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MINI-FOCUS ISSUE: TAVR

ADVANCED

CASE REPORT: CLINICAL CASE

Transcatheter Aortic Valve Replacement of a Bicuspid Aortic Valve in a Heart Transplant Recipient

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ABSTRACT

Patients with heart transplants who present with severe aortic stenosis may be deemed high-risk surgical candidates due to immunosuppression and multiple comorbid conditions. Appropriately selected patients may be successfully treated with transcatheter aortic valve replacement. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2020;2:716–20)

HISTORY OF PRESENT ILLNESS

A 45-year-old male who had undergone orthotopic heart transplantation in 1997 presented 22 years later with progressive dyspnea on exertion. The patient was known to have a bicuspid aortic valve in his transplanted heart (**Figure 1**, Videos 1 and 2).

On presentation, he was afebrile, his blood pressure was 150/90 mm Hg, his heart rate was 95 beats/min, and his oxygen saturation was 95% on room air. Physical examination was significant for mildly elevated neck veins, clear lungs, 3/6 systolic murmur at the right upper sternal border with radiation to the neck and minimal lower extremity edema.

MEDICAL HISTORY. Medical and surgical history included orthotopic heart transplant in 1997 for nonischemic cardiomyopathy, bicuspid aortic valve in the transplanted heart, hypertension, and hyperlipidemia. Transplantation-associated complications included squamous cell carcinoma of the hard palate treated with resection and grafting and end-stage renal disease (ESRD) on hemodialysis, due to cyclosporine toxicity.

LEARNING OBJECTIVES

- To recognize that recipients with transplanted hearts are subject to a slew of complications which may include common valvular diseases.
- To understand the importance that a multidisciplinary heart team approach plays in management of complex structural heart patients.
- To illustrate the expanding role of transcatheter aortic valve replacement to serve complex patients.

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DIFFERENTIAL DIAGNOSIS

The differential diagnosis for shortness of breath in a patient with prior heart transplant is extensive but includes predominantly transplant rejection, which can be acute cellular rejection, humoral rejection, and coronary artery vasculopathy (CAV) (1,2). Lung infections can also present with shortness of breath, due to either community-acquired pneumonia or opportunistic infections by organisms such as cytomegalovirus, Epstein-Barr virus, *Toxoplasma gondii*, *Aspergillus fumigatus*, or *Candida albicans*. Lymphoproliferative disorder of the lung and lung cancer are also in the differential.

INVESTIGATIONS

Blood work showed normal white blood count and mild anemia. B-type natriuretic peptide was elevated (600 ng/l). Creatinine was 8 mg/dl with normal electrolytes. Chest radiographs showed no lung infiltrates. Electrocardiography showed sinus rhythm, first-degree AV block, right bundle branch block, and left posterior fascicular block (**Figure 2**). An echocardiogram demonstrated preserved left ventricular systolic function with apical hypokinesis and severe aortic stenosis (AS) with a peak velocity across the valve of 4.7 m/s and a mean gradient of 52 mm Hg (**Figures 3 and 4**, Videos 3 and 4). Of note, the transplantation team was aware of progressively worsening AS, documented by both clinical examination and echocardiography. The last echocardiogram before the one showing severe AS was performed a year earlier and had shown moderate to severe AS. During that time, the patient was asymptomatic.

MANAGEMENT

Left heart catheterization revealed an 80% stenosis in the mid portion of the left anterior descending artery, which was treated with one drug-eluting stent. The lesion was focal, consistent with coronary artery disease, and lacked features concerning for CAV, but it was deemed significant enough and was fixed in preparation for valve intervention. Approximately 2 to 3 months after undergoing revascularization, the patient was seen by the valve team in the authors' institution and was evaluated by 2 cardiothoracic surgeons. He was deemed to be at high risk for surgical valve replacement due to prior heart transplantation, ESRD, and immunosuppression with a calculated Society of Thoracic Surgery risk score of 12.9%. A computed tomography angiogram demonstrated adequate common femoral artery diameters for a transfemoral approach to transcatheter aortic valve replacement (TAVR). He underwent successful placement of a balloon-expandable transcatheter valve. Immediately after valve deployment, complete heart block (CHB) was noted, and a transvenous pacemaker was placed at the conclusion of the case (**Central Illustration**). He was monitored in the coronary care unit for 24 to 36 h without atrioventricular conduction recovery. He was evaluated by electrophysiology for potential right axillary/subclavian access, but venography confirmed inadequate venous access. Thus, he received a permanent epicardial pacemaker 48 h after the valve placement, through a left thoracotomy. This was further complicated by a moderately sized hemothorax, thought to be due to significant adhesions, which were dissected to place the leads, requiring chest tube placement. The tube remained in place for 4 days and drained 855 cc of blood. He was discharged home 6 days after the valve procedure in a stable condition. His echocardiogram at discharge revealed normal left

