The Texas Heart Institute® Journal

Case Reports

Transcatheter Aortic Valve-in-Valve-in-Valve Replacement in a Young Woman With Transcatheter Structural Valve Deterioration Within a Degenerated Aortic Root Homograft

Jonathan C. Hong, MD, MHS[1]; Vicente Orozco-Sevilla, MD[1,2]; Jose G. Diez, MD[3,4]; Joseph S. Coselli, MD[1,2,5]

[1]Division of Cardiothoracic Surgery, Michael E. DeBakey Department of Surgery, Baylor College of Medicine, Houston, Texas
[2]Department of Cardiovascular Surgery, Texas Heart Institute, Houston, Texas
[3]Division of Cardiology, Department of Medicine, Baylor College of Medicine, Houston, Texas
[4]Department of Cardiology, Texas Heart Institute, Houston, Texas
[5]Cardiovascular Research Institute, Baylor College of Medicine, Houston, Texas

Abstract

Transcatheter aortic valve replacement is a well-established procedure for older patients with symptomatic, severe aortic stenosis. However, data are lacking on its durability and long-term complications, particularly in young patients and patients treated for aortic valve regurgitation. This article describes the case of a 27-year-old woman with complex congenital cardiovascular disease who, after 4 previous aortic valve replacement procedures, presented with structural deterioration of her most recent replacement valve, which had been placed by transcatheter aortic valve replacement inside a failed aortic root homograft 6 years earlier. After the patient had undergone this transcatheter aortic valve replacement procedure to treat aortic valve regurgitation related to her degenerated aortic root homograft, she became pregnant and successfully carried her high-risk pregnancy to term. However, the replacement valve deteriorated during the late stages of pregnancy, resulting in substantial hemodynamic changes between the first trimester and the postpartum period. To avoid repeat sternotomy, a redo transcatheter valve-in-valve replacement procedure procedure was performed through the right carotid artery. Because the patient wanted to have more children and therefore avoid anticoagulation, a SAPIEN 3 transcatheter valve (Edwards Lifesciences) was placed as a bridge to a future, more-durable aortic root replacement. The result in this case suggests that in patients with complex adult congenital pathology, transcatheter aortic valve replacement can be used as a temporizing bridge to subsequent, definitive aortic valve repair.

Keywords: Aortic valve; heart defects, congenital; heart valve prosthesis; reoperation; transcatheter aortic valve replacement

Introduction

Transcatheter aortic valve replacement (TAVR) is a well-established procedure for older patients (eg, older than 70 years) with symptomatic, severe aortic stenosis. This procedure is standard of care for patients at prohibitive or high surgical risk and is considered a reasonable alternative in intermediate- and low-surgical-risk patients.[1,2] However, data are lacking on its durability and long-term complications, particularly in young patients. Here, we describe the case of a 27-year-old woman who had undergone TAVR within a failed aortic root homograft 6 years earlier.[3] She presented with structural deterioration of the bioprosthetic transcatheter replacement valve and had recently had a successful high-risk pregnancy. The patient wanted to continue to avoid anticoagulation because she

Citation: Hong JC, Orozco-Sevilla V, Diez JG, Coselli JS. Transcatheter aortic valve-in-valve-in-valve replacement in a young woman with transcatheter structural valve deterioration within a degenerated aortic root homograft. *Tex Heart Inst J*. 2023;50(2):e227874. doi:10.14503/THIJ-22-7874
Corresponding author: Joseph S. Coselli, MD, Department of Cardiothoracic Surgery, One Baylor Plaza, BCM 390, Houston, TX 77030 (jcoselli@bcm.edu)

planned to have more children. We decided to perform a TAVR-in-TAVR-in-homograft procedure as a temporizing bridge to subsequent aortic root replacement.

Case Report

This report was prepared in accordance with a clinical research protocol approved by the Baylor College of Medicine institutional review board (BCM H-18095). Written informed consent was obtained from the patient.

