JACC: CASE REPORTS VOL. 3, NO. 17, 2021

STRUCTURAL INTERVENTIONS

INTERMEDIATE

CASE REPORT: CLINICAL CASE

Transcatheter Aortic Valve Replacement for Bicuspid Aortic Insufficiency After Valve-Sparing Aortic Root Replacement

Ofir Koren, MD,[a,b] Vivek Patel, MS,[a] Danon Kaewkes, MD,[a,c] Keita Koseki, MD,[a,d] Tarun Chakravarty, MD,[a] Mamoo Nakamura, MD,[a] Cheng Wen, MD,[a] Robert Naami, MD,[a] Raj R. Makkar, MD[a]

ABSTRACT

Bicuspid aortic insufficiency (BAI) patients with root aneurysm often require aortic valve and root replacement in a composite procedure. The valve-sparing root replacement (VSARR) procedure is aimed at preserving the native valve when possible. This case highlights a successful transcatheter aortic valve replacement procedure in a BAI patient previously treated with VSARR. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:1798–1802)

A 59-year-old woman presented with new progressive fatigue, dyspnea on exertion, and shortness of breath at rest. She was known to have a congenital bicuspid aortic valve (BAV) and to have previously undergone aortic surgery involving the ascending aorta because of an aortic aneurysm.

LEARNING OBJECTIVES

- To be familiar with the epidemiology, pathophysiologic aspects, and natural history of bicuspid aortic valve disease.
- To explore the treatment options for bicuspid aortic insufficiency patients with root aneurysms.
- To discuss the preprocedural dilemmas of matching and sizing a transcatheter heart valve in bicuspid aortic insufficiency patients with root replacement.

MEDICAL HISTORY

The patient's medical history included controlled hypertension, asthma, hyperthyroidism, rheumatoid arthritis, and breast cancer treated by surgical lumpectomy and mediastinal radiation.

In 2014, the patient experienced daily chest pain and effort-related dyspnea. Transthoracic echocardiography (TTE) demonstrated a bicuspid aortic valve and mild to moderate aortic insufficiency (AI). A computed tomographic (CT) angiogram revealed a calcified raphe with a fusion of the left and right coronary cusps and an ascending aortic aneurysm at a 4.5-cm maximal diameter. The patient underwent a valve-sparing aortic root replacement (VSARR), known as a David procedure, with use of a 26-mm prosthetic aortic root graft and a 26-mm prosthetic annular graft (**Figure 1**). The valve was decalcified and partially repaired with a free-edge plication with one suture. TTE after the procedure indicated a remaining

From the [a]Cedars-Sinai Medical Center, Smidt Heart Institute, Los Angeles, California, USA; [b]Bruce Rappaport Faculty of Medicine, Technion Israel Institute of Technology, Haifa, Israel; [c]Department of Medicine, Faculty of Medicine, Khon Kaen University, Khon Kaen, Thailand; and the [d]Department of Cardiology, The University of Tokyo Hospital, Tokyo, Japan.
The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received April 28, 2021; revised manuscript received July 15, 2021, accepted July 26, 2021.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2021.07.018

mild to moderate AI with eccentric and posteriorly directed jet.

DIFFERENTIAL DIAGNOSIS

The following diagnoses were considered: coronary artery disease, progressive valvular dysfunction, arrhythmias, and heart failure.

Coronary angiography indicated a patent coronary artery, and 24-hour Holter monitoring demonstrated extra beats and no arrhythmias. TTE and transesophageal echocardiography demonstrated an aortic valve coaptation defect, a pressure half-time of 196 ms, and holodiastolic flow reversal in the proximal abdominal aorta, indicating a more severe AI (Figure 2). The left and right ventricles were of normal size and function.

MANAGEMENT

A heart team recommended transcatheter aortic valve replacement (TAVR) over redo cardiothoracic surgery, considering the surgical risk, the patient's age, previous sternotomy, previous mediastinal radiation, and the patient's preference.

