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ADVANCED

CASE REPORT: CLINICAL CASE

Transcatheter Aortic Valve Replacement in a Patient With Criss-Cross Heart

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ABSTRACT

This paper presents the first transcatheter management of severe aortic regurgitation in a 77-year-old woman with a criss-cross heart—an extremely rare and complex congenital heart disease. The procedure achieved an elimination of aortic regurgitation and resulted in a remarkable improvement of the patient's physical condition. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:934-940) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 77-year-old woman with a criss-cross heart (CCH), a rare congenital heart disease, was admitted to our hospital for evaluation for transcatheter aortic valve replacement (TAVR) due to symptomatic severe aortic regurgitation (AR). Symptoms included dizziness and exertional dyspnea consistent with New York Heart Association (NYHA) functional class III.

LEARNING OBJECTIVES

- To understand the importance of transcatheter aortic valve replacement as an alternative to surgical valve replacement in patients with anatomical anomalies or congenital heart diseases.
- To appreciate the occurrence of rare anatomical anomalies in older patients requiring surgical intervention.

PAST MEDICAL HISTORY

The patient was born with a CCH—a complex and rare condition, accounting for <0.1% of all congenital heart diseases. It is characterized by abnormal rotation of the ventricles around the major axis resulting in a crossing of the ventricular inflows.[1] Our patient presented with superior-inferior ventricles and physiologically intact atrioventricular as well as

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ventriculoarterial connections (**Figure 1,** Video 1). The inferior left ventricle was normal-sized, yet moderately hypertrophic, and the superior right ventricle was extremely small with an end-diastolic volume of only 48 mL. The left and right atria were slightly enlarged, and the hypoplastic tricuspid valve was in an almost sagittal plane, superior to the mitral valve, which was in a frontal position. Additionally, the patient had a ventricular septal defect (VSD), which was closed with a percutaneous occluder.

Presumably due to the underlying heart disease and aortic root dilation, the patient developed moderate AR (regurgitation fraction of 22% 5 years ago) and mild-to-moderate mitral and tricuspid regurgitation, respectively, as well as paroxysmal atrial fibrillation and a left bundle branch block. Furthermore, the patient had moderate precapillary pulmonary hypertension and chronic obstructive pulmonary disease. A mammary carcinoma was also diagnosed and treated in a breast-conserving manner, though the patient had, at that point, declined radiotherapy.

At 77 years of age, she presented with dizziness and dyspnea on exertion (NYHA functional class III). Based on cardiac magnetic resonance imaging (CMR) (regurgitation fraction of 50%) and transthoracic echocardiography (TTE) (pressure half time of 355 ms, effective regurgitant orifice area of 0.28 cm^2, and regurgitant volume of 56 mL [**Figure 2**]), AR was graded as severe. Dilation of the aortic root and of the ascending aorta, up to 44 mm and 48 mm, respectively, as well as dilation of the left ventricle (left ventricular end-diastolic volume of 147 mL and diameter of 48 mm) were diagnosed, and the patient was then admitted to our institution for further investigation and evaluation for aortic valve replacement.

DIFFERENTIAL DIAGNOSIS

Other causes for the symptoms were excluded. No progression of the chronic obstructive pulmonary disease or other pulmonary findings were found in spirometry, x-ray, or computed tomography (CT). Furthermore, a progression of the pre-existing mitral valve regurgitation and a deterioration in left ventricular systolic function (left ventricular ejection

