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CASE REPORT

ADVANCED

CLINICAL CASE

Self-Expanding Transcatheter Pulmonary Valve Implant in the Right Pulmonary Artery

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ABSTRACT

Newer self-expanding transcatheter pulmonary valves (TPVs) are approved for the treatment of severe pulmonary regurgitation in patients with large right ventricular outflow tracts. We present a patient with Tetralogy of Fallot whose right ventricular outflow tract was too large for self-expanding TPV, who was treated successfully with a self-expanding TPV in the right pulmonary artery. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;14:101823) © 2023 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/).

Treatment of chronic pulmonary insufficiency using transcatheter pulmonary valve replacement (TPVR) in the branch pulmonary artery position has been well-described since the first case report of a Melody (Medtronic) TPVR in 2011.[1] In 2021, the Harmony Transcatheter Pulmonary Valve (Medtronic) received Food and Drug Administration approval for the treatment of pulmonary regurgitation (PR) in patients with native or patched right ventricular outflow tracts (RVOTs). There are 2 sizes available, which are named according to the diameter of the respective valves (ie, 22-mm self-expanding transcatheter pulmonary valve [TPV22] and 25-mm self-expanding transcatheter pulmonary valve [TPV25]). We report the first case of branch pulmonary artery implantation of this self-expanding transcatheter pulmonary valve (TPV) in the single right pulmonary artery (RPA) position.

LEARNING OBJECTIVES

- To review novel approaches to TPVR in patients with dilated patched or native RVOTs.
- To discuss technical aspects of branch PA implantation of newer self-expanding transcatheter valves.
- To review historical development of TPVR devices and novel techniques for delivery.

HISTORY OF PRESENTATION

A 41-year-old male with Tetralogy of Fallot (ToF) underwent modified Blalock-Taussig-Thomas shunt at 6 months of age followed by complete repair at 6 years of age. He developed left pulmonary artery (LPA) isolation that could not be corrected at time of his complete repair. Many years later he was referred to adult congenital cardiology with symptoms of worsening dyspnea on exertion and generalized fatigue with functional limitations consistent with New York Heart Association functional class III.

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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.

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ABBREVIATIONS AND ACRONYMS

LPA = left pulmonary artery

MPA = main pulmonary artery

PR = pulmonary regurgitation

RPA = right pulmonary artery

RV = right ventricle

RVEDV = right ventricular end-diastolic volume

RVOT = right ventricular outflow tract

TPV = transcatheter pulmonary valve

TPV22 = 22-mm self-expanding transcatheter pulmonary valve

TPV25 = 25-mm self-expanding transcatheter pulmonary valve

ToF = Tetralogy of Fallot

TPVR = transcatheter pulmonary valve replacement

MEDICAL HISTORY

The patient had ToF, right aortic arch with mirror image branching, status post left modified Blalock-Taussig-Thomas shunt at 6 months, and transannular patch repair at age 6 years. He developed LPA isolation with associate left lung hypoplasia. Other comorbidities included hypertension, obesity (body mass index: 41.4 kg/m^2), mixed moderate restrictive and obstructive lung disease, and depression.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis was right ventricular (RV) dilation secondary to sequelae from chronic severe PR, pulmonary hypertension, and undiagnosed cardiomyopathy.

INVESTIGATIONS

Standard 12-lead electrocardiogram revealed normal sinus rhythm, right bundle branch block (QRS duration 186 ms). Echocardiogram showed a dilated RV and mild tricuspid regurgitation. Cardiac magnetic resonance imaging demonstrated a severely dilated RV with right ventricular end-diastolic volume (RVEDV) index of 179 mL/m^2, RV end systolic volume of 96 mL/m^2, and an RVEDV:left ventricular end-diastolic volume ratio of 2.5. There was mild biventricular dysfunction and severe pulmonary regurgitation (regurgitant fraction: 80%). Discontinuity of the LPA from the main pulmonary artery (MPA) was confirmed along with the presence of numerous small aortopulmonary collaterals feeding the left lung.

