

# Unidirectional-valved Pulmonary to Systemic Shunt for Idiopathic Pulmonary Hypertension

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## Background

- ❖ Idiopathic pulmonary arterial hypertension (IPAH) is a progressive disease that leads to right ventricular strain and eventual failure
- ❖ Patients with Eisenmenger physiology have been shown to fare better than those with IPAH presumably due to preservation of RV function via afterload reduction into systemic circulation. In order to mimic this physiology, the application of a pulmonary to systemic shunt has been proposed in patients with end-stage IPAH.
- ❖ A Potts shunt that allows bidirectional shunting or a unidirectional-valved shunt (UVS) which allows for only right-to-left shunting at the level of the great vessels have been proposed as potential approaches.

## Patient 1 Presentation

- ❖ 5-year-old male with IPAH, supra-systemic pulmonary artery pressures, PVR of 24 indexed Wood units and moderate to severe right ventricular dysfunction.
- ❖ Despite percutaneous atrial septostomy, oral Bosentan, and injectable Treprostinil via subcutaneous pump, he had worsening symptoms and an acute decompensation secondary to respiratory rhinoviral illness and was therefore offered a pulmonary-to-systemic shunt

## Patient 2 Presentation

- ❖ 5-year-old female with IPAH, labile pulmonary artery pressures, PVR of 18 indexed Wood units and low cardiac output with severe right ventricular dysfunction.
- ❖ Despite percutaneous atrial septostomy, oral sildenafil, and continuous Treprostinil via central line, she continued to require inhaled nitric oxide given intermittent pulmonary hypertensive crises and was therefore offered a pulmonary-to-systemic shunt

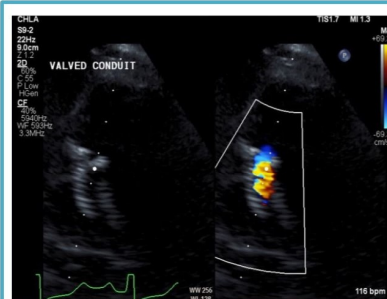
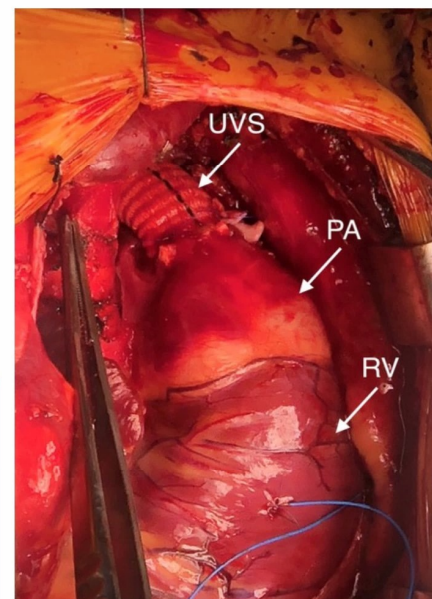


Figure 1. Postoperative TEE demonstrating right-to-left flow through the UVS

Figure 2. Postoperative TEE demonstrating right-to-left shunting through the valved ASD

Figure 3. UVS fashioned from a 9mm cryopreserved pulmonary homograft sewed inside a 10mm Hemashield graft

Figure 4. In-situ view of the UVS



## Surgical Management and Technique

- ❖ A valved pulmonary-to-systemic shunt from main pulmonary artery to descending aorta placed just distal to left subclavian artery takeoff
- ❖ A valved ASD patch was fashioned using autologous pericardium with an 8mm central fenestration and overlying flap on its left atrial side suture for two-thirds of its circumference
- ❖ The UVS would allow for right ventricular unloading at supra-systemic pulmonary pressures while preventing pulmonary over-circulation when systemic pressures are higher than pulmonary pressures

## Patient 1 Follow-up

- ❖ Post-operative TEE demonstrated intermittent right-to-left shunting at the atrial level through the valved ASD and right-to-left flow seen in the UVS
- ❖ Recovered well after surgery and was discharged home on post-operative day 5
- ❖ Current regimen is IV Treprostinil, PO Bosentan, PO Sildenafil and nighttime oxygen
- ❖ Bidirectional shunting across UVS was seen at one-year follow up

## Patient 2 Follow-up

- ❖ Post-operative TEE demonstrated no interatrial shunting and no right-to-left shunting through UVS
- ❖ Recovered well after surgery and was discharged home on post-operative-day 31. Majority of this prolonged time was pursuing insurance approval and supplies
- ❖ Current regimen is IV Treprostinil, PO Bosentan, PO Sildenafil, and around the clock oxygen
- ❖ Bidirectional shunting across the UVS was seen at six-month follow up

## Conclusion

- ❖ Valved unidirectional pulmonary-to-systemic shunt is a feasible approach in patients with supra-systemic IPAH or recurrent pulmonary hypertension crises not being adequately decompressed after percutaneous atrial septostomy while minimizing the effect of left-to-right shunt when pulmonary pressures become sub-systemic. To our knowledge, these are the youngest patients undergoing this procedure in the literature. With serial follow-up, both patients developed bidirectional shunting across the UVS but clinically have continued to do well. Continued follow-up will be needed to determine durability of this intervention.

1. Baruteau AE, et al. M. Potts shunt in children with idiopathic pulmonary arterial hypertension: long-term results. Ann Thorac Surg. 2012 Sep;94:817-24.
2. Rosenzweig EB, et al. A novel unidirectional-valved shunt approach for end-stage pulmonary arterial hypertension: Early experience in adolescents and adults. J Thorac Cardiovasc Surg. 2021 Apr;161:1438-1446.e2.

There are no disclosures to report.