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CASE REPORT

ADVANCED

CLINICAL CASE: TECHNICAL CORNER

Transcatheter Systemic AV Valve-in-Valve Implantation in a Patient With LTGA and Ebstein Anomaly

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ABSTRACT

We present the first in-human transcatheter systemic atrioventricular valve-in-valve implantation in a 37-year-old patient with Ebstein anomaly, levo-transposition of the great arteries, and prior systemic valve replacement. She had severe bioprosthetic valve regurgitation and reduced systolic function. She had high surgical risk and was planned for transcatheter intervention. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:551-555) © 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/).

CASE PRESENTATION

A 37-year-old woman with levo-transposition of the great arteries (L-TGA) and Ebstein anomaly of the tricuspid valve (TV) underwent systemic (tricuspid) valve replacement with a 29 mm Carpentier-Edwards Perimount valve 10 years prior. Her medical history was significant for complete heart block requiring a dual-chamber transvenous pacemaker, chronic systolic heart failure (New York Heart Association functional class III), and a history of metastatic adenoid cystic carcinoma of the skull base with metastasis to the lungs and lymph nodes (stage IV). She presented with worsening shortness of breath and decreasing exercise tolerance over the prior year. Echocardiogram revealed a dilated systemic (right) ventricle with a severe decrease in systolic function (right ventricular ejection fraction: 35%) and severe bioprosthetic valve degeneration with regurgitation (Video 1). Because of her comorbidities and surgical history, the patient was deemed high risk for surgical valve replacement and referred for transcatheter valve-in-valve implantation after discussion with the congenital heart team.

LEARNING OBJECTIVES

- To describe the clinical evaluation, multimodality imaging, and preprocedural planning involved in transcatheter atrioventricular valve-in-valve implantation in a patient with levo-transposition of the great arteries and Ebstein anomaly.
- To describe the procedural details of transcatheter systemic atrioventricular valve-in-valve implantation in a patient with levo-transposition of the great arteries and Ebstein anomaly.

PROCEDURE DESCRIPTION

The procedure was performed under general anesthesia in a biplane angiography suite. After obtaining bilateral femoral venous access, heparin and

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ABBREVIATIONS AND ACRONYMS

AV = atrioventricular

L-TGA = levo-transposition of the great arteries

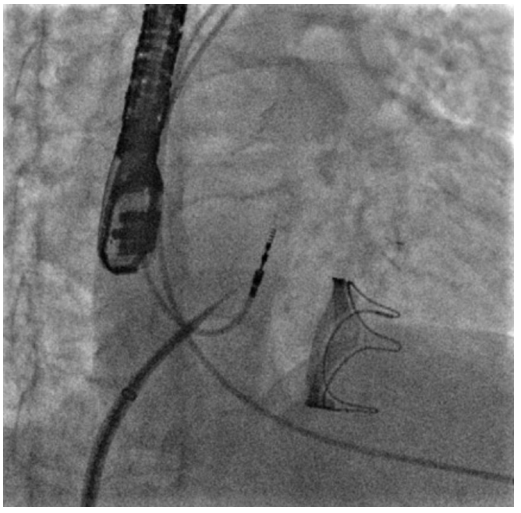
TEE = transesophageal echocardiogram

TV = tricuspid valve

antibiotics were administered. Transesophageal echocardiogram (TEE) imaging was used to guide transseptal puncture with a BRK-1 needle and SL0 sheath, with special care taken to avoid interfering with the pacemaker leads (**Figure 1**). The puncture site was selected to be level with the midline of the bioprosthetic valve on fluoroscopy in the 20° right anterior oblique projection and at the inferior edge of the fossa ovalis on TEE (Video 2). Once the septum was crossed, baseline hemodynamics were obtained that were significant for V waves of 56 mm Hg and a mean pressure of 32 mm Hg (**Figure 2A**).