The patient was a 27-year-old Chinese American woman with congenital aortic valve stenosis and congenital vascular anomalies, including the absence of the infrarenal abdominal aorta. She had a complex surgical history, which was described previously.[3] As a child, she underwent repair of a ventricular septal defect via a left thoracotomy (at age 2 years), mechanical aortic valve replacement through a median sternotomy (at age 5 years), a Konno procedure for aortic root enlargement and bioprosthetic aortic valve replacement via a repeat sternotomy (at age 10 years), and homograft aortic root replacement via a third-time sternotomy (at age 14 years). At age 21 years, she underwent TAVR (with a 26-mm SAPIEN XT [Edwards Lifesciences]) via a redo left thoracotomy transapical approach to treat moderate to severe aortic valve stenosis and severe aortic valve regurgitation related to calcification and degeneration of the aortic root homograft, which otherwise remained in place. The patient was closely followed up with imaging surveillance and remained well for several years.

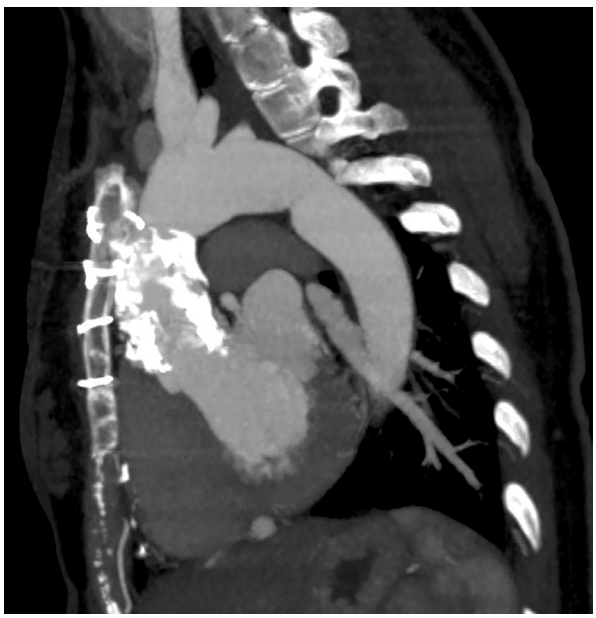

Fig. 1 Preoperative computed tomography angiogram in sagittal oblique view shows the calcified aortic root homograft, the prior transcatheter aortic valve (SAPIEN XL), and their proximity to the chest wall.

Abbreviations and Acronyms

TAVR transcatheter aortic valve replacement

At age 27 years, the patient underwent an uncomplicated pregnancy, was induced at 38 weeks for cholestasis, and vaginally delivered a healthy child. After her pregnancy, she reported increased shortness of breath on exertion (New York Heart Association Class II). A transthoracic echocardiogram revealed sudden structural valve deterioration with severe calcific degeneration and stenosis. The peak gradient was 116 mm Hg, and the mean gradient was 79 mm Hg. There was a mild paravalvular leak along the posterior and anterior aspect of the annulus. Her left ventricle had normal size and function. There was eccentric, posteriorly directed mild to moderate mitral regurgitation and trace tricuspid regurgitation. A computed tomographic angiogram (Fig. 1) showed a 23.9- × 24.6-mm annulus at the basal plane of the 26-mm SAPIEN XT valve. Her congenital absence of the abdominal aorta precluded standard retrograde deployment of the transcatheter valve. Notably, the carotid and subclavian arteries were patent and free of stenosis bilaterally and thus were suitable for transcatheter valve deployment.

The patient planned to have more children and wanted to continue to avoid anticoagulation for at least a few more years. For this reason, as well as the high risk associated with a fourth-time sternotomy and the patient's likelihood of requiring a redo root replacement (given the severely calcified aortic root homograft) within the next several years, our multidisciplinary heart valve team decided to perform a TAVR-in-TAVR through the right carotid artery to bridge her to future aortic intervention.

