JSHD
Journal of Structural Heart Disease

Case Report

Journal of Structural Heart Disease, April 2018,
Volume 4, Issue 2:42-49
DOI: https://doi.org/10.12945/j.jshd.2018.042.17

Received: October 18, 2017
Accepted: November 28, 2017
Published online: April 2018

Transcatheter Aortic Valve Replacement in Transposition of the Great Arteries Following Arterial Switch Operation

Joanna Ghobrial, MD, MSc[1*], Eric H. Yang, MD[2], Olcay Aksoy, MD[2], William Suh, MD[2], Jamil Aboulhosn, MD[1, 2]

[1] Division of Cardiology, Ahmanson/UCLA Adult Congenital Heart Disease Center, University of California, Los Angeles, Los Angeles, California, USA

[2] Division of Cardiology, University of California, Los Angeles, Los Angeles, California, USA

Abstract

Transcatheter aortic valve replacement (TAVR) for the treatment of congenital heart disease has been mostly limited to patients with bicuspid aortic valve stenosis. We present a case of a 19 year old Jehovah's witness patient with D-Transposition of the great arteries (D-TGA) who underwent an arterial switch operation, followed by a valve sparing aortic root repair with a hemashield graft, who later developed severe aortic valve regurgitation and successfully underwent a transcatheter aortic valve replacement (TAVR) with the Edwards Sapien-3 valve.

Key Words

Transcatheter Aortic Valve Replacement (TAVR) • Transposition of the Great Arteries (TGA) • Arterial Switch Surgery

Introduction

The minimally invasive technique of transcatheter aortic valve replacement (TAVR) may be ideal for the growing population of adult congenital heart disease (ACHD) patients, many of whom may be at increased risk of adverse events associated with surgical valve replacement. TAVR has had limited application in the ACHD population to this point, mostly utilized for the treatment of bicuspid aortic valve (BAV) stenosis or valve-in-valve replacement of dysfunctional aortic bioprostheses. Aortic stenosis or aortic regurgitation (AR) in ACHD patients may be due to isolated valvular abnormality such as BAV or as a component of additional congenital anomalies such as hypoplastic left heart syndrome. Both aortic stenosis and AR can occur following aortic valve repair or replacement with bioprostheses, homografts, or pulmonary autografts as in the Ross procedure or in arterial switch operations. We herein present a case of TAVR in an ACHD patient with D-transposition of the great arteries (D-TGA) post status arterial switch and subsequent valve sparing aortic root repair with resultant severe AR.

Case Presentation

A 19-year-old woman of Jehovah's Witness faith with D-TGA, membranous ventricular septal defect, and subpulmonic stenosis underwent a right modified Blalock-Taussig-Thomas shunt and Blalock-Hanlon atrial septectomy at 1 year of age; arterial switch operation, sub-pulmonic stenosis resection, and atri-

Fax +1 203 785 3346
E-Mail: jshd@scienceinternational.org
http://structuralheartdisease.org/

© 2018 Journal of Structural Heart Disease
Published by Science International Corp.
ISSN 2326-4004

Accessible online at:
http://structuralheartdisease.org/

* Corresponding Author:
Joanna Ghobrial, MD, MSc
Heart and Vascular Institute
Cleveland Clinic Foundation
9500 Euclid Avenue/J2-4, Cleveland, OH 44195, USA
Tel. +1 216 444 5923; Fax: +216 445 6163; E-Mail: joannaghobrial@gmail.com