Cardiac reconstructed CT confirmed a type 1 bicuspid aortic valve and a raphe between fused left and right cusps (Figure 3). The annular measurement indicated a perimeter of 73.2 mm, an area of 403.9 mm^2, and a mean diameter of 22.1 mm. The neocoronary ostia were located 18.0 mm (for the right coronary artery) and 15.7 mm (for the left coronary artery) above the annulus. The left ventricular outflow tract (LVOT) area measured 475 mm^2, and the sinus of the Valsalva area (SOV) was 642 mm^2. The calcium score, measured over the annulus area using an 850 Hounsfield unit cutoff, indicated a small amount (0.1 mm^3) of residual calcium (Figure 4). The right femoral artery was accessible with a minimal lumen diameter of 6.3 × 6.5 mm measured at the right external iliac artery.

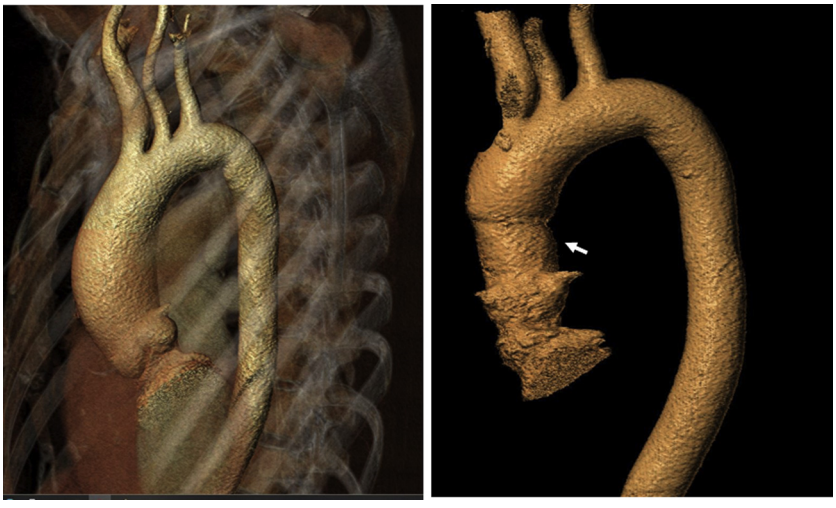
Preprocedural dilemmas were mainly focused on the choice between the 23- or 26-mm balloon expandable Sapien-3 (Edwards Lifesciences) and the 29-mm self-expanding Evolut-PRO+ devices (Medtronic). We considered various parameters such as the actual annular diameter, graft diameter and length, neocoronary ostia height, SOV and LVOT areas, the lack of significant calcification, and the desired effective orifice area to avoid patient-prosthesis mismatch (indexed effective orifice area [EOA] of 0.93 for Sapien-3 and 1.16 for Evolut-PRO+).

Based on the preprocedural CT measurements, both transcatheter heart valves (THVs) were found to be suitable. We decided to use the 26-mm balloon-expandable Sapien-3 THV over a self-expandable THV, mainly considering future coronary access given the patient's age and the lack of calcium distribution over the annulus (1). We successfully deployed the THV over the prosthetic graft, using a