ABBREVIATIONS AND ACRONYMS

AS = aortic stenosis

AV = atrioventricular

CAV = coronary artery vasculopathy

CHB = complete heart block

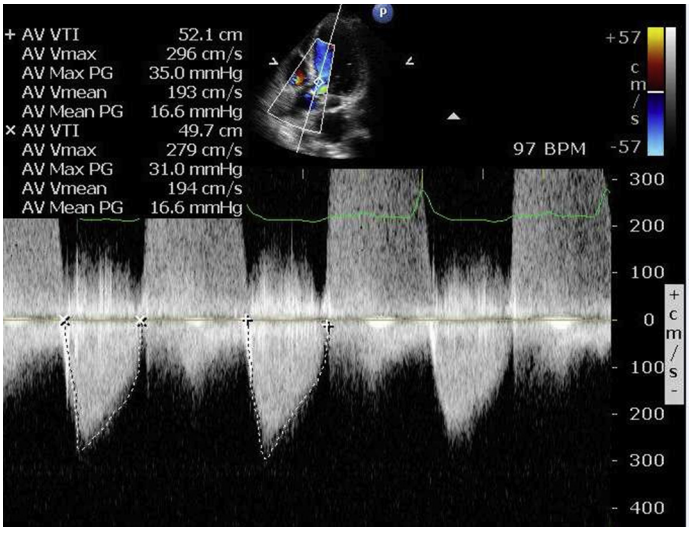
ESRD = end-stage renal disease

TAVR = transcatheter aortic valve replacement

TEE = transesophageal echocardiogram

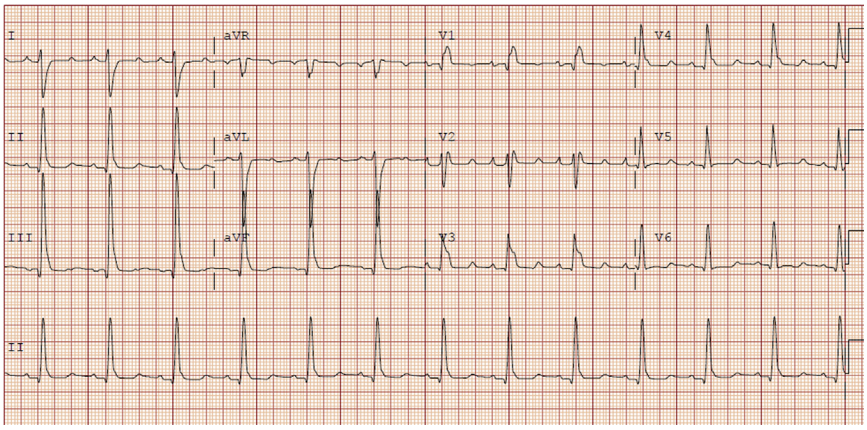
TTE = transthoracic echocardiogram

FIGURE 1 Continuous-Wave Doppler



Continuous-wave Doppler across the aortic valve demonstrates a mean gradient of 16.6 mm Hg, consistent with normal functioning of an aortic valve in 2014. AV VTI = aortic valve velocity time integral (52.1 cm); AV Vmax = aortic valve peak (maximum) velocity (2.9 m/s); AV Max PG = aortic valve peak gradient (35 mm Hg); AV Vmean = aortic valve mean velocity (1.9 m/s); BPM = beats/min.

FIGURE 2 Initial Electrocardiogram Demonstrating First-Degree Atrioventricular Block



Initial electrocardiogram demonstrating first-degree atrioventricular block, right bundle branch block and left posterior fascicular block.

ventricular function and a well-seated prosthesis in the aortic position with a mean gradient of 8 mm Hg and trace paravalvular regurgitation (Video 5).