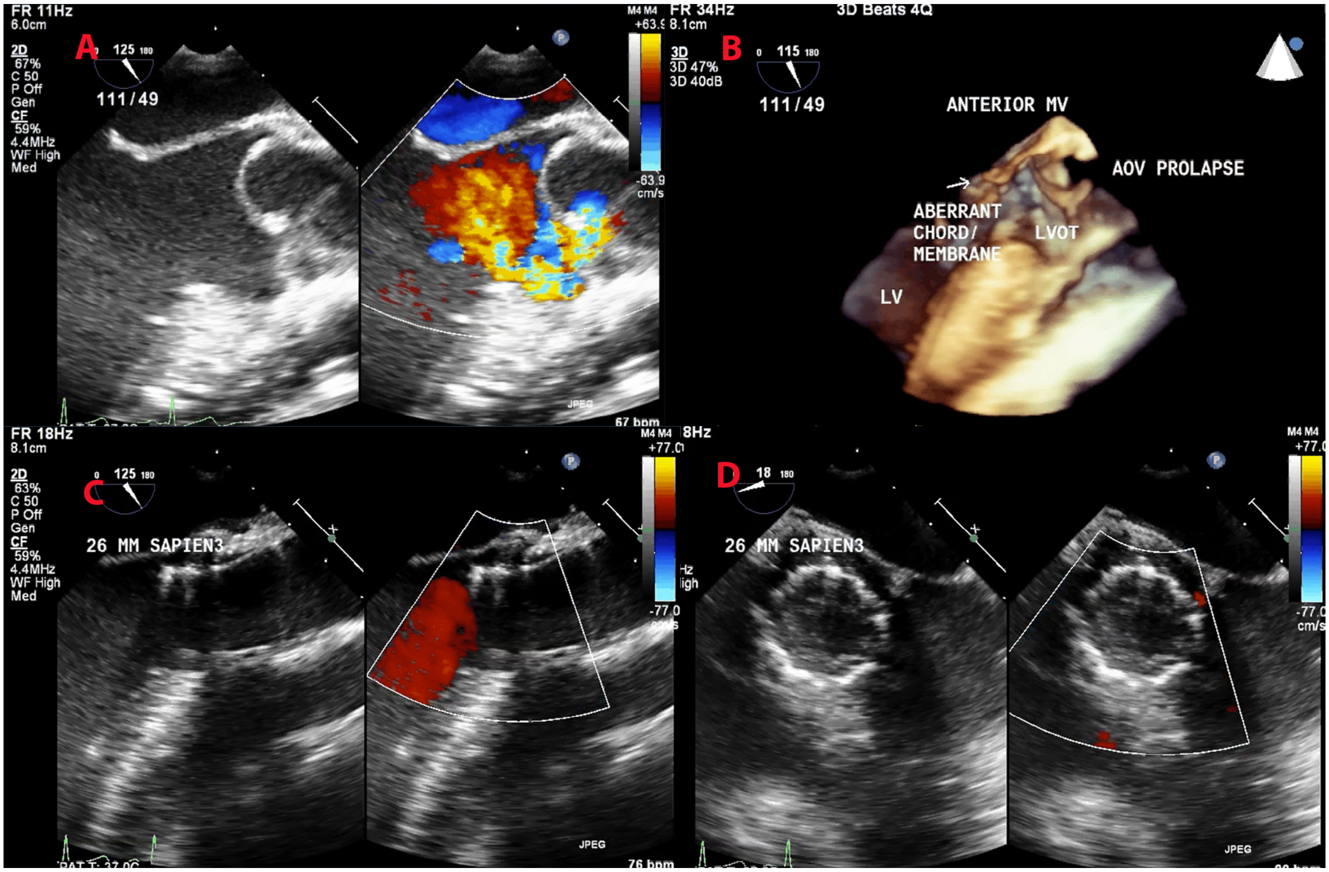
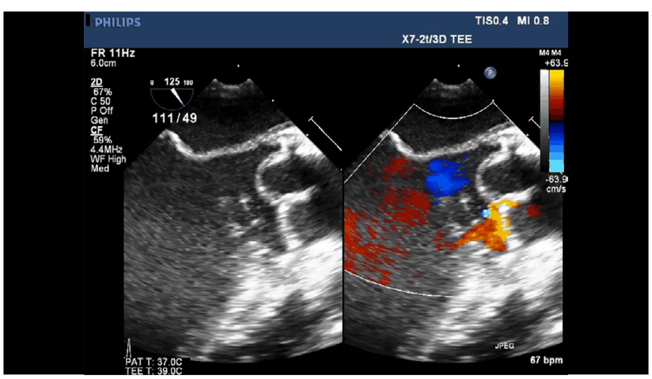
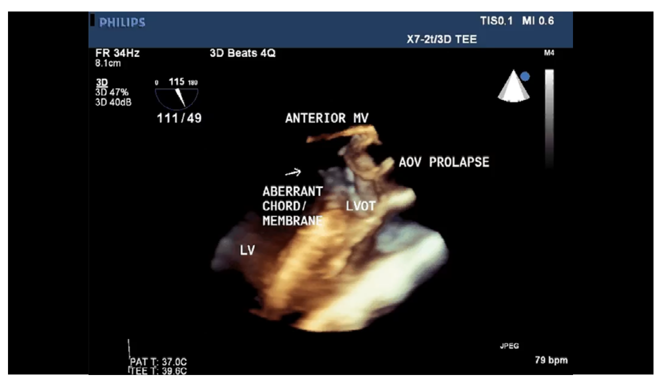


Figure 1. Transesophageal echocardiography (TEE) of transcatheter aortic valve replacement (TAVR) in D-transposition of the great arteries (D-TGA) status post arterial switch with a valve sparing root repair using a hemashield graft and repair of severe aortic regurgitation using a 26-mm Edwards Sapien-3 valve. *Panel A*. TEE two-dimensional (2D) long axis view of the aortic valve demonstrating left coronary cusp prolapse and severe regurgitation. *Panel B*. TEE three-dimensional (3D) long axis view of the aortic valve demonstrating left coronary cusp prolapse and aberrant chord in the left ventricular outflow tract. *Panel C*. TEE 2D long axis view of the aortic valve status post deployment of a 26-mm Sapien-3 valve with no residual regurgitation and trace perivalvular regurgitation. *Panel D*. TEE 2D short axis view of the aortic valve status post deployment of a 26-mm Sapien-3 valve with no residual regurgitation and trace perivalvular regurgitation.



Video 1. TEE 2D long axis view of the aortic valve with color compare showing the prolapsing aortic valve leaflet and severe regurgitation. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.01.



Video 2. TEE 3D view of the aortic valve prolapse and aberrant chord. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.02.