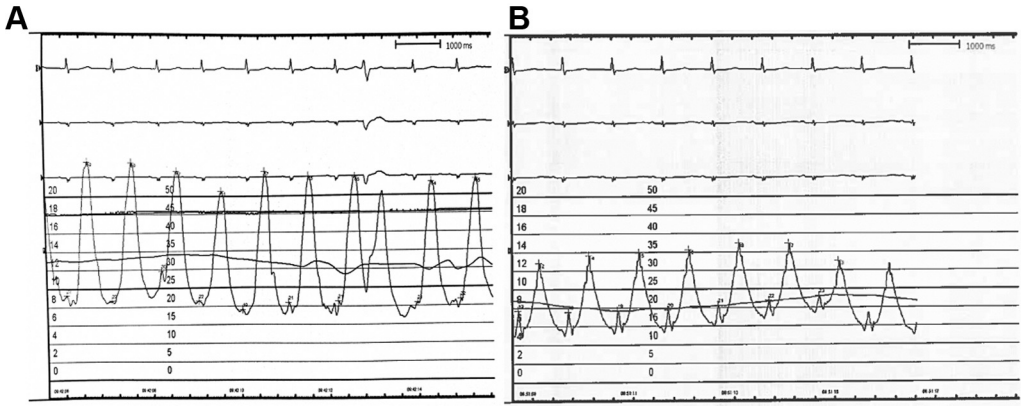
A left internal mammary artery catheter was advanced into the left atrium and used to introduce a Confida wire (Medtronic) into the right ventricular apex. After predilating the septum with the hydrophilic transcatheter valve delivery system dilator, a Z-Med II 10 × 3 cm balloon (B. Braun Medical, Inc) was passed over the wire and used to further dilate the septum under TEE guidance (**Figure 3,** Video 3). Through contralateral venous access, a balloon-tipped pacing catheter was advanced into the morphologic left ventricle for rapid pacing during valve deployment. A 29-mm Edwards Sapien S3 valve (Edwards Lifesciences) was prepared so that the skirted inflow of the valve was on the atrial side or proximal on the delivery system and advanced through the sheath. The system was inverted such that flexing the delivery catheter curved to the patient's left. The valve was advanced across the septum and into the surgical valve while adding torque to facilitate crossing and avoid getting caught on the septum. Once in proper position, the heart was rapidly paced at 180 beats/min and the valve balloon inflated slowly to deploy the valve (Video 4). After implantation, the valve regurgitation was noted to have dropped to trace on TEE with normal working leaflets (**Figure 4,** Videos 5 and 6). Mean left atrial pressure dropped immediately after implant to 18 mm Hg with V waves of 28 mm Hg (**Figure 2B**). The sheath was removed and Perclose sutures (Abbott)

FIGURE 1 Fluoroscopic Image of Performing Transseptal Puncture



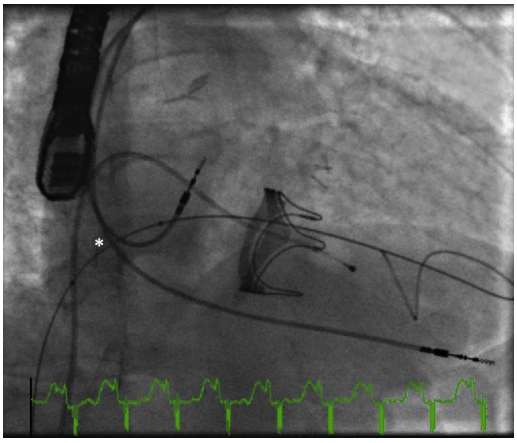
Note the position of the needle relative to the transvenous pacing leads and bioprosthetic valve.

FIGURE 2 Invasive Left Atrial Pressure Measurements Showing Marked Improvement Post-Valve Implantation



Left atrial pressure tracing obtained through transseptal puncture before **(A)** and after **(B)** valve implantation.

FIGURE 3 Dilation of Atrial Septum With 10 × 3 mm Balloon



Note the coaxial wire position relative to the bioprosthetic valve, the wire positioned in the right ventricular apex, and the balloon-tipped pacing catheter in the left ventricle. *Position of septal dilation balloon.

tightened to achieve hemostasis. The atrial septal perforation was not closed postimplantation. Follow-up care and an echocardiogram at 1 year revealed normal valve function with no significant regurgitation.

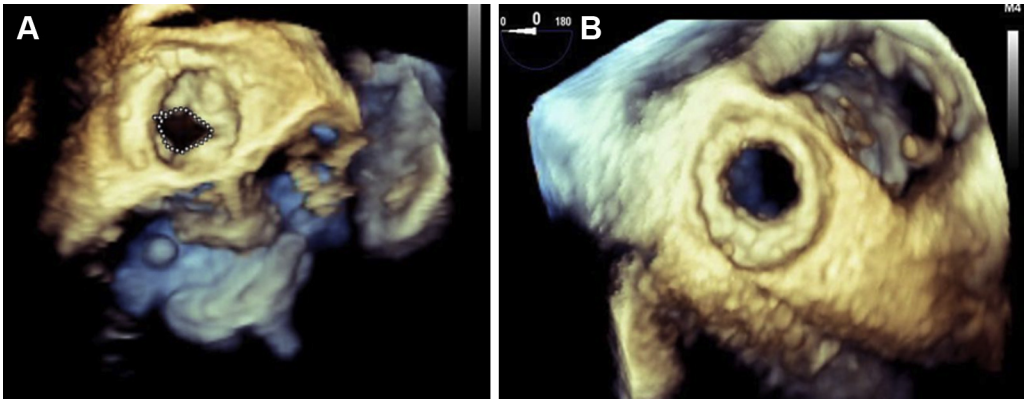
PROCEDURAL CONSIDERATIONS

Preprocedural planning and anatomical assessment are key steps before undertaking transcatheter TV replacement. When performing a valve-in-valve implantation in the mitral position of a normal heart, care must be taken to assess the distance between the surgical valve and the opposite ventricular wall, as well as anticipated residual ventricular outflow tract area, to avoid an outflow obstruction. This is due to the aorto-mitral continuity and proximity of the anterior aspect of the systemic AV valve to the left ventricular outflow tract and aortic valve in a congenitally normal heart. Conversely, patients with L-TGA have greater separation between the right AV valve and the outflow tract and, therefore, a much lower risk of outflow tract obstruction. In our patient, the cardiac computed tomography imaging-predicted distance from the valve to the outflow tract after implantation was 22 mm, which made outflow tract obstruction highly unlikely (Figure 5) as did the predicted outflow tract area of 800 mm2 postimplantation.