ABBREVIATIONS AND ACRONYMS

AR = aortic regurgitation

CCH = criss-cross heart

CMR = cardiac magnetic resonance

CT = computed tomography

NAVR = native aortic valve regurgitation

NYHA = New York Heart Association

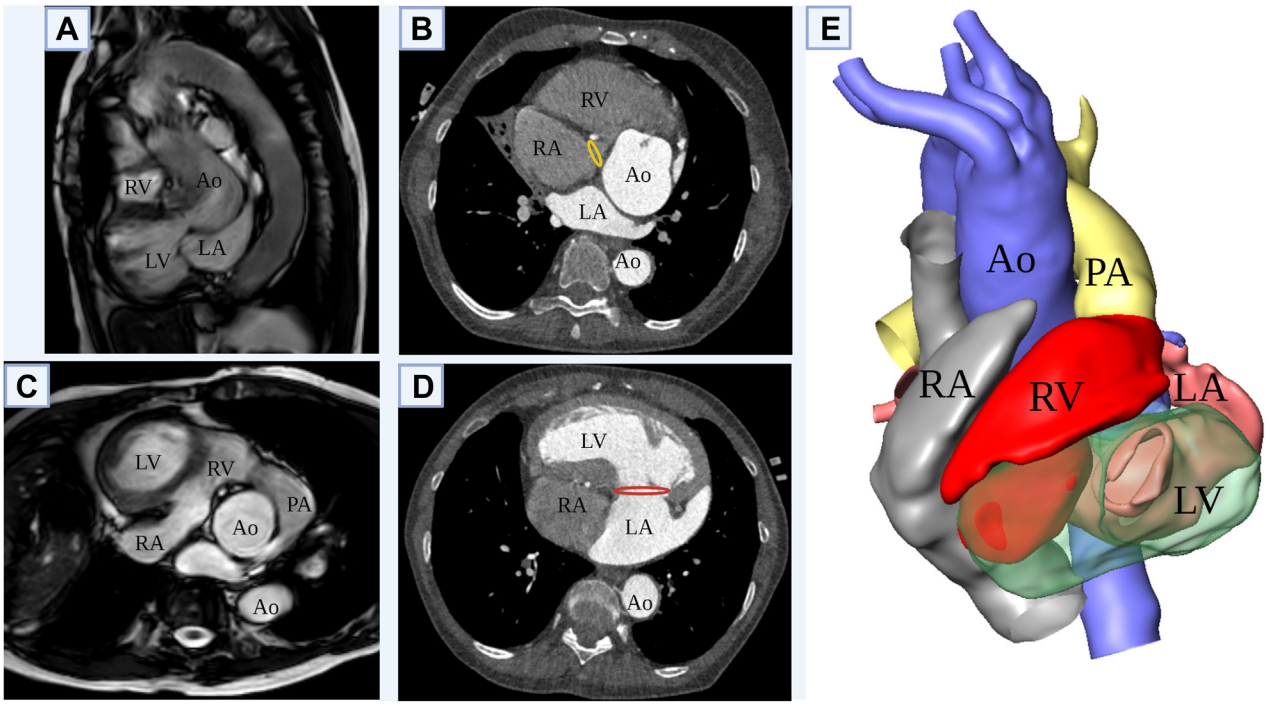
SAVR = surgical aortic valve replacement

TAVR = transcatheter aortic valve replacement

TTE = transthoracic echocardiography

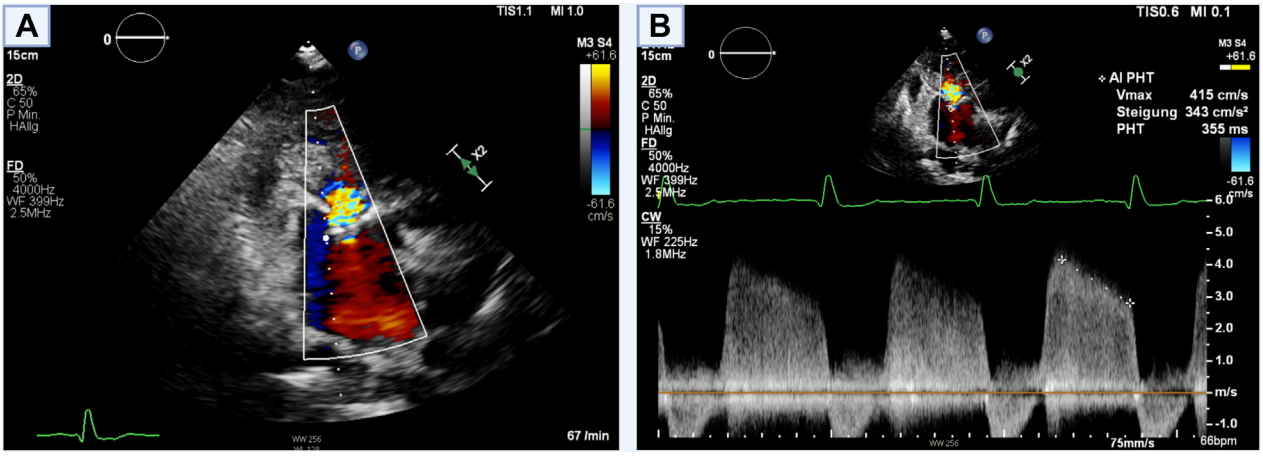
VSD = ventricular septal defect

FIGURE 1 Criss-Cross Heart Anatomy



Magnetic resonance imaging **(A, C)** and computed tomography **(B, D)** showing left and right ventricular relationships and connections as well as the tricuspid **(yellow ring in B)** and mitral valve **(red ring in D)**. **(E)** Three-dimensional volume rendering of the criss-cross heart. Created with BioRender.com. Ao = aorta **(blue)**; LA = left atrium **(pink)**; LV = left ventricle **(green)**; PA = pulmonary artery/trunk **(yellow)**; RA = right atrium **(grey)**; RV = right ventricle **(red)**.