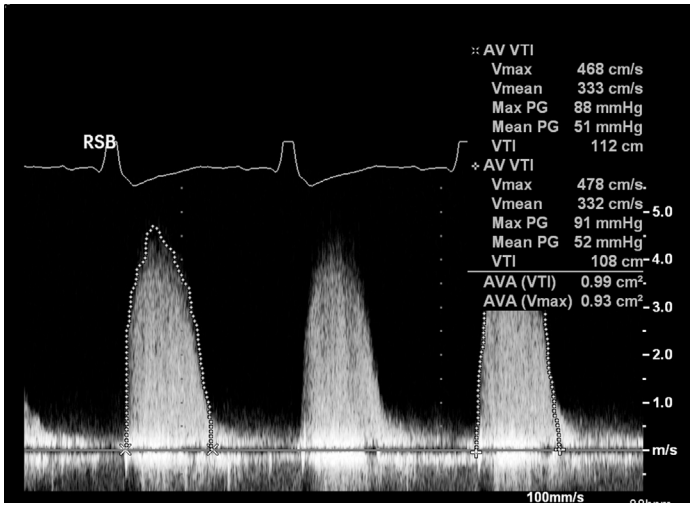
DISCUSSION

For a heart to be considered for transplantation, donors must be younger than 55 years of age, although select donors over the age of 55 years are acceptable for older recipients without a history of chest trauma or cardiac disease, appropriate hemodynamics on minimal or no inotropic support, and a normal or nearly normal echocardiogram results (3). In the absence of stenosis, the presence of a bicuspid valve in the donor does not exclude the heart from transplantation, as the average life span of an adult recipient after heart transplantation is approximately 11 years. Our patient lived 22 years after receiving a heart transplant, and the donor bicuspid valve likely underwent accelerated calcification due to hemodialysis.

TAVR is feasible in patients with bicuspid aortic valve, and research has shown that the overall complication rate is comparable to TAVR in trileaflet valves (4-6). TAVR in bicuspid aortic valve is more prone to paravalvular regurgitation and adverse procedural events, such as aortic rupture, although that was not demonstrated with the newer generation devices. The major difference is the higher propensity for paravalvular leak in patients with bicuspid valve. Cumulative all-cause mortality was found to be similar in these 2 groups, but more research is needed (6).

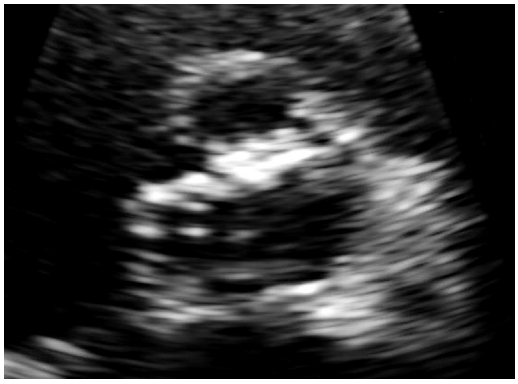
Data with regard to TAVR in patients who have previously received a heart transplant, however, are limited to case reports only (7-10). Four previous cases reported successful TAVR in patients with previous heart transplants, all of whom had a trileaflet aortic valve. These case reports suggest TAVR may

FIGURE 3 Continuous-Wave Doppler Using a Pedoff Probe



Continuous-wave Doppler using a Pedoff probe in the right upper sternal border, revealing a peak velocity of 4.7 m/s and a mean gradient of 52 mm Hg, consistent with severe aortic stenosis. Abbreviations are as in Figure 1.

