Delayed aneurysmal complication of bicuspid aortic valve disease after heart transplantation

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Central Message

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Video clip is available online.

CLINICAL SUMMARY

A 72-year-old man with bicuspid aortic valve (BAV) disease underwent aortic valve replacement (AVR) in 1993, and subsequently developed cardiomyopathy secondary to sarcoidosis for which he underwent orthotopic heart transplantation in 1995. Posttransplantation, he developed mild hypertension in the range of 130 to 140 mm Hg (systolic), but otherwise was doing well clinically and undergoing routine echocardiographic surveillance. In late December 2017, he presented to an outside hospital with chest discomfort. Echocardiography at that time showed normal function of the transplanted heart and an ascending aortic diameter of 3.6 cm. As part of a workup for presumed pneumonia, he underwent a computed tomography (CT) scan of the chest, which incidentally showed a distal ascending aortic/arch aneurysm in the native aorta with maximal diameter of 7.2 cm.

It is difficult to establish the temporal progression of the aneurysm based on echocardiography reports alone. For example, in 2015 the mid ascending aorta was “mildly dilated,” and a subsequent report in 2016 failed to mention the ascending aorta at all, demonstrating the limitations of echocardiography as an imaging modality to adequately delineate the distal ascending aorta and arch.

Upon completion of the preoperative evaluation, the patient underwent reoperative cardiac surgery for aneurysm resection. Right common femoral artery and vein were accessed for cardiopulmonary bypass before entering the chest, because the aorta was right underneath the sternum. Intraoperative inspection revealed a chronic

pseudoaneurysm of the native ascending aorta directed posteriorly between the right pulmonary artery and superior vena cava. The donor aorta was nonaneurysmal and not involved in the disease process. After careful dissection, the ascending aorta was freed and a crossclamp was applied. The aneurysm was resected and transverse hemiarch replacement was performed under deep hypothermic circulatory arrest with retrograde cerebral perfusion. Upon completion of the distal anastomosis, cardiopulmonary bypass was resumed, and ascending aortic replacement was performed (Video 1). Follow-up clinic visit at 1 and 6 months showed stable repair by CT imaging (Figure 1).

VIDEO 1. Dr Prashanth Vallabhajosyula describes the operative steps of aneurysmal resection in a patient with delayed bicuspid aortic valve related aneurysm formation after heart transplantation, and discusses the importance of computed tomography scan surveillance of the thoracic aorta in patients with bicuspid aortic valve disease. Video available at: [https://www.jtcvs.org/article/S0022-5223\(19\)30591-4/fulltext](https://www.jtcvs.org/article/S0022-5223(19)30591-4/fulltext).

FIGURE 1. Computed tomography scan of the chest in a patient with a delayed aneurysmal complication of bicuspid aortic valve disease more than 20 years after heart transplantation. A, Preoperative. B, Postoperative.

DISCUSSION

To our knowledge, this is the first reported case in the literature of a recipient native ascending aortic aneurysm in a patient with BAV disease after cardiac transplantation. Other reports have described thoracic aortic aneurysms in the recipients' native aorta unrelated to BAV disease,¹ or complications of the donor aorta associated with BAV disease in the donor heart.^{2,3} Nevertheless, a retrospective review of 243 explanted hearts found that 7 patients had native (recipient) BAV disease, suggesting that the prevalence of BAV disease in the heart transplantation population may be as high as 2.8%.⁴

The etiology of this aneurysm is likely a combination of risk factors related to both BAV disease and heart transplantation. Current theory suggests that ascending aortic dilatation in BAV disease results from hemodynamic and embryologic influences.⁵ Because any hemodynamic abnormalities associated with the morphologically bicuspid valve should have been eliminated at the time of this patient's AVR, this case underscores the importance of the embryologic component of aortopathy in BAV disease. Aneurysm formation is also more prevalent after heart transplantation than in the general population due to factors unrelated to BAV disease.⁶ For example, immunosuppression may directly increase the rate of atherosclerosis and weaken aortic wall tissue. The patient was taking prednisone (5 mg daily), which has been associated with aneurysm expansion in animal models,¹ and tacrolimus (0.5 mg twice per day), which is known to induce hypertension. Moreover, hemodynamic changes related to improved cardiac output after heart transplantation could exacerbate the problem.¹

Although the exact cause for aneurysm formation in this patient may not be known, this case demonstrates the necessity for follow-up thoracic aortic imaging in BAV patients. The 2018 American Association for Thoracic Surgery guidelines for BAV aortopathy recommend regular interval

imaging of the thoracic aorta regardless of baseline diameter to screen for aneurysm formation.⁷ Even in patients with normal aortic dimensions, the thoracic aorta should be imaged at least every 3 to 5 years (if stable).⁷ Furthermore, these recommendations apply to BAV patients even after AVR because of the continued risk of aortic complications.⁷ At our institution, surveillance CT imaging is not a routine part of follow-up care after heart transplantation. Therefore, future studies should assess the need for surveillance imaging of the thoracic aorta in patients after heart transplantation, especially in the presence of BAV disease and other risk factors for aneurysm formation.

CONCLUSIONS

Surgery on the ascending aorta can be done safely with minimal surgical complications many years after cardiac transplantation. Routine surveillance with follow up CT imaging of the thoracic aorta may be warranted in BAV patients after cardiac transplantation.

References

1. Fukuhara S, Stephens EH, Glotzbach JP, Borger MA. Repair of ascending aortic aneurysms following cardiac transplantation. *J Card Surg.* 2016;31:778-80.
2. Stephens EH, Fukuhara S, Neely RC, Takayama H. Aortic root replacement for bicuspid aortopathy following heart transplantation. *J Card Surg.* 2017;32:667-9.
3. van der Merwe J, Casselman F, Verstreken S, Stockman B. Acute ascending aorta dissection in a bicuspid aortic valve cardiac allograft 11 years after orthotopic cardiac transplantation. *Eur J Cardiothorac Surg.* 2017;51:191.
4. Roberts WC, Roberts CC, Ko JM, Hall SA, Capehart JE. Cardiac transplantation in adults with aortic valve disease with focus on the bicuspid aortic valve. *Am J Cardiol.* 2012;109:1212-4.
5. Mordt I, Tzemos N. Bicuspid aortic valve disease: a comprehensive review. *Cardiol Res Pract.* 2012;2012:196037.
6. Muluk SC, Steed DL, Makaroun MS, Pharm S, Kormos RL, Griffith BP, et al. Aortic aneurysm in heart transplant recipients. *J Vasc Surg.* 1995;22:689-94.
7. Borger MA, Fedak PWM, Stephens EH, Gleason TG, Girdauskas E, Ikonomidis JS, et al. The American Association for Thoracic Surgery consensus guidelines on bicuspid aortic valve-related aortopathy: full online-only version. *J Thorac Cardiovasc Surg.* 2018;156:e41-74.