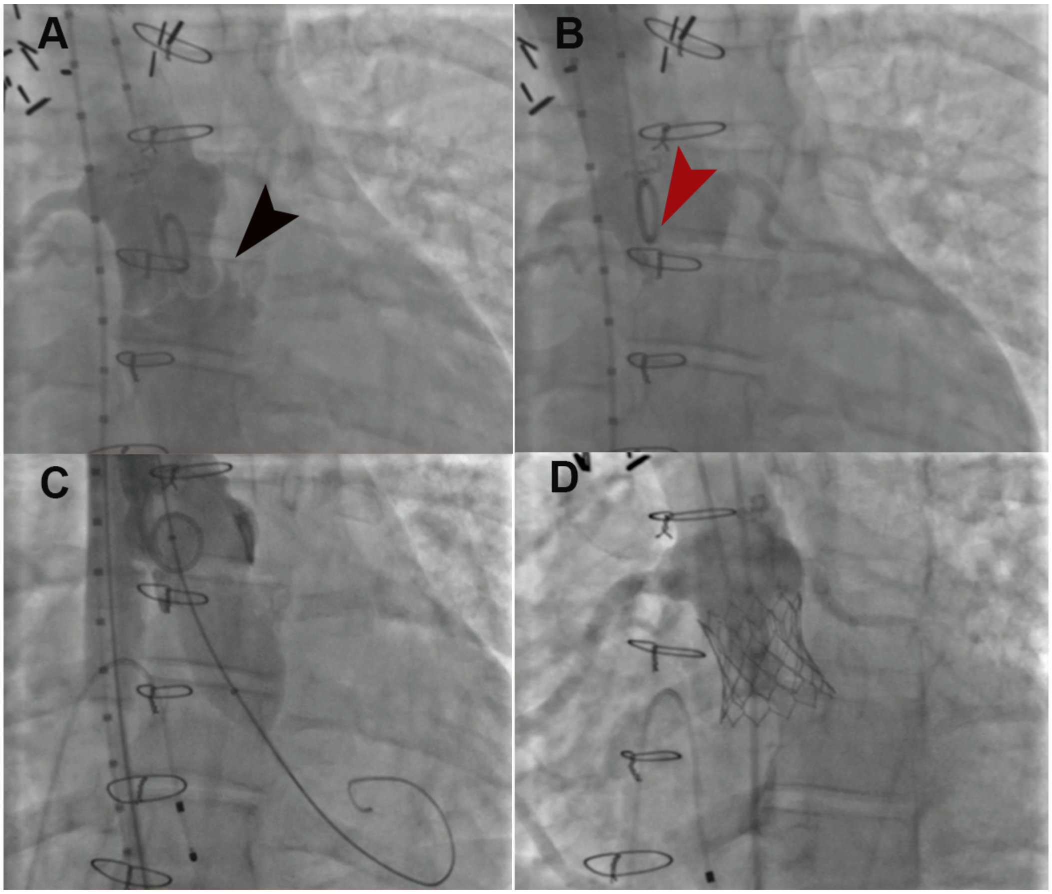
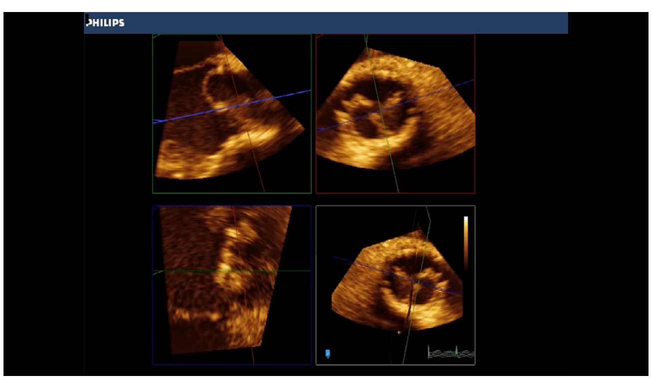


Figure 2. Angiography of transcatheter aortic valve replacement in D-TGA status post arterial switch with a valve sparing root repair using a hemashield graft and repair of severe aortic regurgitation with a 26-mm Edwards Sapien-3 valve. *Panel A.* Aortic root angiogram demonstrating the severely regurgitant valve with opacification of the left ventricle and re-implanted coronary arteries at a safe height from the aortic valve annulus (**black arrow** marks the aortic valve leaflets). *Panel B.* The aortic valve in an open position (**red arrow**). *Panel C.* Balloon sizing of the aortic valve annulus and aortic valve angioplasty with a waist measuring 19–21 mm. *Panel D.* Aortic root angiogram status post 26-mm Edwards Sapien-3 valve deployment with no residual regurgitation, no perivalvular regurgitation, and normal coronary artery flow.



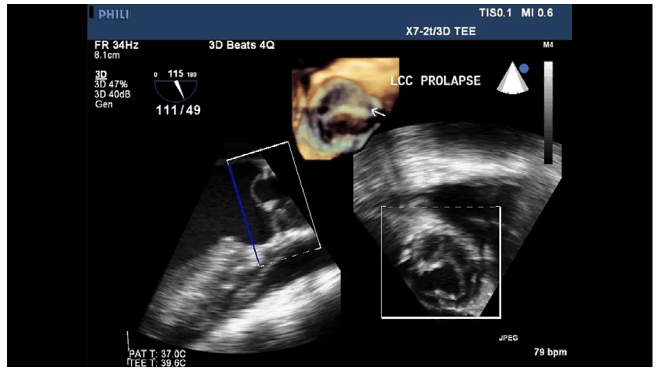
Video 3. TEE 3D views of the aortic valve demonstrating the left coronary cusp prolapse; views also used for annular dimensions. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.03.

al and ventricular septal defect closure at 2 years of age; and valve-sparing aortic root replacement with a 22-mm Hemashield graft and subaortic membrane resection at 15 years of age. The last operation was complicated by cognitive injury and memory loss, and her recovery was protracted. She subsequently developed symptomatic severe AR. Because of her refusal to receive blood transfusions due to her Jehovah's Witness faith and history of three surgeries, two median sternotomies, and post-operative cognitive injury, she was considered high risk for repeat surgical intervention and was referred for off-label use of TAVR.