FIGURE 2 Preprocedural Transthoracic Echocardiography of the Aortic Valve



Transthoracic echocardiography revealing a severe aortic valve regurgitation **(A)** and showing the flow profile through the aortic valve **(B)**. Created with BioRender.com.

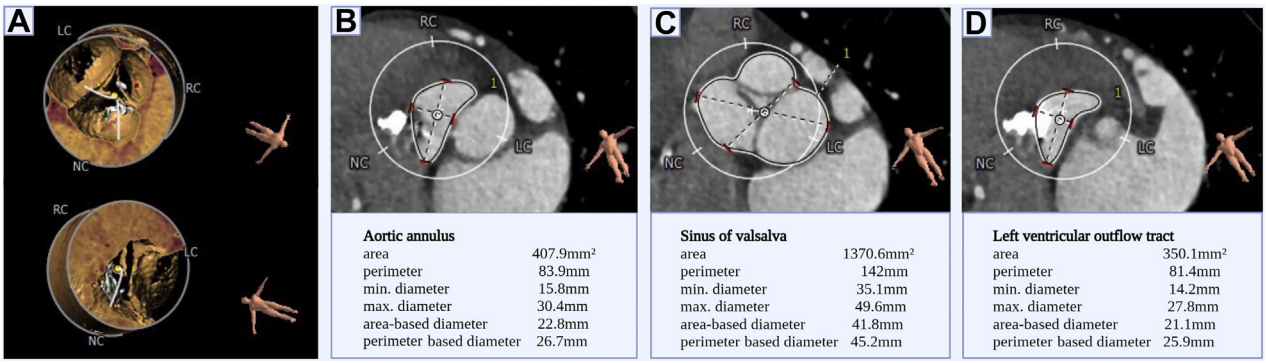
fraction 59%) were ruled out by TTE. Additionally, fluoroscopic imaging of the coronary arteries showed no stenosis of the left coronary artery with atypical anatomy in CCH, and electrocardiography did not reveal any new abnormalities.

INVESTIGATIONS

CT showed an asymmetric, tricuspid aortic valve with an aortic annulus area of 407.9 mm^2, as well as sinus of Valsalva area of 1,370.6 mm^2 and left ventricular outflow tract area of 350.1 mm^2 (Figure 3). Additionally, a right heart catheterization had been performed and showed elevated right ventricular pressures (systolic pressure 40 mm Hg, end-diastolic pressure 12 mm Hg).

