. We also thank Lindsey Hamblin for helping us edit the paper.



## Funding

Natural Science Foundation of Guangdong Province China (grant number 2016A030313792), Medical Science Research Foundation of Guangdong Province China (grant number 2016115114137325), and Chinese Medicine Research Foundation of Guangdong Province China (grant number 20161003).

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

**Author Contributions:** X.F. was the attending doctor in charge of surgery and the treatment process, the leading surgeon in the operation and was also involved in supervision and revising the article. J.H. was the primary author for writing and revising this article. J.Z. was the senior consultant who helped in planning the therapy and assisted in the operation. J.P. was the co-author of this article who helped collecting data and editing the article. X.F. and J.Z. holds the overall responsibility.

## References

- Caspary L. Inflammatory diseases of the aorta. *VASA* 2016;**45**:17–29.
- Bougioukas I, Mikroulis D, Popov AF, Bougioukas G. Re-do aortic operation in a young patient for aggressive Takayasu's arteritis. *J Cardiothorac Surg* 2012;**7**:91.
- Miyata T. The Asia Pacific meeting for vasculitis and ANCA workshop 2012: surgical treatment for Takayasu's arteritis. *Clin Exp Nephrol* 2014;**18**:296.
- Ishikawa K, Maetani S. Long-term outcome for 120 Japanese patients with Takayasu's disease. Clinical and statistical analyses of related prognostic factors. *Circulation* 1994;**90**:1855–1860.
- Kaku Y, Aomi S, Tomioka H, Yamazaki K. Surgery for aortic regurgitation and aortic root dilatation in Takayasu arteritis. *Asian Cardiovasc Thorac Ann* 2015;**23**:901–906.
- Ando M, Kosakai Y, Okita Y, Nakano K, Kitamura S. Surgical treatment for aortic regurgitation caused by Takayasu's arteritis. *J Card Surg* 2010;**13**:202–207.
- Miyata T, Sato O, Deguchi J-O, Kimura H, Namba T, Kondo K, Makuuchi M, Hamada C, Takagi A, Tada Y. Anastomotic aneurysms after surgical treatment of Takayasu's arteritis: a 40-year experience. *J Vasc Surg* 1998;**27**:438–445.
- Adachi O, Saiki Y, Akasaka J, Oda K, Iguchi A, Tabayashi K. Surgical management of aortic regurgitation associated with takayasu arteritis and other forms of aortitis. *Ann Thorac Surg* **84**:1950–1953.
- Miyata T, Ohara N, Shigematsu H, Konishi T, Yamaguchi H, Kazama S, Ohshiro H, Kawaguchi S, Ishimaru S. Endovascular stent graft repair of aortopulmonary fistula. *J Vasc Surg* 1999;**29**:557–560.
- Ham SW, Kumar SR, Rowe VL, Weaver FA. Disease progression after initial surgical intervention for Takayasu arteritis. *J Vasc Surg* 2011;**54**:1345–1351.
- Kawazoe K, Eishi K, Kawashima Y. New modified Bentall procedure: carrel patch and inclusion technique. *Ann Thorac Surg* 1993;**55**:1578–1579.
- Kim SW, Park PW, Kim WS, Sung K, Lee YT, Jun T-G, Jeong DS. Long-term results of aortomitral fibrous body reconstruction with double-valve replacement. *Ann Thorac Surg* 2013;**95**:635–641.
- Rosero H, Nathan PE, Rodney E, Vasavada B, Sacchi TJ. Aorta to right atrium fistula with congestive heart failure resulting from a patent Cabrol shunt after repair of aortic dissection. *Am Heart J* 1994;**128**:608–609.
- Watanabe Y, Matsushita S, Okawa S, Yamabuki K, Gomi S, Hiyama T. Aortic root replacement for prosthetic aortic valve detachment without regurgitation and with enlarged Valsalva's sinuses and complete atrioventricular block caused by Takayasu's aortitis. *Jpn J Thorac Cardiovasc Surg* 2003;**51**:201–204.
- Minami H, Wakita N, Kawanishi Y, Kitano I, Sakata M, Shida T. Aortic aneurysm and aortic regurgitation following aortic valve replacement due to Takayasu's arteritis. *Jpn J Thorac Cardiovasc Surg* 2002;**50**:43–45.