ABBREVIATIONS AND ACRONYMS

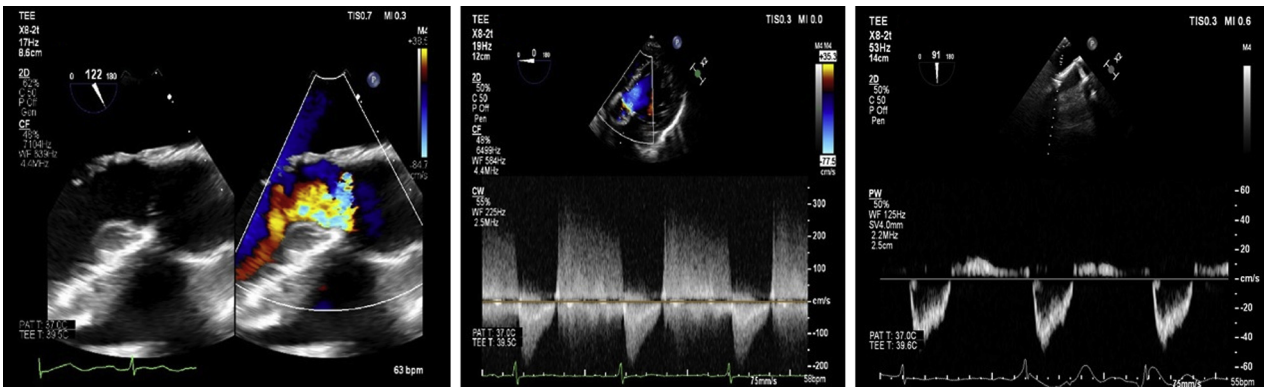
AI = aortic insufficiency
BAI = bicuspid aortic insufficiency
BAV = bicuspid aortic valve
EOA = effective orifice area
LVOT = left ventricular outflow tract
SOV = sinus of Valsalva
TAVR = transcatheter aortic valve replacement
THV = transcatheter heart valve
TTE = transthoracic echocardiography
VSARR = valve-sparing aortic root replacement

FIGURE 1 Computed Tomographic Angiographic View of Ascending Aorta Before and After Valve-Sparing Aortic Root Replacement Surgery



The **white arrow** points to aortic root graft.

FIGURE 2 Significant Aortic Insufficiency Seen on Midesophageal Transesophageal Echocardiographic Views



standard TAVR technique via transfemoral approach (Figure 5).

DISCUSSION

BAV is among the most common congenital heart abnormalities seen in the adult population, affecting 1% to 2% of adults worldwide; men constitute about 70% to 80% of the cases (1). These patients often exhibit various anatomic and functional changes, such as accelerated calcification and valvular stenosis, coronary artery anomalies, and coronary artery disease. BAV aortopathy is a term coined to describe the high prevalence of aortic dilation, aneurysm, and dissection seen in BAV patients (2).

Over the past years, a modified surgical technique offers BAV patients with root aneurysms to replace the dilated segment while preserving the valvular components (3). The VSARR was associated with lower surgical complications such as heart block and cerebral hemorrhage and had a higher survival rate than the traditional Bentall surgical procedure, which also uses replacement of the aortic valve (4).

TAVR is the criterion standard for aortic stenosis patients across all risk groups, and the ongoing experience over the past decade encourages centers to include BAV patients with aortic stenosis. The initial results are promising and show similar outcomes in comparison with tricuspid TAVR patients (5-6).

Nowadays, SAVR remains the treatment of choice for pure AI bicuspid patients because these patients usually present with larger annular size and insufficient calcification for THV anchoring, making TAVR more challenging.

Recently, a new nonrandomized large-scale study found no significant difference in in-hospital