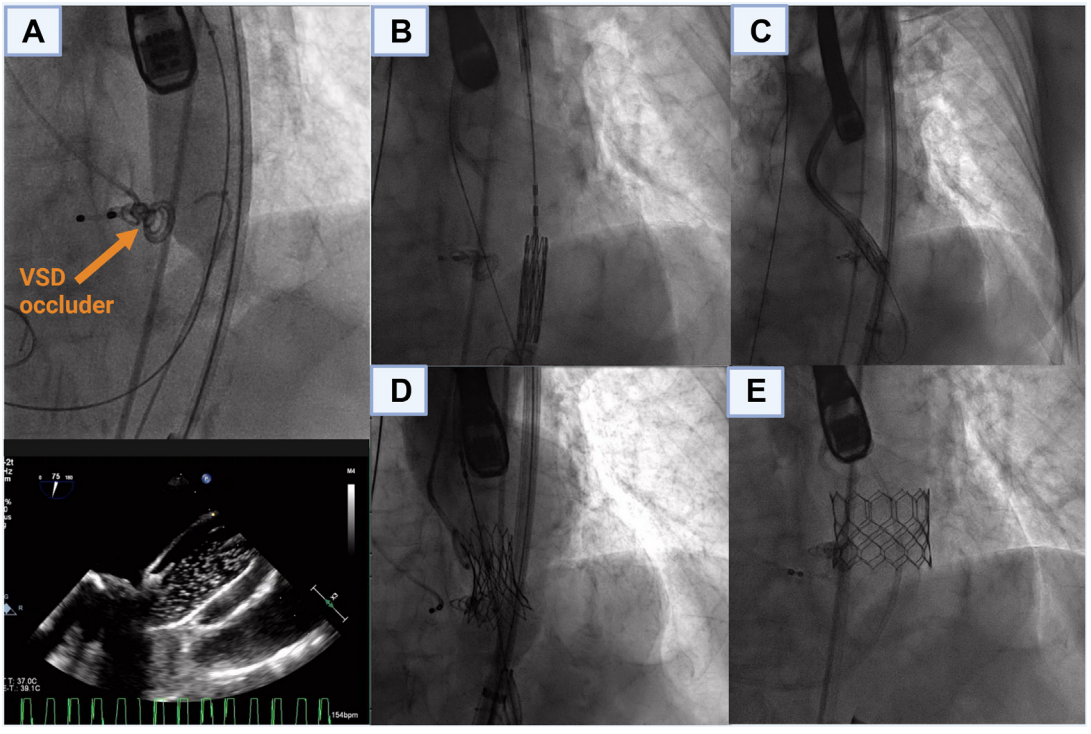
After thorough evaluation, the institutional heart team decided to perform a TAVR due to the challenging anatomy in spite of an estimated low arithmetic surgical risk (EuroSCORE II = 2.61%) and respecting the patient's wish. By implementing CT angiography and 3-dimensional CT reconstruction, the possible access routes and potential prosthetic valve were evaluated. Preference was given to a

FIGURE 3 Preprocedural Computed Tomography of the Aortic Valve, Sinus of Valsalva, and Left Ventricular Outflow Tract



Three-dimensional computed tomography reconstruction of the aortic valve from different views **(A)**; the aortic valve is asymmetric, tricuspid, and slightly thickened with negligible minor calcification. Computed tomography and respective dimensions of the aortic annulus **(B)**, sinus of Valsalva **(C)**, and left ventricular outflow tract **(D)**. Created with BioRender.com.

FIGURE 4 Sequence of the Transthoracic Aortic Valve Replacement



Sizing with a 25-mm balloon **(A)**, stepwise delivery of the prosthetic valve **(B, C)**, implantation during rapid-pacing **(D)**, final aortic valve angiography **(E)**. Close proximity of the ventricular septal defect occluder **(arrow)** and prosthetic valve is noticeable. Created with BioRender.com.

retrograde transfemoral access over a transthoracic or trans-subclavian access route. Because of tortuosity of the retrograde access route, dilatation of the aortic root and possible interference with the VSD occluder, a balloon-expandable strategy with a partially steerable delivery catheter was chosen, in particular the Edwards SAPIEN 3 system (Edwards Lifesciences) with a known high device success rate in pure AR.[2] Contrarily, these circumstances may have hindered the deployment of a self-expandable valve. Furthermore, the narrow left ventricular outflow tract in the presence of an aneurysmatic aortic root complicated the valve size decision, hence had to be made intraprocedurally.

MANAGEMENT

The following specifics were added to the otherwise standard TAVR (**Figure 4,** Video 2): The patient was intubated for the duration of the procedure and transesophageal echocardiography was used throughout the procedure in addition to the angiography to support the valve deployment through the abnormal aorta and heart. As expected, sizing with a 25-mm balloon did not seal the aortic annulus completely, indicating the implantation of a 29-mm valve prosthesis.

Aortic valve sizing is a critical topic for the given anatomy. The circle method is used by the authors on a regular basis,[3] but only for bicuspid stenotic patients—especially with a tapered anatomy. In case of AR, we have not used the circle method yet. In addition, the annulus in this case was very eccentric (16 × 30 mm), which made standard aortic valve sizing rather difficult. Our decision-making was as follows: a 26-mm valve would have been chosen for a regular aortic stenosis patient. However, given the lack of calcification, we opted for a 29-mm SAPIEN 3 valve; to be able to do this in a safe manner, we used balloon sizing in this particular case (25-mm balloon),