General anesthesia was induced in the hybrid cardiovascular operating room. The anesthesia team placed a left radial artery line for continuous hemodynamic monitoring. A transesophageal echocardiography probe was inserted. Cerebral oximetry was monitored throughout the procedure. A 5F, balloon-tipped bipolar pacing catheter was advanced through a 6F sheath in the left internal jugular vein and positioned in the right ventricle. Pacing thresholds were interrogated. A 4-cm incision was made in the right neck, anterior to

the sternocleidomastoid muscle. The right common carotid artery was exposed and controlled with vessel loops. Heparin was administered for an activated clotting time of more than 300 seconds. Percutaneous access was obtained in the right ulnar artery with a 6F sheath, and a pigtail catheter was placed in the aortic root. An aortic root angiogram was performed to establish the spatial relationship between the prior TAVR and the coronary arteries previously reimplanted onto the aortic root homograft (Fig. 2). Systolic blood pressure was maintained at greater than 120 mm Hg. The right common carotid artery was accessed with a large-bore needle, and a Glidewire (Terumo) was placed in the ascending aorta. A 6F sheath was placed, followed by an Amplatz left 2 catheter (Boston Scientific) and a straight wire, which was advanced into the left ventricle. A pigtail catheter was placed. This was exchanged for an Amplatz extra-stiff wire.

The delivery system sheath was inserted, and a 23-mm SAPIEN 3 transcatheter bioprosthetic valve (Edwards Lifesciences) was advanced and deployed with rapid ventricle pacing. After deployment, the delivery catheter was retracted, and angiographic contrast material was injected into the root to confirm the valve's position, spatial orientation, and proximity to the coronary arteries (Fig. 3). A transesophageal echocardiogram revealed satisfactory positioning and function of the TAVR. The patient's left ventricular end-diastolic pressure was 24 mm Hg at baseline and 19 mm Hg after the procedure. There was no gradient across the TAVR on pullback. The introducer sheath was removed, and the carotid artery was closed with 6-0 Prolene sutures (Ethicon, Inc). Protamine was administered. Cerebral oximetry was stable throughout the procedure.

The patient was extubated in the postoperative intensive care unit and was neurologically intact. A transthoracic echocardiogram showed that the aortic valve had a peak gradient of 59 mm Hg, a mean gradient of 32 mm Hg, and a dimensionless obstructive index of 0.3. No prosthetic aortic valve regurgitation was seen. The patient had no conduction abnormalities. She was discharged on postoperative day 2 in stable condition. At 14-month clinical follow-up after her bridge TAVR-in-TAVR procedure, this patient remained well, without any limitations regarding her daily activities, and remained employed. At 14-month echocardiographic follow-up, results showed that the aortic valve function was essentially unchanged since repair; the aortic valve had a mean gradient of 35 mm Hg and a dimensionless obstructive index of 0.3. In addition, the left ventricular ejection fraction of 55% to 60% indicated normal function.

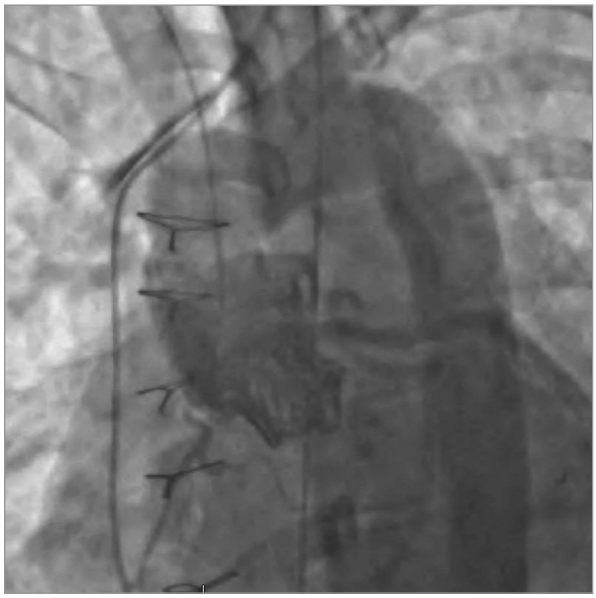

Fig. 2 Predeployment aortic root angiogram shows the prior transcatheter aortic valve in relation to the aortic root and coronary arteries.