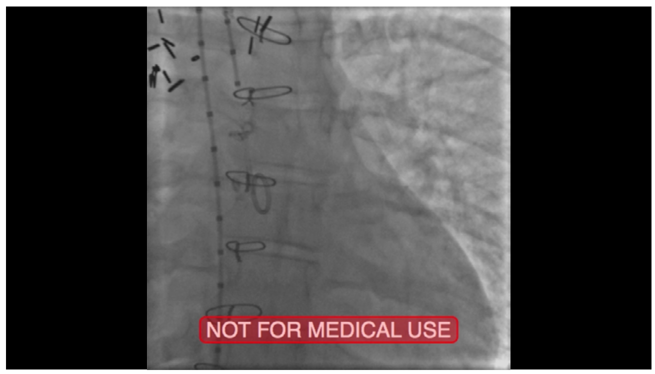
Procedure

Pre-procedural electrocardiography-gated cardiac computed tomographic angiography (CTA) showed the valve annular dimensions to be 21 × 20 × 23 mm, adequate clearance of the coronary ostia to the annulus, and no aortic valve or ascending aortic calcification. Abdominal/pelvic CTA demonstrated adequate femoral arterial size for a transfemoral valve replacement approach without significant tortuosity.

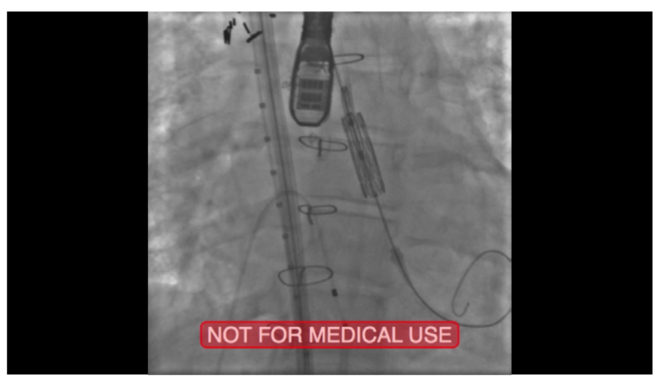
The procedure was performed under general anesthesia. Intra-procedural transesophageal echocardiography (TEE) showed severe eccentric aortic regurgitation due to malcoaptation of the aortic valve leaflets and prominent prolapse of the left coronary cusp leaflet (Figure 1; Videos 1, 2, 3, and 4). There was a false chord extending from the anterior mitral valve leaflet to the left ventricular outflow tract without functional obstruction (Video 2). The aortic valve annulus measured 20 × 20 × 21 mm by TEE, mostly consistent with the CTA annular dimensions. Aortic root angiograms (Figure 2; Video 5) showed severe AR and patent re-implanted coronary arteries without evidence of atherosclerotic disease with a distance from the valve level of 27–30 mm and minimal concern for coronary obstruction. A 23 mm × 4 cm balloon was used to perform balloon sizing with rapid ventricular pacing at 180 bpm; the waist on the balloon was noted to be 21 mm. A 26-mm Edwards Sapien-3 valve (Edwards LifeSciences, Irvine, CA, USA) was prepped and mounted onto the Commander delivery system in the usual manner and advanced into the 14-F sheath, and the valve was assembled in the descending aorta. Once across the valve, aortic angiography was performed to ensure appropriate position of the Sapien valve. Rapid pacing at 180 bpm was initiated, and the valve was deployed using the nominal volume of 23 ml (Video 6). Aortic root angiography was performed, which demonstrated a competent aortic valve (Figure 2; Video 7). Post-deployment TEE showed a well-positioned 26-mm Edwards Sapien-3 valve with trace perivalvular regurgitation at the left coronary cusp region and no central regurgitation (Figure 1; Videos 8 and 9). The delivery sheath was pulled and perclose sutures were deployed; however, right iliac angiography revealed that the right femoral artery was occluded at the site of the perclose su-



Video 4. TEE 2 and 3D views of the aortic valve showing the left coronary cusp prolapse. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.04.



Video 5. Baseline aortic root angiogram demonstrating the severely regurgitant aortic valve and that the re-implanted coronary arteries were at a safe height from the aortic annulus. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.05.



Video 6. Deployment of a 26-mm Sapien-3 valve with aortic root angiogram prior to deployment and rapid pacing during deployment. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.06.

