

CASE REPORT

ADVANCED

CLINICAL CASE

Transcatheter Valve Implantation in Reversed Potts Shunt in Pulmonary Arterial Hypertension

Keeping the Shunt Reversed



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ABSTRACT

The reversed Potts shunt is designed to offload the right ventricle in severe pulmonary arterial hypertension. We present a case of bidirectional flow across a reversed Potts shunt leading to pulmonary edema, with clinical improvement after implantation of a transcatheter valve in the shunt to maintain unidirectional flow. **(Level of Difficulty: Advanced.)**

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A 19-year-old woman with idiopathic pulmonary arterial hypertension (iPAH) after surgical valved reversed Potts shunt (rPS) placement presented in a decompensated state with hypoxemia requiring continuous oxygen supplementation, pulmonary edema, and right ventricular (RV) failure. On examination, the patient was

underweight. Cardiac examination was significant for a loud P2 auscultated at the apex, 3/6 systolic ejection murmur heard throughout precordium, and a palpable RV lift. No lower extremity edema or jugular vein distension observed. Crackles were appreciated bilaterally at the mid-lower lung fields posteriorly. No ascites or organomegaly was noted. She was transferred to the intensive care unit with invasive monitoring for inotropic and pressor support, and for adjustment of pulmonary vasodilators to reduce left-to-right shunting.

LEARNING OBJECTIVES

- To understand the role of a reversed Potts shunt in idiopathic pulmonary arterial hypertension.
- To understand the potential detrimental effect of left-to-right shunting across a reversed Potts shunt.
- To understand the novel technique of implanting a transcatheter bovine jugular valve in a reversed Potts shunt to maintain unidirectional flow.

MEDICAL HISTORY

The patient was diagnosed at the age of 4 years with iPAH, and underwent placement of a surgical valved rPS, from the left pulmonary artery (LPA) to the descending aorta (DsAo), at the age of 16 years. The valved rPS is designed to reduce RV afterload by allowing right-to-left shunting, minimizing cyanosis

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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****DsAo** = descending aorta**iPAH** = idiopathic pulmonary
arterial hypertension**PA** = pulmonary artery**rPS** = reversed Potts shunt**RV** = right ventricular

of the upper body. The valve prevents left-to-right shunting and pulmonary overcirculation.^{1,2} In her specific case, the valved rPS was fashioned with the use of a 10-mm Fusion graft (Getinge), and a 12-mm valved Contegra conduit (Medtronic) that was trimmed to a length shorter than the graft and positioned in the middle of it, leaving the distal edges of the graft intact at the smaller diameter.

DIFFERENTIAL DIAGNOSIS

The clinical history, presentation, and physical examination findings were indicative of pulmonary edema. The differential diagnosis included acute left heart failure, decompensated RV failure, a primary pulmonary process such as alveolar hemorrhage or infection, or pulmonary overcirculation due to left-to-right flow across the rPS.