MANAGEMENT

The patient met criteria for pulmonary valve replacement. Given his comorbidities and desire to avoid surgical intervention, the patient was evaluated for self-expanding TPV implantation. Standard multidetector computed tomography (CT) with retrospective gating was performed per protocol. The patient was deemed not a candidate for RVOT placement due to the lack of necessary contact of the valve stent apparatus to the RVOT in both systole and diastole (**Figure 1A**). The patient was re-evaluated for RPA placement and was deemed a candidate for both TPV22 and TPV25 (**Figure 1B**). Based on the preoperative CT scan fit analysis and the angiogram performed during the case, the decision was made to implant a TPV22 (Video 1).

The TPV22 was prepped and loaded onto the proprietary delivery system following the manufacturer's instructions. The valve delivery system was advanced through a 26-French DrySeal Flex Introducer Sheath (Gore) until positioned just distal to the landing zone. The valve was deployed with careful fluoroscopic and angiographic assessment of the first, second, and then final row of struts. Angiography confirmed proper positioning of the valve apparatus, and so it was released from the delivery system (**Figure 2**). A postimplantation angiogram showed no valvar or perivalvular leak and good apposition to the vessel wall (Video 2). Peak gradient from distal RPA to MPA/RV was 6 mm Hg.

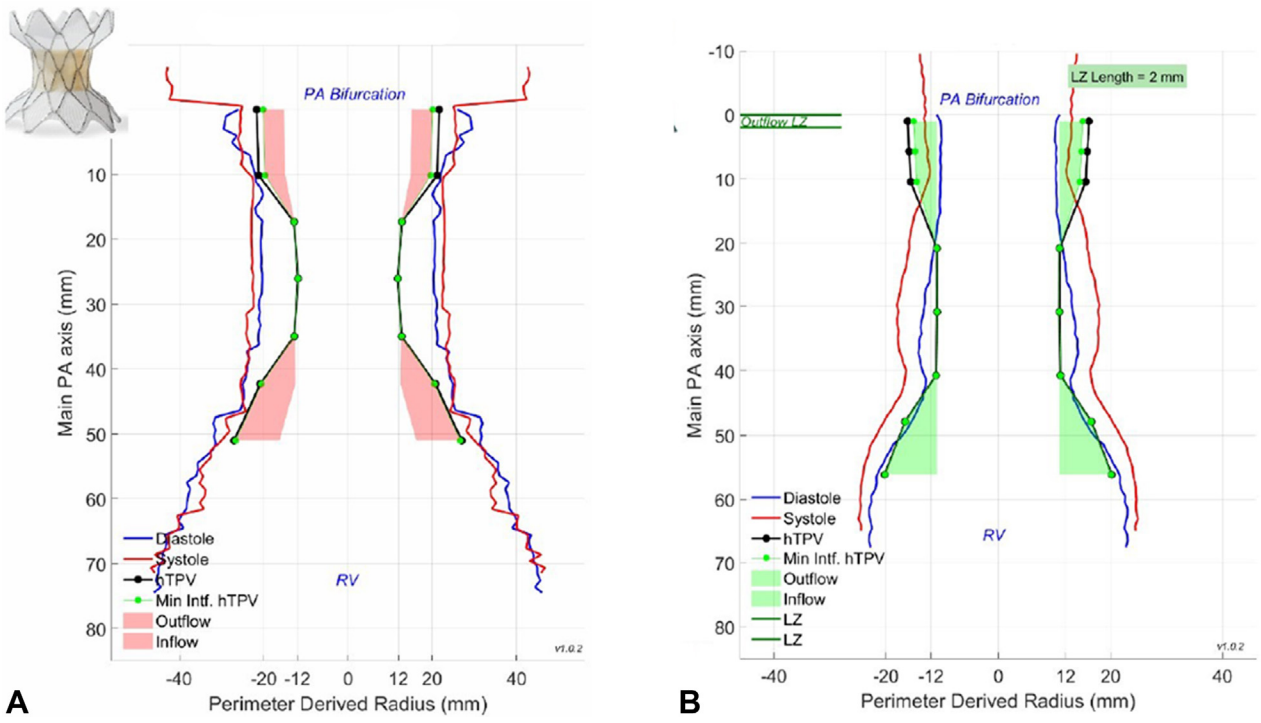
DISCUSSION

Patients who have undergone transannular patch repair of TOF or pulmonary valvotomy/balloon valvuloplasty for congenital pulmonary stenosis often develop chronic severe PR that leads to dilation of the RV and the RVOT/MPA. For these patients there are now multiple Food and Drug Administration-approved transcatheter valves, which can be used for percutaneous intervention.

However, despite these recent advancements, not all patients are good candidates for traditional TPVR and alternative approaches should be considered. One such approach is TPV placement in the branch pulmonary artery location, which was first described in an ovine model in 2011.[1] Gillespie et al[2] were the first to demonstrate success of this procedure in humans the same year. Subsequently, Qureshi et al[3] reported successful TPVR implantation in the branch pulmonary arteries in 32 of 34 patients with oversized RVOTs with reduction in RVEDV in 2018. Bansal et al[4] reported significant reduction of RVEDV using cardiac magnetic resonance imaging after successful TPVR implantation in the branch pulmonary arteries of 7 patients.[4] These studies showed TPVR in the branch pulmonary arteries could be a viable alternative to surgical valve replacement in certain high-risk patients. The second alternative would be to use a hybrid approach for TPVR, involving surgical plication of the MPA or RVOT to create an adequate landing zone for one of the available valves, as described by Sosnowski et al.[5]

For our patient we chose to place the TPV22 in the proximal RPA and did so with excellent outcome when his RVOT proved to be too large even for the TPV25. The TPV22 is a porcine pericardial tissue valve mounted on a self-expanding nitinol frame. The TPV22 has an outer diameter of 41 mm at the proximal