FIGURE 3 Type 1 Bicuspid Aortic Valve in Various Imaging Modalities

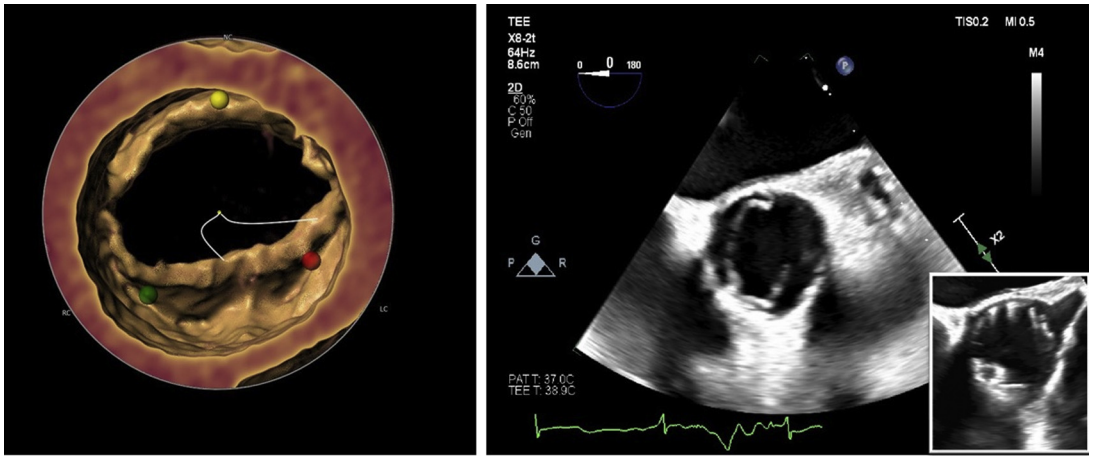
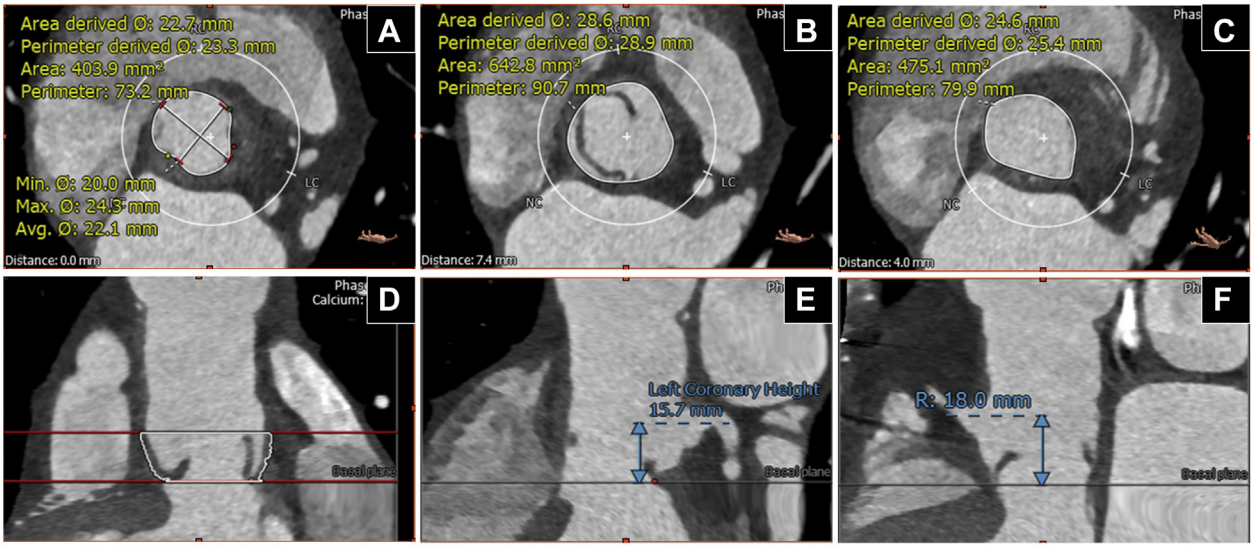


FIGURE 4 Preprocedural Reconstructed Computed Tomographic Measurements



mortality between SAVR and TAVR in AI patients (7), which adds to numerous additional promising data and hope for AI patients.

BAI patients after root replacement make the percutaneous approach even more challenging. Choosing a proper THV should be balanced with the desired EOA, future coronary access, native annulus size, and graft morphology. The use of TAVR in high-risk patients with severely reduced left ventricular function, lack of aortic valve calcification, and significantly large annular size should be limited until further data support its efficacy and safety. Given the limited data of TAVR on long-term outcomes in BAV patients, TAVR may be considered in high-risk patients in whom VSARR has failed.