The location of transseptal puncture was critical for procedural success. The transseptal puncture was performed at a low point on the septum: the inferior edge of the fossa ovalis and coaxial to the valve plane to provide a more streamlined valve delivery. Care was taken to pass the catheter under the pacemaker lead and up the superior vena cava before pulling back to avoid disrupting the pacemaker lead located in the right atrial appendage.

The atrial septum can be more thickened in patients who have previously undergone surgical AV valve replacement as the septum is accessed for exposure to implant the valve and then surgically closed after. The long, gently tapered hydrophilic

FIGURE 4 3-Dimensional Transesophageal Echocardiogram Images Pre- and Post-Valve-in Valve-Implantation



(A) 3-dimensional transesophageal echocardiogram image of degenerated bioprosthetic valve with decreased opening area. **(B)** Significantly improved opening area after valve-in-valve implantation.

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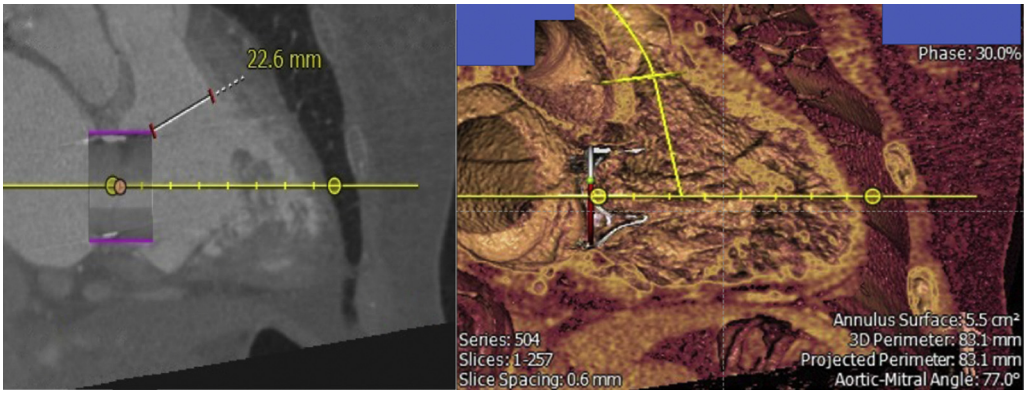
FIGURE 5 Cardiac Computed Tomography Image Showing the Distance From the Predicted Valve Implant Location to the Septum (22 mm)



dilator present in the valve kit was used to predilate the septum because it has superior crossing characteristics. Carefully predilating the septum before advancing the valve ensures that the uncovered valve does not get caught on the thickened septum, which can result in significant complications. Recently, large-bore Gore Dry-Seal sheathes (65 cm, 26-F) (Gore and Associates) have been used to protect the TV and facilitate delivery of the transcatheter valve to intended implantation sites in patients with dysfunctional right ventricular outflow tracts.[1,2] Anchoring the guidewire in the right ventricle presents a challenge in that the morphologic right ventricular trabeculations prevent the coiled wire from reaching the apex, limiting the landing zone for the delivery sheath and nose-cone.

DISCUSSION

Transcatheter TV replacement for patients with severe native TV dysfunction has evolved over the past few years, with various patient selection and procedural aspects as previously described.[3,4] Successful valve-in-valve replacements in prior dysfunctional surgical valves have been reported in the international multicenter Valve-in-Valve International Database registry,[5] but knowledge and experience are limited for these interventions in patients with complex congenital heart disease.[6] Eicken et al[7] reported the first case of percutaneous tricuspid valve-in-valve implantation for the treatment of systemic bioprosthetic AV-valve dysfunction in a patient after an atrial switch operation for transposition of the great arteries. To our knowledge, the current patient is the first reported case of a systemic AV valve-in-valve transcatheter intervention in a patient with Ebstein anomaly and L-TGA.

Limited surgical options are available in patients with complex congenital anatomy and previous surgical valve replacement. Redo TV replacement and repair have been shown to have significant operative mortality and complications.[8] Surgical TV repair was also noted to have a not-insignificant degree of recurrent regurgitation, with >14% of patients having moderate or severe regurgitation 1-week post-procedure.[8,9] Higher preoperative regurgitation grade and poor systemic ventricular function were noted to be risk factors for worsening tricuspid regurgitation after the procedure. Considering this patient's cardiac history and poor ventricular function, surgical intervention was considered a higher risk, and the transcatheter approach yielded an optimal outcome.

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Dr Abudayyeh is a physician advisor for Edwards Lifesciences; and is an education consultant for Gore. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS **bioprosthetic valve degeneration, Ebstein anomaly, levo-transposition of the great arteries, systemic atrioventricular valve, transcatheter valve-in-valve replacement**

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