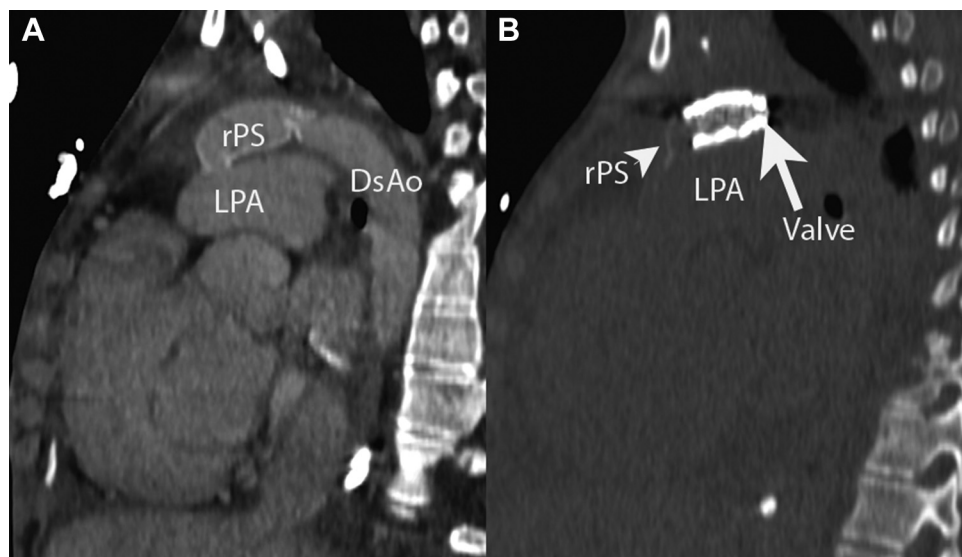
INVESTIGATIONS

Echocardiography demonstrated normal left ventricular size and function, severe RV dilation and dysfunction, and RV systolic pressure of 102 mm Hg (systolic blood pressure 119 mm Hg). New bidirectional flow across the valve within the rPS was noted. Computed tomographic imaging with contrast

demonstrated the rPS to be patent, measuring 12 mm at the midsection and 9-10 mm at the proximal and distal edges, and pulmonary edema was seen (Figure 1). Infection investigation was unremarkable.

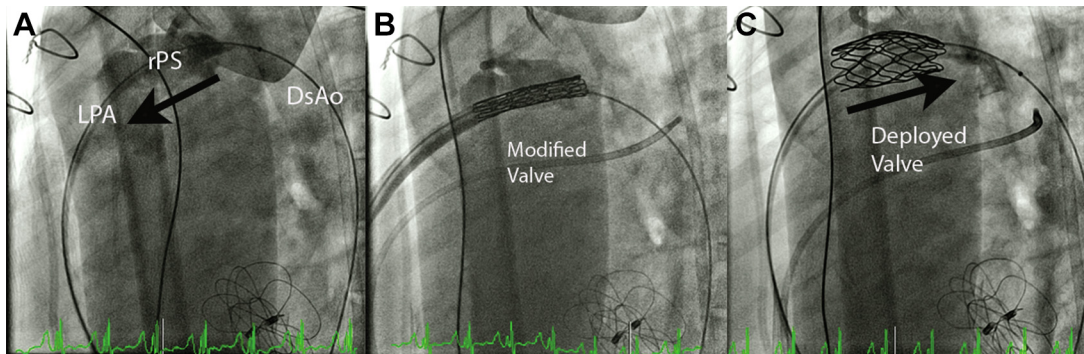
MANAGEMENT

After multidisciplinary discussion, she was taken to the cardiac catheterization laboratory for valve implantation within the rPS. Her right atrial pressure was 11 mm Hg, RV pressure 90/16 mm Hg, and PA pressure 90/45 (64) mm Hg, with a simultaneous aortic pressure of 90/56 (71) mm Hg, wedge pressure of 14 mm, superior vena cava saturation of 50%, PA saturation of 60%, ascending aorta saturation of 84%, and DsAo saturation of 84%. Angiography demonstrated bidirectional shunting and an incompetent deteriorated rPS valve. We modified the 18-mm Ensemble transcatheter valve delivery system to fit a 20-mm Melody (Medtronic) valve by removing the 18-mm balloon and using a 15-mm balloon instead, maintaining the external sheath of the delivery system. Once the valve was crimped onto the balloon, we advanced it with the tip of the balloon slightly inflated over a stiff wire. After valve deployment, angiography demonstrated resolution of left to right shunting (Figure 2, Videos 1 to 3), and her hemodynamics were stable as previous with the

FIGURE 1 Computed Tomography Imaging of the Reversed Potts Shunt Pre and Post Intervention

(A) Computed tomographic (CT) sagittal view of the surgical reversed Potts shunt (rPS). (B) Noncontrast CT sagittal view of the valve stent frame (arrow) within the rPS (arrowhead). DsAo = descending aorta; LPA = left pulmonary artery.