FIGURE 4 Still Frame of a Short Axis of the Aortic Valve Prior to Transcatheter Aortic Valve Replacement by Transesophageal Echocardiogram

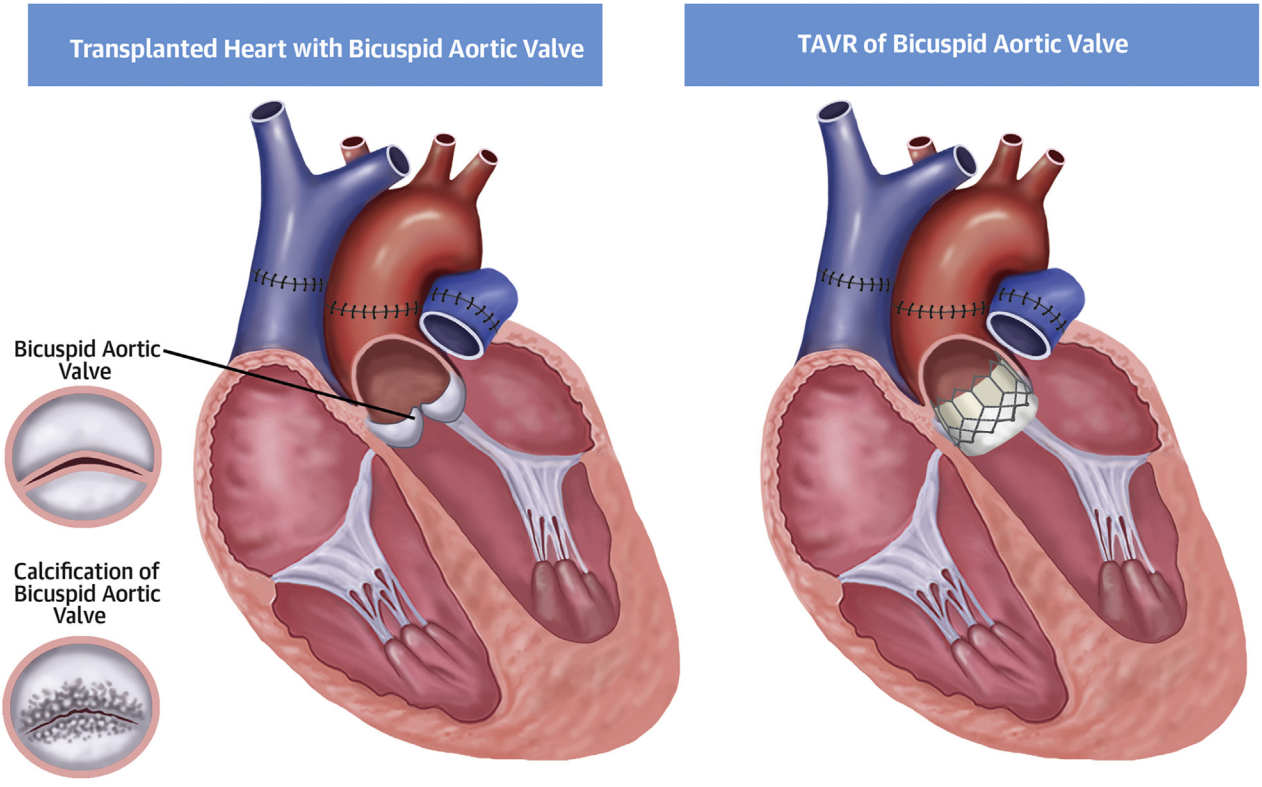


represent a feasible alternative in patients with severe AS in a transplanted heart.

Furthermore, this patient's course was complicated by CHB. Periprocedural pacemaker rates in patients with a bicuspid versus a tricuspid valve have not been well studied. A retrospective study suggested there are no significant differences (6). It is unknown whether TAVR in a transplanted heart bicuspid valve is associated with a higher risk for CHB and need for pacemaker compared to TAVR in native hearts.

FOLLOW-UP. The patient was seen in follow-up at 1 and 6 months, and he was free of symptoms, without diastolic murmur on examination. The patient is scheduled for follow-up echocardiogram at 1 year after the procedure.

CENTRAL ILLUSTRATION TAVR in Bicuspid Aortic Valve of Transplanted Heart



Beale, R. et al. J Am Coll Cardiol Case Rep. 2020;2(5):716-20.

TAVR = transcatheter aortic valve replacement.

CONCLUSIONS

Heart transplant patients are subject to many complications over their lifetimes, including common diseases such as aortic stenosis. TAVR may be a feasible alternative treatment in patients with transplanted hearts who develop severe AS.

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KEY WORDS bicuspid valve, heart transplant, transcatheter aortic valve replacement

APPENDIX For supplemental videos, please see the online version of this paper.