FIGURE 1 **TPV Perimeter Plot**

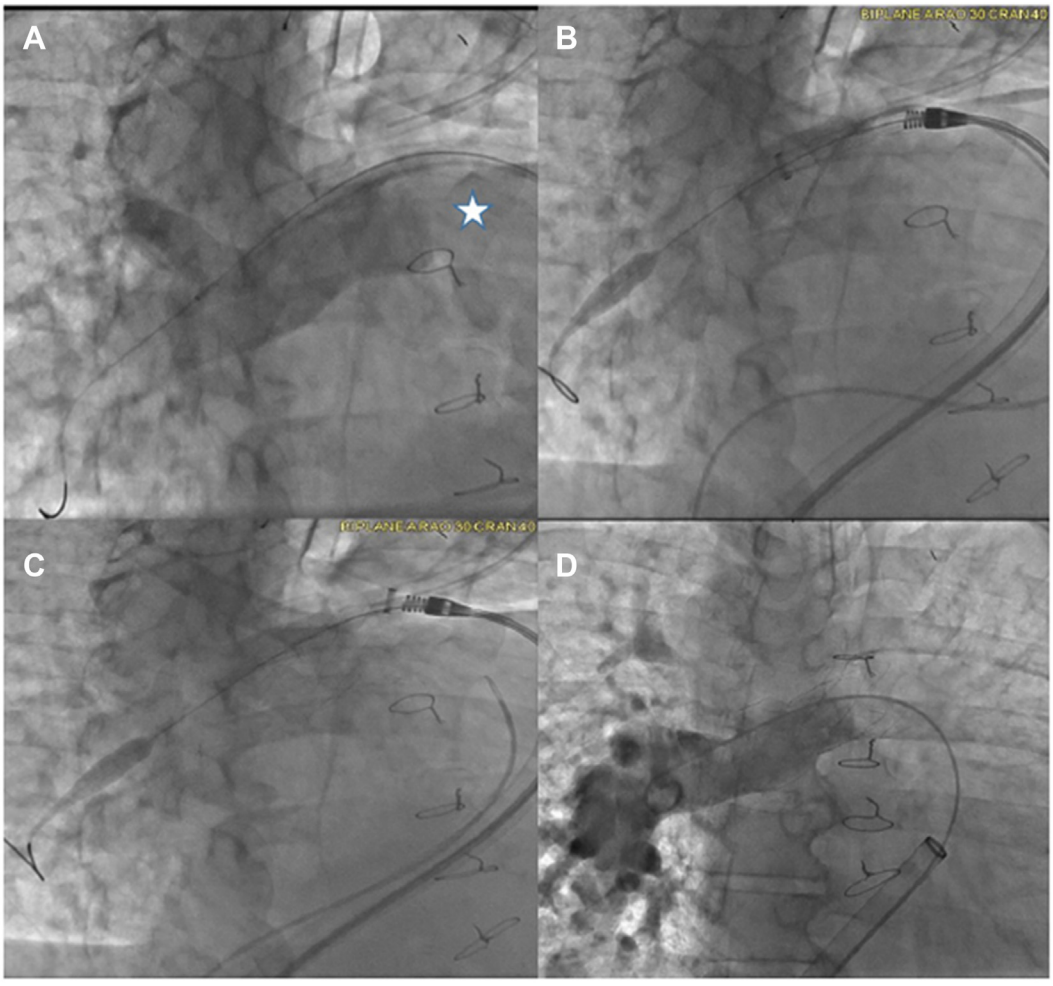


TPV Perimeter Plot (created using 3mensio Structural Heart, Pie Medical Imaging) for the TPV25 in the RVOT position showing minimal expected interference between the valve frame and RVOT **(A)** and for the TPV22 in the RPA position showing adequate interference between valve frame and branch pulmonary artery **(B)**. Reproduced with permission of Medtronic. The TPV25 is intended to be implanted in a section of the main PA (between the RVOT and main PA bifurcation). The device manufacturer has allowed use of the perimeter plot in this article to assist the authors in describing their clinical rationale for using the device off-label. The TPV25 manufacturer does not promote the use of its products outside the IFU. IFU = indication for use; PA = pulmonary artery; RPA = right pulmonary artery; RVOT = right ventricular outflow tract; TPV = transcatheter pulmonary valve; TPV22 = 22-mm self-expanding transcatheter pulmonary valve; TPV25 = 25-mm self-expanding transcatheter pulmonary valve.

end, 23.5 mm at the valve housing, 32 mm at the distal end, and is 55 mm in length. The proprietary delivery system for both the TPV22 and TPV25 is a 25-F coil-loading catheter with an integrated sheath.[6] Although both the TPV22 and TPV25 devices could have been used based on the fit analysis provided by the manufacturer, our angiograms showed less pulsatile dilation of the distal portion or the RPA and our concern was that the TPV25 would be oversized distally at the nominal diameter of 43 mm. We also considered placing a 29-mm Sapien S3 (Edwards Lifesciences) in the RPA. However, our concern for this device was that the proximal portion of the valve apparatus would not have good apposition with the vessel wall, which measured consistently >30 mm in systole, posing an extremely high chance of significant perivalvular leak. Therefore, the TPV22 was selected.

Following a standard hemodynamic cath and angiographic evaluation, a 26-F long sheath was advanced over a stiff wire to the proximal RPA. It was fairly difficult to advance the large and relatively stiff large bore sheath to that position and took considerable inward force and coordination between both sheath and wire operators. However, advancement of the TPV22 delivery system through the sheath to the bifurcation point of the RPA was not difficult having already obtained such distal position with the large bore sheath. Taking the time and effort to get the long 26-F sheath into the proximal RPA was key as the turn from MPA to RPA was acute and could have posed difficulty to advancing the delivery system around the turn on its own. We thought taking the time to get sheath position in the RPA would be safer than having to put excessive inward pressure on the valve delivery