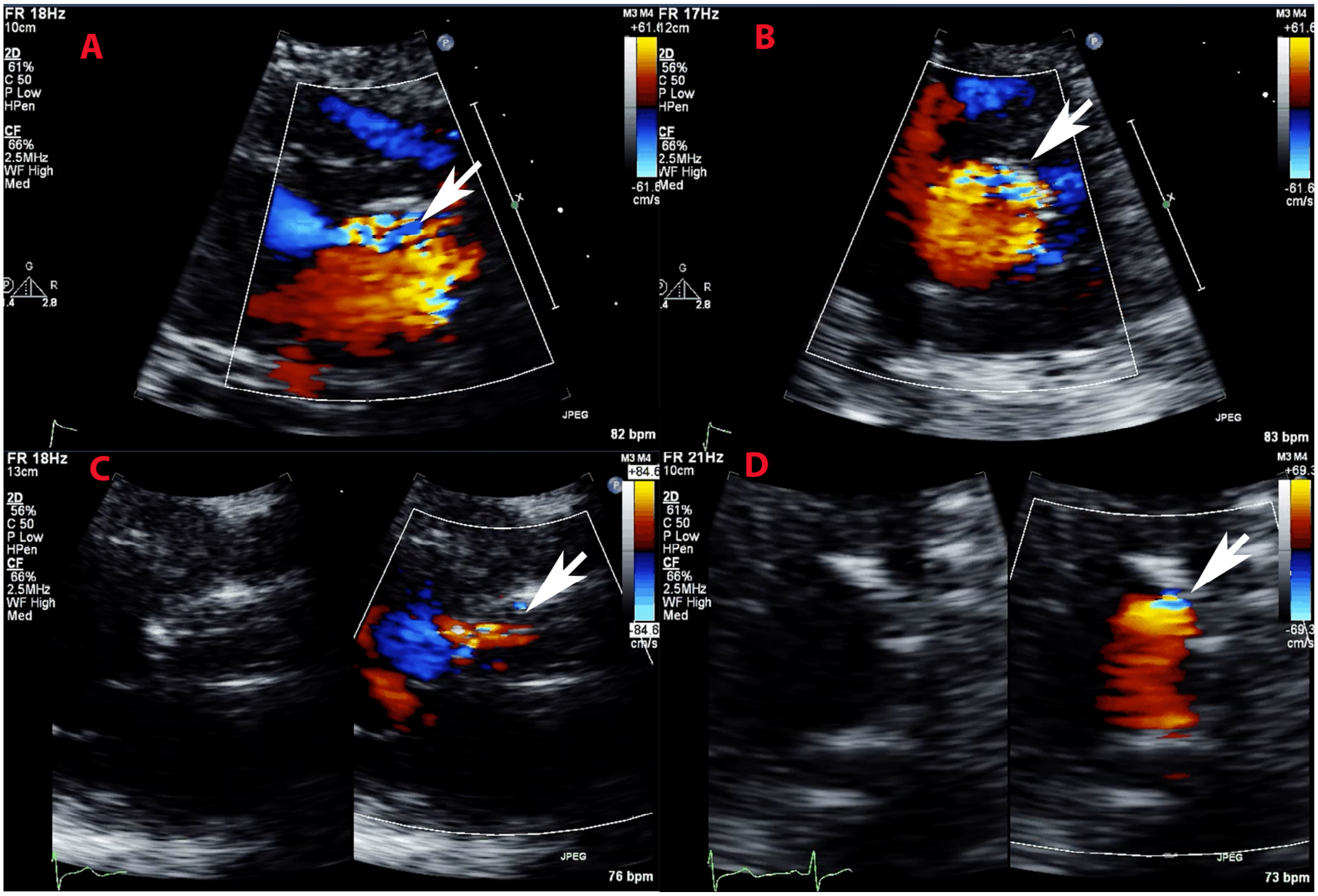
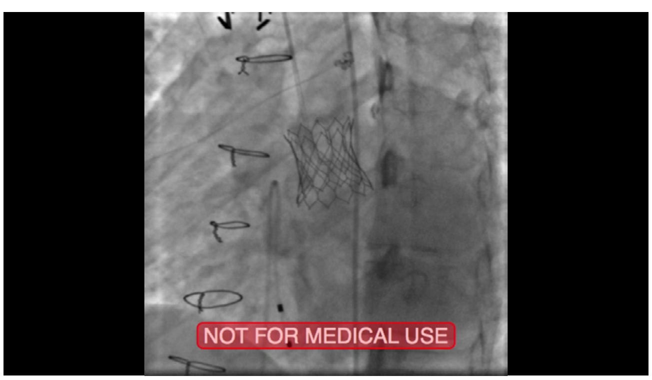
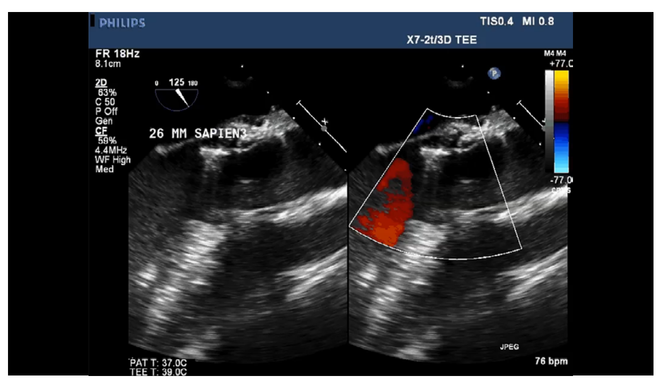


Figure 3. Transthoracic echocardiography (TTE) post-initial procedure and post-dilation of the aortic prosthesis. *Panel A.* TTE 2D long axis view of the aortic valve prosthesis with color Doppler demonstrating perivalvular regurgitation (**white arrow**). *Panel B.* TTE 2D short axis view of the aortic valve prosthesis with color Doppler demonstrating perivalvular regurgitation (**white arrow**). *Panel C.* TTE 2D long axis view of the aortic valve prosthesis with color Doppler demonstrating reduced perivalvular regurgitation post dilation (**white arrow**). *Panel D.* TTE 2D short axis view of the aortic valve prosthesis with color Doppler demonstrating reduced perivalvular regurgitation post dilation (**white arrow**).