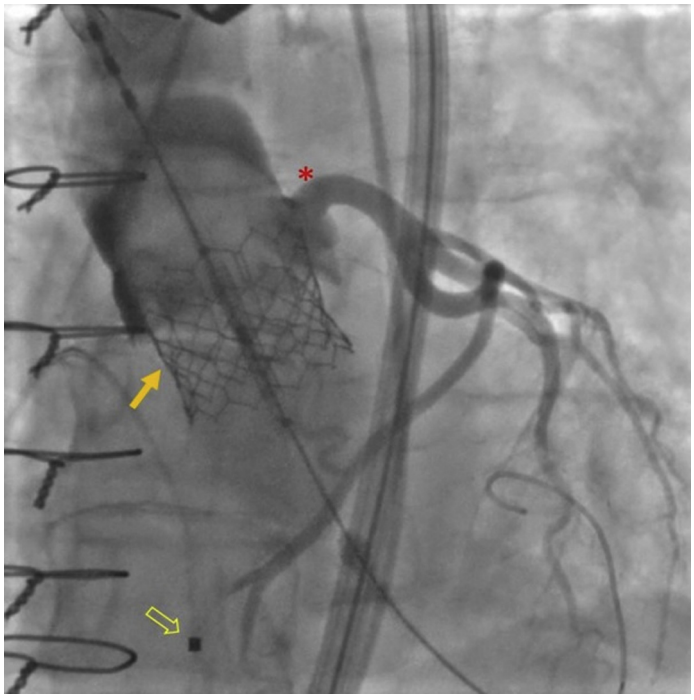
Our case highlights the need, the feasibility, and the dilemmas of the TAVR procedure in patients with failed VSARR and patients with bicuspid aortic regurgitation.

FOLLOW-UP

Post-TAVR echocardiography indicated a well-seated THV with normal leaflet motion and a mean gradient of 6 mm Hg. There was no significant central aortic insufficiency or paravalvular leak (Video 1).

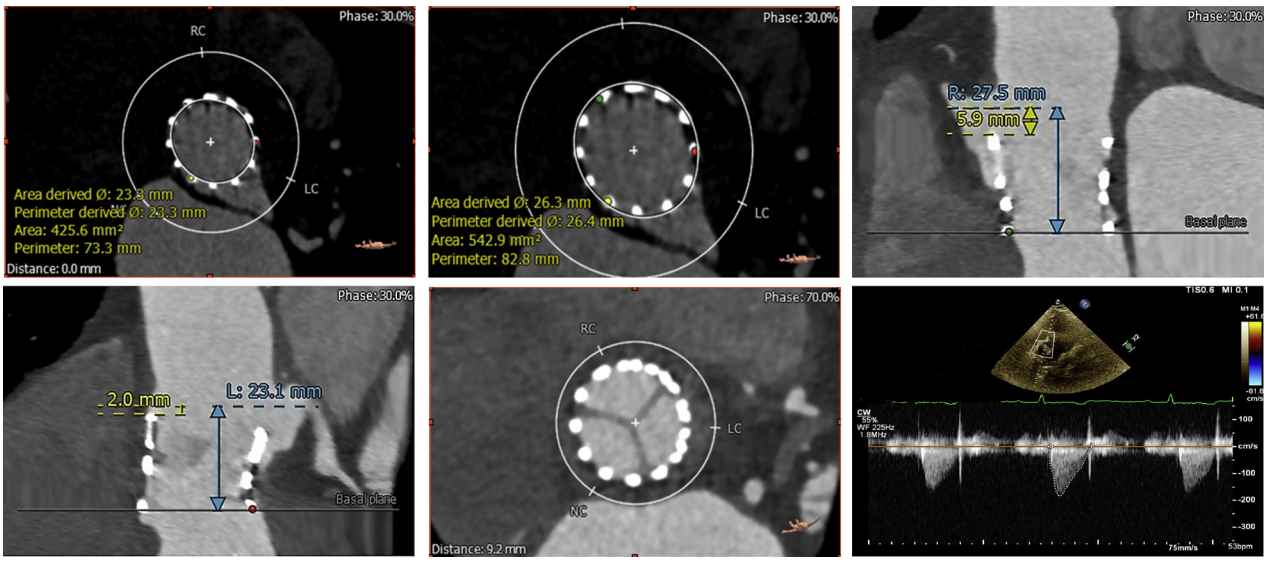
The Doppler-velocity index was 0.45, and the neoeffective orifice area (EOA) was 1.6 cm^2/m^2. The patient was discharged 2 days after the procedure feeling much better without dyspnea or shortness of breath. A 30-day post-TAVR CT indicated an annular