FIGURE 2 Step-Wise Delivery



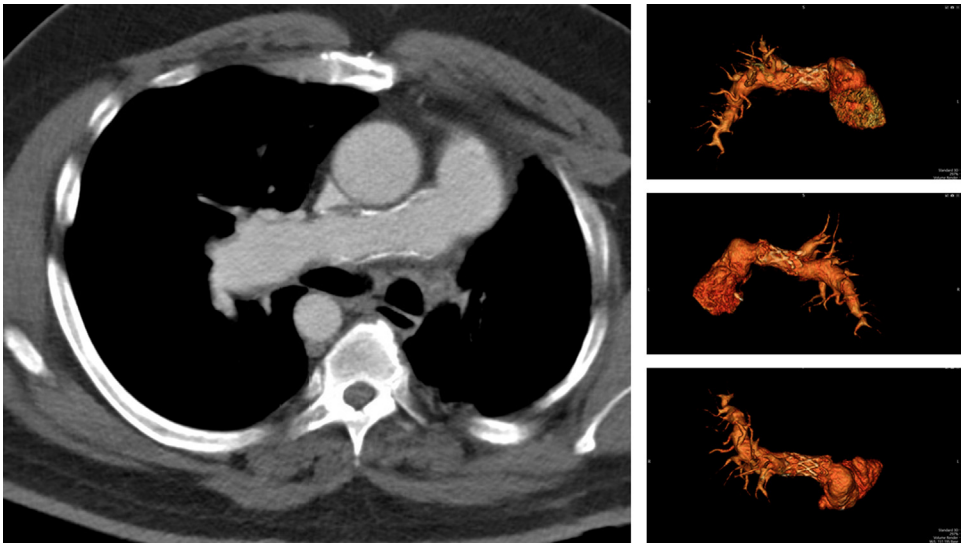
Step-wise delivery of the TPV22. **(A)** Dilated RPA and MPA with free PR **(star)**. The valve is unsheathed **(B)** and released **(C)**. The postdelivery angiogram shows a competent valve in the RPA position **(D)**. MPA = main pulmonary artery; PR = pulmonary regurgitation; other abbreviations as in Figure 1.

system. Unsheathing the TPV22 was also very difficult and took considerable inward force on the inner shaft while trying to unsheathe the valve in a controlled fashion as not to lose our distal position at the takeoff of the right upper pulmonary artery. Although the operators' experience with this valve was limited at the time with this being just the third case, the experience or the RPA implantation sounds similar to what other have described with horizontal deployments into RVOTs that have acute turns as it dives posteriorly. Considerable attention and slow movements are necessary to maintain the distal end of the valve within the desired landing zone.

FOLLOW-UP

Our patient experienced immediate improvement in symptoms at baseline and with exertion. At 4 months postimplantation, his echocardiogram revealed no regurgitation or perivalvular leak and peak gradient of 12 mm Hg. However, it is useful to note the limited utility of transthoracic echocardiography to assess valves implanted in the pulmonary arteries, especially with larger adult patients, and a high parasternal view provides the best alignment for color Doppler interrogation. CT scan at 8 months showed the valve apparatus nicely with delineation of the right upper lobe pulmonary

FIGURE 3 Still Frame and 3D CT Reconstruction



Still frame and 3D CT reconstruction shows the well-seated TPV22 in the RPA 9 months after implantation. 3D = 3-dimensional; CT = computed tomography; other abbreviations as in Figure 1.

artery patency clearly demonstrated (Figure 3). A cardiac magnetic resonance imaging at 9 months postimplantation showed the valve well seated in the RPA with good function, improved RV volumes (RVEDV, indexed 128 mL/m^2, RVEDV:left ventricular end-diastolic volume 1.7), and improved LV function.

CONCLUSIONS

This is the first ever implantation of a self-expanding transcatheter valve in the branch pulmonary artery position. Although the deployment is technically difficult, self-expanding TPV implantation in a branch pulmonary artery represents a potential alternative to surgery when standard RVOT implantation is not possible.

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KEY WORDS congenital heart defect, pulmonic valve, Tetralogy of Fallot, valve replacement

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