Video 7. Post-valve deployment aortic root angiogram demonstrating no residual aortic regurgitation or perivalvular regurgitation and normal coronary artery flow. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.07.



Video 8. TEE 2D long axis view of the aortic valve status post deployment of the Sapien-3 valve. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.08.

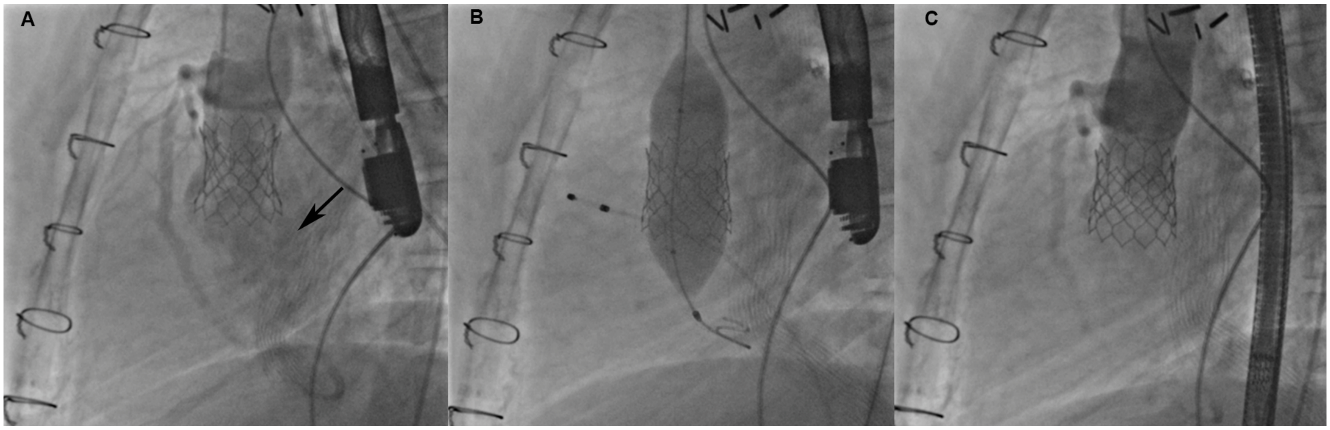
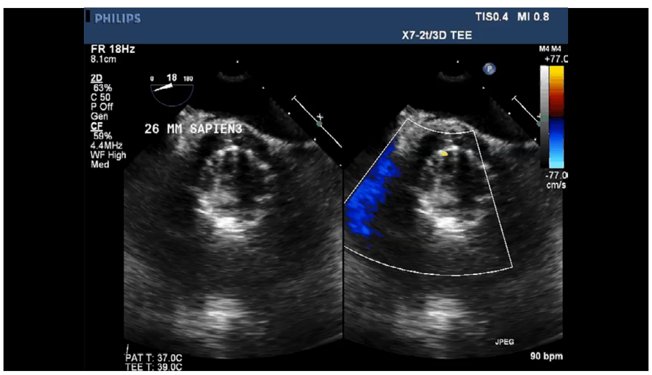
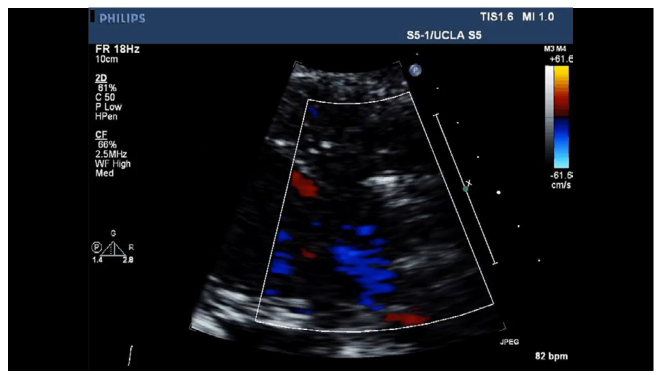


Figure 4. Angiography and angioplasty of the transcatheter aortic valve 10 months post-initial procedure**.** *Panel A.* Aortic root angiogram in the lateral projection demonstrating downward migration of the transcatheter valve of approximately 4–5 mm with severe perivalvular regurgitation (**black arrow**). *Panel B.* Angioplasty of the transcatheter Sapien 3 valve using a 24-mm VIDA balloon resolving the residual waist. Note the transvenous pacemaker in the right ventricle. *Panel C.* Post-angioplasty aortic root angiogram in the lateral projection demonstrating only mild residual perivalvular regurgitation.