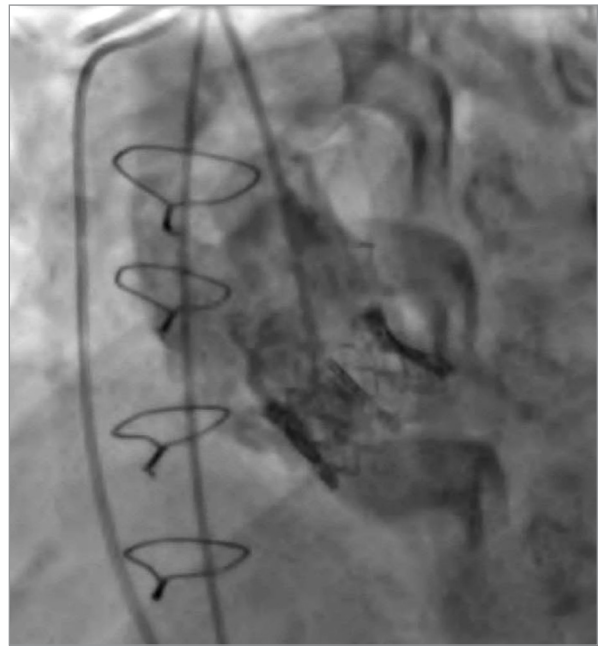

Fig. 3 Postdeployment aortic root angiogram shows satisfactory positioning of the transcatheter aortic valve (SAPIEN 3) inside a previously placed SAPIEN XL within a calcified and degenerated aortic root homograft.

Discussion

The patient was a young woman in the third decade of life with a failing transcatheter bioprosthetic aortic valve within a stenotic, calcified, and degenerated aortic root homograft. Because of her congenital disease, she had a complex surgical history that included 3 sternotomies. The following treatment strategies were considered: (1) redo sternotomy, bioprosthetic aortic valve replacement, and possible aortic root replacement; (2) redo sternotomy, pulmonary autograft (ie, Ross) procedure to replace the degenerated aortic root homograft, and additional homograft replacement in the pulmonary position; and (3) valve-in-valve TAVR within the aortic root homograft (to avoid redo sternotomy).

Undoubtedly, the patient will eventually require a redo sternotomy to replace her aortic root. The best long-term strategy would be to use a mechanical composite valve graft to replace the aortic root because mechanical valves have well-established durability. However, the patient expressed a desire to have more children and avoid vitamin K antagonists in the short term, despite understanding the long-term risks. Bioprosthetic aortic valve replacement (or aortic root replacement with a bioprosthetic composite valve graft) would only be a short-term solution because the lifespan of bioprosthetic valves is generally limited to 10 to 15 years.[4] A pulmonary autograft procedure in a prior Konno enlargement would be complicated by a calcified right ventricular outflow patch anchored to the pulmonic valve, so we were not certain that that approach would offer a durable long-term result.

Valve-in-valve TAVR is an alternative to surgical re-replacement. Some studies have shown better short-term outcomes with this procedure than with redo surgical aortic valve replacement.[5] We decided to do a redo valve-in-valve TAVR as a temporizing bridge to definitive long-term intervention. This would allow the patient to avoid anticoagulation and expose her to the least risk in the short term.

Although standard valve-in-valve TAVR has a favorable operative risk profile, it carries potential complications. It is a risk factor for patient-prosthesis mismatch, which is associated with poorer survival, functional status, left ventricular mass regression, and quality of life.[6] Our patient had some patient-prosthesis mismatch, with a postprocedure mean gradient of 32 mm Hg.

Coronary access is also a concern after redo valve-in-valve TAVR. When the second TAVR is deployed, the leaflets of the first prosthesis are vertically displaced, resulting in a covered cylinder that can interfere with blood flow into the coronary arteries. A recent study found that coronary angiography was not feasible in more than 30% of patients after TAVR-in-TAVR.[7] However, our patient was unlikely to undergo procedures requiring coronary access in the near future, given her young age and lack of atherosclerotic comorbidities.