FIGURE 5 Fluoroscopic View During Transcatheter Aortic Valve Replacement



Solid yellow arrow points to fully expandable Sapien-3 transcatheter heart valve. **Open yellow arrow** points to tip of a right ventricle temporary pacemaker. **Red asterisk** points over the left main coronary artery.

FIGURE 6 Reconstructed Computed Tomographic and Echocardiographic View 30 Days After Transcatheter Aortic Valve Replacement



area of 542.9 mm², a mean diameter of 26.3 mm, and patency of the coronary ostia (Figure 6).

CONCLUSIONS

Patients with BAV exhibit valvular dysfunction and aortic root aneurysms and have been traditionally treated with the replacement of aortic components, including replacement of the aortic valve with a prosthetic valve. Nowadays, with the ongoing use of TAVR in BAV patients and the evolution of the valve-sparing aortic root replacement technique, patients can also have a minimally invasive therapeutic option and lifelong freedom from the use of anticoagulant agents. Our case highlights this clinical scenario, the periprocedural dilemmas, and the feasibility of the procedure.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

Dr. Makkar has received grant support from Edwards Lifesciences Corporation; is a consultant for Abbott Vascular, Cordis, and Medtronic; and holds equity in Entourage Medical. Dr Chakravarty is a consultant, proctor, and speaker for Edwards Lifesciences and Medtronic; is a consultant for Abbott Lifesciences; and is a consultant and speaker for Boston Scientific. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Raj R. Makkar, Cedars-Sinai Smidt Heart Institute, 127 S. San Vicente Boulevard, Advanced Health Sciences Pavilion, Third Floor, Suite A3100, Los Angeles, California 90048, USA. E-mail: Raj.Makkar@cshs.org. Twitter: @dr_ofir.

REFERENCES

1. Ochiai T, Chakravarty T, Yoon SH, et al. Coronary access after TAVR. *J Am Coll Cardiol Intv*. 2020;13(6):693–705.

2. Ehrlich T, de Kerchove L, Vojacek J, et al. State-of-the art bicuspid aortic valve repair in 2020. *Prog Cardiovasc Dis*. 2020;63(4):457–464.

3. Youssefi P, El-Hamamsy I, Lansac E. Rationale for aortic annuloplasty to standardise aortic valve repair. *Ann Cardiothorac Surg*. 2019;8(3):322–330.

4. Patel PM, Wei JW, McPherson L, Binongo J, Leshnower BG, Chen EP. Bicuspid aortic valve sparing root replacement. *J Card Surg*. 2021;36(1):118–123.

5. Yoon SH, Webb JG, Leon MB, Makkar R. Transcatheter aortic valve replacement in bicuspid aortic valve stenosis. *Prog Cardiovasc Dis*. 2020;63(4):482–487.

6. Makkar RR, Thourani VH, Mack MJ, et al. Five-year outcomes of transcatheter or surgical aortic-valve replacement. *N Engl J Med*. 2020;382(9):799–809.

7. Alharbi AA, Khan MZ, Osman M, et al. Transcatheter aortic valve replacement vs surgical replacement in patients with pure aortic insufficiency. *Mayo Clin Proc*. 2020;95(12):2655–2664.

KEY WORDS aortic aneurysm, aortic valve insufficiency, bicuspid aortic valve disease, David procedure, transcatheter aortic valve replacement, valve-sparing aortic replacement

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