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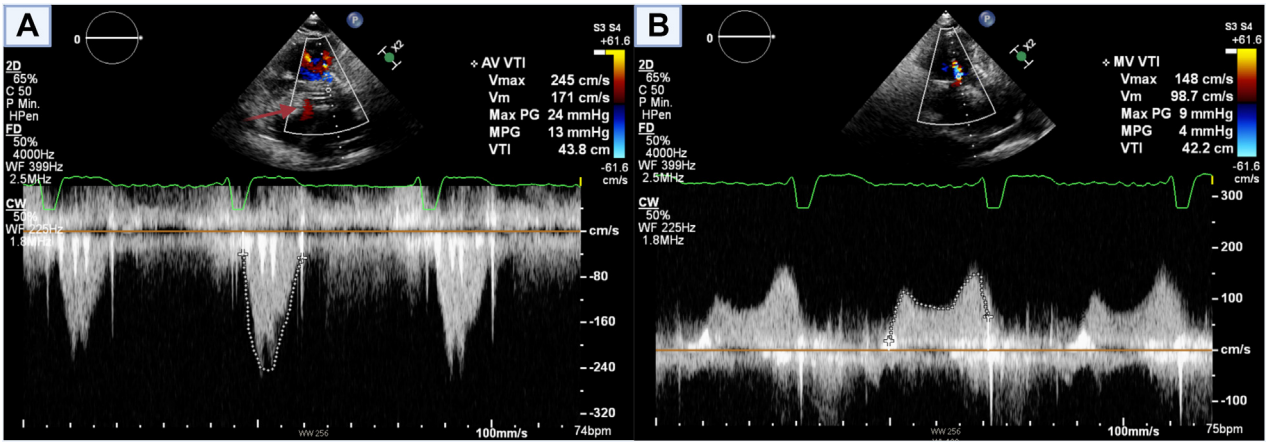
FIGURE 5 Postprocedural Transthoracic Echocardiography



Postprocedural flow through the aortic valve **(A)** and mitral valve **(B)**. Both valves show a remaining minor regurgitation with significantly improved flow profiles compared to the preprocedural imaging. A paravalvular leakage **(arrow)** is also noticeable. Created with BioRender.com.

which led to the impression that a 29-mm bioprosthesis would fit. Figure 4 shows only minor crowing supporting our "up-sizing" choice (29 mm).

After valve deployment, the final angiography and transesophageal echocardiography showed a proper position of the valve without paravalvular leakage and low transvalvular gradients. However, postprocedural TTE and CMR revealed a remaining minor paravalvular leak and a regurgitant fraction of 7% (Figure 5). Nevertheless, the volume flow rate and velocity in the ascending aorta were significantly reduced (Figure 6). No hemodynamic compromise was observed throughout the procedure and no atrioventricular block occurred.

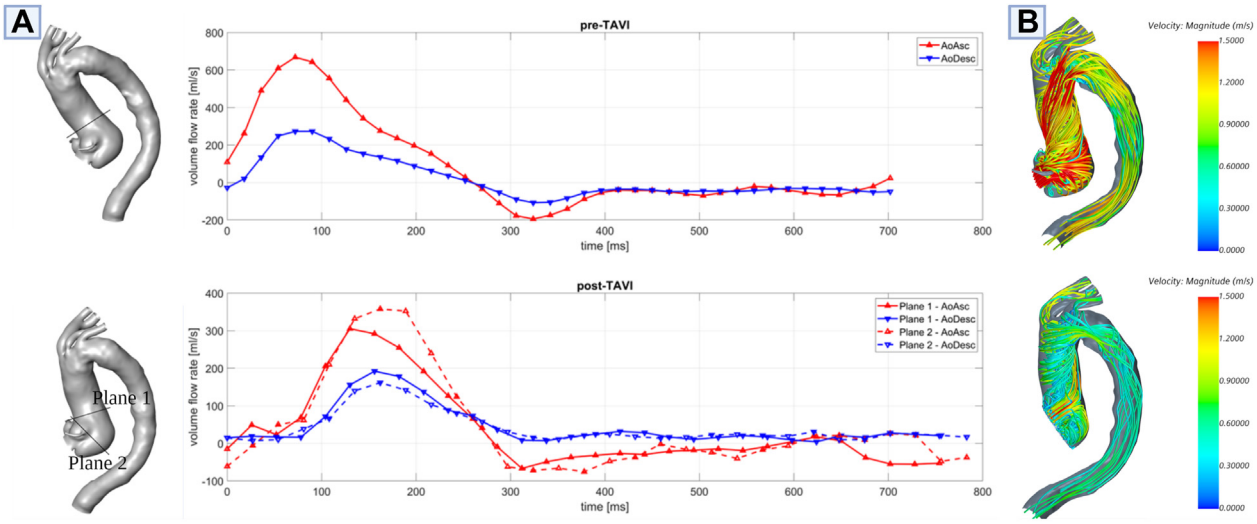
Instantaneous relief of heart failure symptoms was achieved and no complications according to the Valve Academic Research Consortium 3 (VARC-3)[4] besides a minor paravalvular leakage occurred. Ultimately, the patient was discharged home on the third postoperative day with an improved resilience (NYHA functional class I).