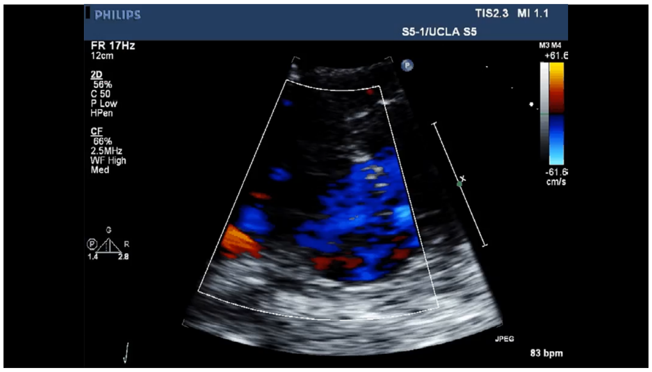
Video 9. TEE 2D short axis view of the aortic valve status post deployment of the Sapien-3 valve. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.09.



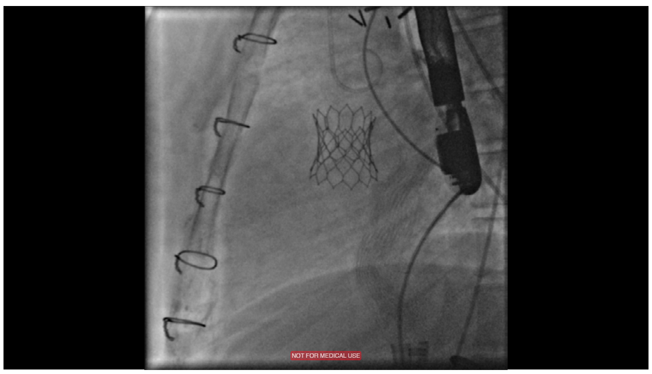
Video 10. TTE 2D long axis view of the aortic valve upon follow-up after the procedure demonstrating progressive perivalvular regurgitation. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.10.

tures. Surgical cutdown and open repair of the right femoral artery were performed with a good final result and return of normal peripheral pulses.

The patient was monitored in the surgical intensive care unit. Her B-natriuretic peptide level decreased from 121 pg/ml pre-procedure to 39 pg/ml. Her post-operative echocardiogram showed no evidence of central or perivalvular regurgitation. She was started on aspirin and clopidogrel and discharged on postoperative day 2. On initial clinic follow-up within 1 month post-procedure, she reported minimal improvement in her exertional symptoms, and her echocardiogram showed progressive perivalvular regurgitation (Figure 3; Videos 10 and 11). In the next few months following the initial TAVR, the patient reported worsening exertional symptoms. Given the perivalvular AR, she was brought back to the cardiac catheterization laboratory. Cineangiography demonstrated slight downward migration of the aortic prosthesis with evidence of severe perivalvular regurgitation (Figure 4; Video 12). This was hypothesized to be due to inadequate post-dilation of the Sapien valve in a patient with primary AR. A 24-mm Vida balloon (BARD Peripheral Vascular, Tempe, AZ, USA) was used to perform high-pressure dilation, which resulted in improved valve apposition to the walls of the hemashield graft and resolution of the perivalvular AR (Figure 4; Videos 13 and 14). The patient toler-

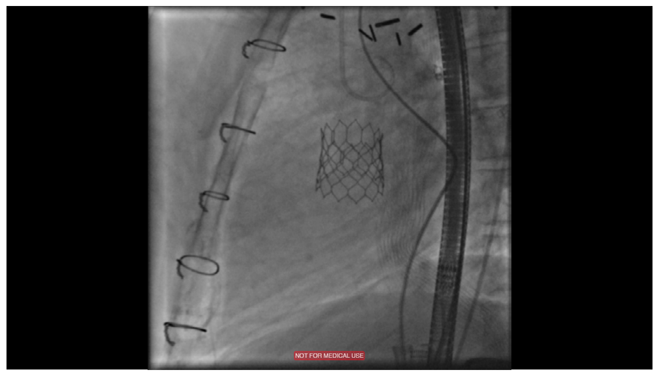


Video 11. TTE 2D short axis view of the aortic valve upon follow-up after the procedure demonstrating progressive perivalvular regurgitation. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.11.



Video 12. Aortic root angiogram in the lateral projection 10 months post-initial valve deployment demonstrating downward migration of the valve and severe perivalvular regurgitation. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.12.

Video 13. Balloon dilation of the Sapien 3 valve using a 24-mm VIDA balloon. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.13.

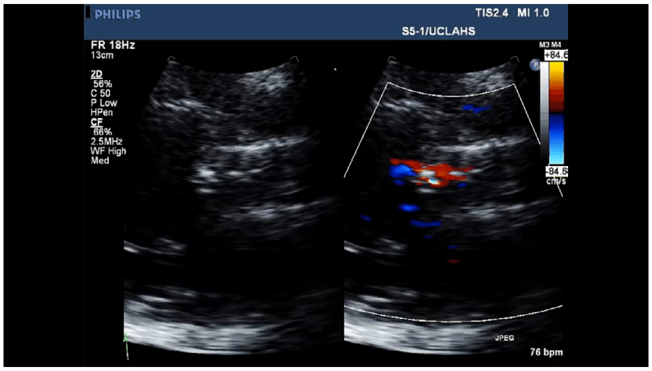


Video 14. Aortic root angiogram post-dilation demonstrating better apposition of the Sapien 3 valve and only mild residual perivalvular regurgitation. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.14.