FIGURE 2 Fluoroscopy and Angiography of the Reversed Potts Shunt and Transcatheter Valve Placement



(A) Angiographic lateral view of reversed Potts shunt (rPS) with contrast flowing predominately from left to right (arrow). (B) Positioning of the modified valve into the rPS. (C) Deployed valve with resolution of left-to-right flow.

exception of drops in PA saturation to 49% and DsAo saturation to 80%.

DISCUSSION

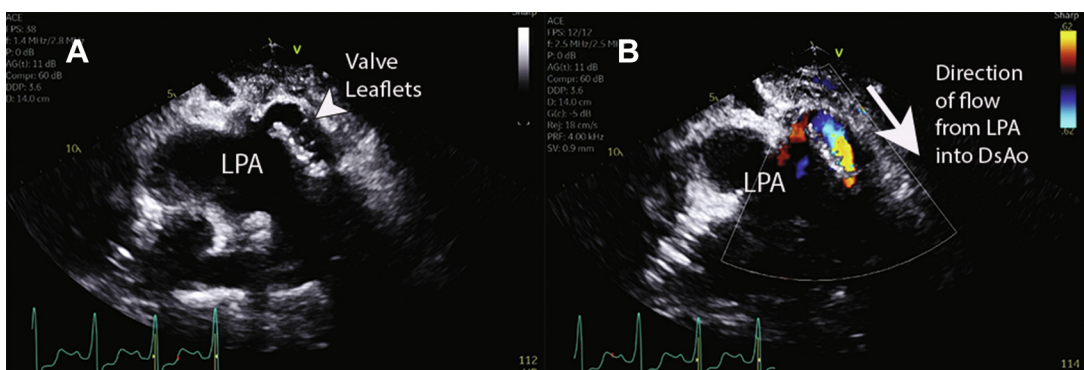
Idiopathic PAH is a progressive disease despite advances in medical therapy, and lung transplantation has a 5-year survival of approximately 59.4%.^{3,4} The rPS is a palliative surgical or interventional option for this group of patients.⁵ It has been proven effective in children and adults with severe symptomatic PAH who have suprasystemic RV pressures. The rPS has been shown to significantly improve functional status and increase transplant-free survival in the majority of recipients.^{3,6,7} The hemodynamics of rPS demonstrate that left-to-right shunting can be detrimental,

leading to pulmonary overcirculation and edema, as seen in the present case, and unidirectional right-to-left flow is preferable.^{8,9} The transcatheter bovine jugular valve, although originally intended to be deployed at 18-22 mm in diameter in the pulmonary position, can be functional at diameters as narrow as 12-16 mm. The valve in this case prevented left-to-right flow across the rPS and thus pulmonary overcirculation, functioning as it was originally intended: a valved unidirectional rPS.

FOLLOW-UP

The patient's respiratory status improved, she was weaned off oxygen supplementation, as well as inotropic and pressor support over a few days, and

FIGURE 3 Echocardiographic View of the Valve Within the Reversed Potts Shunt and Color Doppler Demonstrating Right-to-Left Flow Across the Valve



(A) Echocardiographic view of the valve within the reversed Potts shunt, the arrowhead indicating the leaflets. (B) Color Doppler demonstrating right-to-left flow across the valve. Abbreviations as in Figure 1.

her pulmonary vasodilator, epoprostenol, was adequately up-titrated. Echocardiography demonstrated a functioning valve within the rPS without regurgitation nor stenosis (Figure 3, Videos 4 and 5). The patient is currently active on the lung transplant list.

CONCLUSIONS

This novel use of a modified transcatheter bovine jugular valve in a rPS in patients with severe PAH

assists in maintaining unidirectional right-to-left shunting to offload the RV.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS idiopathic pulmonary arterial hypertension, reversed Potts shunt, transcatheter valve implantation

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