Explanting TAVR valves is considerably more challenging than explanting surgically placed valves because of the crowded aortic root and endothelization of the implanted valve.[8] In a study of Medicare-insured patients, TAVR explantation was associated with a 30-day mortality rate of 13% and a 1-year mortality rate of 23%, albeit in an older adult patient cohort.[9] Our patient would probably require a root-replacement procedure, given the calcified aortic root homograft, and we believe that explanting an intra-annular TAVR-in-TAVR would not greatly increase the complexity of a future intervention.

Our result in this case suggests that although TAVR can be used as a temporizing bridge to subsequent, definitive aortic repair in patients with complex adult congenital pathology, pregnancy can place bioprosthetic transcatheter valves at great risk that is possibly related to hemodynamic changes, hormonal changes, or immune system changes that could cause rapid leaflet degeneration. This case is highly unusual and raises concern about future applications of TAVR in young patients.

Published: 14 March 2023

Conflict of Interest Disclosures: Dr Coselli consults for, receives royalties and a departmental educational grant from, and participates in clinical trials for Terumo Aortic; consults and participates in clinical trials for Medtronic, Inc and W.L. Gore & Associates; and serves as a co-investigator for CytoSorbents. Drs Hong, Orozco-Sevilla, and Diez have no disclosures.

Funding/Support: Dr Coselli's work is partly supported by the Cullen Foundation.

Acknowledgments: The authors wish to thank Stephen N. Palmer, PhD, ELS, of the Department of Scientific Publications at the Texas Heart Institute, and Susan Y. Green, MPH, of the Division of Cardiothoracic Surgery, Baylor College of Medicine, for editorial assistance.

References

1. Nishimura RA, Otto CM, Bonow RO, et al. 2017 AHA/ACC focused update of the 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical

Practice Guidelines. *J Am Coll Cardiol.* 2017;70(2):252-289. doi:10.1016/j.jacc.2017.03.011
2. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *J Am Coll Cardiol.* 2014;63(22):e57-e185. doi:10.1016/j.jacc.2014.02.536
3. Díez JG, Schechter M, Dougherty KG, Preventza O, Coselli JS. Transcatheter aortic valve-in-valve replacement instead of a 4th sternotomy in a 21-year-old woman with aortic homograft failure. *Tex Heart Inst J.* 2016;43(4):334-337. doi:10.14503/THIJ-15-5128
4. Pibarot P, Herrmann HC, Wu C, et al. Standardized definitions for bioprosthetic valve dysfunction following aortic or mitral valve replacement: JACC state-of-the-art review. *J Am Coll Cardiol.* 2022;80(5):545-561. doi:10.1016/j.jacc.2022.06.002
5. Deharo P, Bisson A, Herbert J, et al. Transcatheter valve-in-valve aortic valve replacement as an alternative to surgical re-replacement. *J Am Coll Cardiol.* 2020;76(5):489-499. doi:10.1016/j.jacc.2020.06.010
6. Herrmann HC, Daneshvar SA, Fonarow GC, et al. Prosthesis-patient mismatch in patients undergoing transcatheter aortic valve replacement: from the STS/ACC TVT registry. *J Am Coll Cardiol.* 2018;72(22):2701-2711. doi:10.1016/j.jacc.2018.09.001
7. Nai Fovino L, Scotti A, Massussi M, et al. Coronary angiography after transcatheter aortic valve replacement (TAVR) to evaluate the risk of coronary access impairment after TAVR-in-TAVR. *J Am Heart Assoc.* 2020;9(13):e016446. doi:10.1161/JAHA.120.016446
8. Fukuhara S, Brescia AA, Shiomi S, et al. Surgical explantation of transcatheter aortic bioprostheses: results and clinical implications. *J Thorac Cardiovasc Surg.* 2021;162(2):539-547 e1. doi:10.1016/j.jtcvs.2019.11.139
9. Hirji SA, Percy ED, McGurk S, et al. Incidence, characteristics, predictors, and outcomes of surgical explantation after transcatheter aortic valve replacement. *J Am Coll Cardiol.* 2020;76(16):1848-1859. doi:10.1016/j.jacc.2020.08.048