DISCUSSION

With the increasing popularity and success of TAVR for the treatment of symptomatic aortic valve stenosis,[5-7] off-label uses of TAVR for the treatment of other valvular diseases such as native aortic valve regurgitation (NAVR) are becoming more common.[5] In fact, TAVR for NAVR showed adequate results in high-risk patients[8] and the Trilogy Valve System (JenaValve Technology) was approved for the treatment of severe NAVR in Europe,[9] although the use of the latter was contraindicated in our patient because of the dilated ascending aorta. Moreover, in their multicenter registry, Yoon et al[2] were able to show that TAVR using the new-generation devices (Evolut R [Medtronic], SAPIEN 3, JenaValve, Lotus [Boston Scientific], Direct Flow [Direct Flow Medical], Acurate [Boston Scientific], Portico [Abbott Cardiovascular], and J-Valve [JC Medical]) was associated with improved procedural outcomes in treating patients with pure native AR compared with the early-generation devices (CoreValve [Medtronic] and SAPIEN XT). Nevertheless, surgical aortic valve replacement (SAVR) remains the therapeutic standard, unless special indications, such as a high estimated surgical risk or hostile surgical access, are given.[6] Additionally, NAVRs are often in the presence of aneurysms of the aortic root and ascending aorta.[8] Depending on the diameter of the annulus and aortic root, commercially available prosthetic valves (maximum diameter of 29 mm) are prone to stray from the correct position, let alone sufficiently reduce the regurgitation. Therefore, if the diameter of the ascending aorta exceeds 45 mm, surgery of the aortic root or ascending aorta in addition to SAVR is required.[6]

On the other hand, surgical intervention in patients with complex anatomical anomalies such as CCH is especially challenging, but is not taken into account by current risk models such as the EuroSCORE II or STS PROM (Society of Thoracic Surgeons Predicted Risk of Mortality). Although, in these patients, a better view of the anatomy and surgical field

FIGURE 6 **Preprocedural and Postprocedural Volume Flow Rates**



Based on 4-dimensional cardiac magnetic resonance imaging, preprocedural **(A, top)** and postprocedural **(A, bottom)** volume flow rates through the ascending aorta **(red)** and descending aorta **(blue)**, as well as preprocedural **(B, top)** and postprocedural **(B, bottom)** flow velocities through the aorta were estimated. Created with BioRender.com. TAVI = transcatheter aortic valve implantation.

as given by a conventional approach may be beneficial, the anomalous anatomy in such patients may hinder the feasibility of SAVR. In this particular case, the ascending aorta and aortic root were both borderline dilated, indicating SAVR; however, because of the eccentric and narrow annulus, as well as a possible interference with the VSD occluder, a transcatheter approach was preferred. Moreover, the aortic root and valve were difficult to reach due to the unusual superior position of the right ventricle, hence SAVR was deemed futile, and a TAVR was performed instead.

Because patients with CCHs may differ strongly in anatomical properties,[1,10] SAVR may be feasible and indeed preferable in other cases such as patients with side-by-side ventricular arrangement. Furthermore, CCH is often accompanied by various additional congenital malformations, thus concomitant procedures may be required. For these reasons, meticulous preoperative imaging is necessary regardless of the procedure.

FOLLOW-UP

Three months after the procedure, CMR confirmed the proper function of the prosthetic valve with a remaining minor regurgitation of 3% and showed a significantly reduced left ventricular congestion (end-diastolic volume 79 mL).

CONCLUSIONS

In an inoperable patient with challenging anatomy, TAVR was successful in treating the symptomatic NAVR in spite of complicating factors such as the abnormal aortic annulus and left atrioventricular relationship, the aneurysmatic aorta, no valvular calcifications, or the VSD occluder. In this patient with a complex congenital heart disease, extensive evaluation using different imaging and discussion in an interdisciplinary team was crucial in deciding the appropriate management, including selection of the intervention, approach, and prosthetic valve, as well as excluding necessary concomitant procedures.

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KEY WORDS aortic valve regurgitation, criss-cross heart, TAVI, TAVR

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