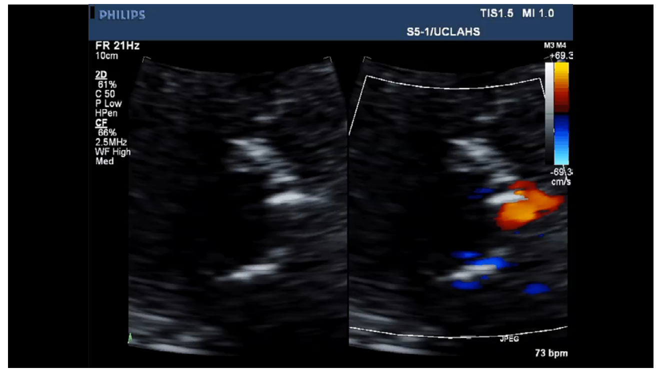
ated the procedure well, and follow-up echocardiography 4 months post-procedure demonstrated mild perivalvular regurgitation and stable valve position (Figure 3; Videos 15 and 16).

Discussion

TAVR in the ACHD population has been mostly limited to patients with calcific stenotic BAV. Initially, some studies showed a higher risk of perivalvular regurgitation in BAV TAVR with earlier generation valves including the CoreValve (Medtronic Inc, Minneapolis, MN, USA) and Sapien XT (Edwards Lifesciences) [1]. In later studies, however, this risk was mitigated with the use of newer generation valves such as the Lotus (Boston Scientific, Marlborough, MA, USA) and Sapien-3 (Edwards Lifesciences), which have a sealing system that decreases perivalvular regurgitation [2]. Nevertheless, the use of TAVR in the ACHD population has been scant, and many patients present with predominant AR without calcification. The present case illustrates the benefits and potential pitfalls of TAVR in an ACHD patient with primary AR. Although the hemashield graft served as an anchor for valve deployment well below the coronary artery buttons, there was evidence of gradual downward migration and development of severe perivalvular regurgitation despite initial valve placement at nominal balloon volume, indicating a potential role for deployment at greater than nominal volumes and the importance of adequate post-dilation. Several newer generation transcatheter valves are currently in preclinical or ear-



Video 15. TTE 2D long axis view of the aortic valve post-dilation demonstrating reduction in perivalvular regurgitation. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.15.



Video 16. TTE 2D short axis view of the aortic valve post-dilation demonstrating reduction in the perivalvular regurgitation. View supplemental video at https://doi.org/10.12945/j.jshd.2018.042.17.vid.16.

ly clinical testing for the treatment of primary AR and are equipped with an anchoring system that serves to stably and accurately position the valves in patients with predominant AR to prevent the migration and resultant perivalvular regurgitation seen in this study [3, 4]. These new platforms will expand the use of TAVR in ACHD patients to further decrease the need for repeat surgeries in this patient population. To our knowledge, this is the first published case of transcatheter deployment of an Edwards Sapien-3 valve in the aortic position in a patient with D-TGA post status arterial switch with predominant AR.

In conclusion, TAVR in patients with congenital heart disease and pulmonary conduit or bioprosthetic valve dysfunction has become the new standard of care. These minimally invasive and effective techniques have allowed for a reduction in the number of open cardiac surgeries needed over the lifetime of a CHD patient. Advances in technology are increasing the applications of TAVR in lesions that were previously only surgically treated. TAVR in ACHD patients with predominant AR is a new frontier, and the use of the newer-generation valves may prove promising.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

Comment on this Article or Ask a Question

References

1. Bauer T, Linke A, Sievert H, Kahlert P, Hambrecht R, Nickenig G, et al. Comparison of the effectiveness of transcatheter aortic valve implantation in patients with stenotic bicuspid versus tricuspid aortic valves (from the German TAVI Registry). Am J Cardiol. 2014;113:518-521. DOI: 10.1016/j.amjcard.2013.10.023
2. Yoon SH, Lefevre T, Ahn JM, Perlman GY, Dvir D, Latib A, et al. Transcatheter aortic valve replacement with early- and new-generation devices in bicuspid aortic valve stenosis. J Am Coll Cardiol. 2016;68:1195-1205. DOI: 10.1016/j.jacc.2016.06.041
3. Kumar R, Latib A, Colombo A, Ruiz CE. Self-expanding prostheses for transcatheter aortic valve replacement. Prog Cardiovasc Dis. 2014;56:596-609. DOI: 10.1016/j.pcad.2014.03.002
4. Treede H, Rastan A, Ferrari M, Ensminger S, Figulla H-R, Mohr F-W. JenaValve. EuroIntervention. 2012;8 Suppl Q:Q88-Q93. DOI: 10.4244/eijv8sqa16

Cite this article as: Ghobrial J, Yang EH, Aksoy O, Suh W, Aboulhosn J. Transcatheter Aortic Valve Replacement in Transposition of the Great Arteries Following Arterial Switch Operation. Structural Heart Disease. 2018;4(2):42-49. DOI: https://doi.org/10.12945/j.jshd.2018.042.17