

A novel unidirectional-valved shunt approach for end-stage pulmonary arterial hypertension: Early experience in adolescents and adults



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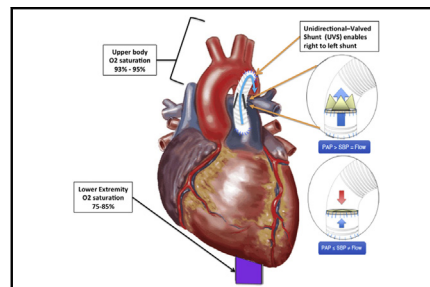
ABSTRACT

Objectives: Despite advances in treatment of idiopathic pulmonary arterial hypertension (IPAH), there remains no medical cure, and patients can experience disease progression leading to right heart failure, progressive exercise intolerance, and death. The reversed Potts shunt (left pulmonary artery to descending aorta) was reintroduced for treatment of end-stage IPAH to permit decompression of the suprasystemic right ventricle by right to left shunting, with preservation of upper body oxygenation. The shunt has the potential to delay the need for lung transplantation and offer a treatment for those who are transplant ineligible. To optimize shunt design and avoid the potential complications of bidirectional shunting, we developed a novel approach using a unidirectional-valved shunt (UVS) in patients with IPAH with suprasystemic pulmonary arterial pressure and poor right ventricular function.

Methods: A single-center retrospective review was performed of UVS cases done at Columbia University Medical Center–New York Presbyterian between November 1, 2016, and May 1, 2019.

Results: Five patients (4 female; ages 12-22 years) underwent UVS. All had suprasystemic pulmonary arterial pressure, poor right ventricular function, and World Health Organization functional class IV symptoms at baseline. All patients are alive and transplant-free at latest follow-up (range 3-33 months; median 6 ± 11 months).

Conclusions: The UVS may offer an alternative solution to lung transplantation in adolescents and young adults with IPAH. Longer-term follow-up is needed to determine the ultimate impact of unidirectional unloading of the right ventricle in these patients and to determine whether the UVS will enable a broader approach to the treatment of patients with IPAH. (*J Thorac Cardiovasc Surg* 2021;161:1438-46)



UVS approach diverts cyanotic blood to the lower body and maintains upper body oxygenation.

CENTRAL MESSAGE

Early experience with novel unidirectional-valved shunt approach for end-stage idiopathic pulmonary arterial hypertension—an alternative to the reversed Potts shunt—is discussed.

PERSPECTIVE

A novel unidirectional-valved shunt approach was applied to alleviate right heart strain, ensure adequate cardiac output, and maintain upper body oxygenation in patients with end-stage idiopathic pulmonary arterial hypertension. This approach, unlike the reversed Potts, shunt avoids bidirectional shunting in these fragile patients and may provide another alternative to lung transplantation.

See Commentaries on pages 1447, 1449, and 1451.

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Abbreviations and Acronyms

ECMO	= extracorporeal membrane oxygenation
FC	= functional class
IPAH	= idiopathic pulmonary arterial hypertension
PAH	= pulmonary arterial hypertension
PAP	= pulmonary arterial pressure
PH	= pulmonary hypertension
RV	= right ventricle
UVS	= unidirectional-valved shunt
WHO	= World Health Organization
WSPH	= World Symposium on Pulmonary Hypertension



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The treatment of end-stage idiopathic pulmonary arterial hypertension (IPAH) remains challenging due to the lack of a medical cure. Despite advances in medical therapy for group 1 pulmonary arterial hypertension (PAH), some patients experience disease progression that leads to right heart failure, progressive exercise intolerance, and increased risk for death.¹ For these patients, interventional and surgical approaches may be lifesaving. Transcatheter atrial septostomy is an effective palliative measure, but it is limited because an atrial septostomy sized sufficiently could lead to profound systemic cyanosis.^{2,3} Lung transplantation is severely limited by its durability, given the 50% to 70% 5-year survival.⁴⁻⁶ Furthermore, the need for retransplantation and the long-term complications of lung transplantation are major concerns, especially for younger patients. Clearly, alternative strategies are needed for patients with IPAH to prolong survival and defer time to transplantation.

There have been recent reports of using the Potts shunt to palliate children with end-stage IPAH to decompress the suprasystemic right ventricle by right to left shunting.⁷⁻¹⁰ Originally developed as a shunt from the descending aorta to the left pulmonary artery for infants with cyanotic heart disease, a “reversed” Potts shunt was created to shunt suprasystemic cyanotic blood from the pulmonary to the systemic circulation, which decompresses the right ventricle (RV) while sparing the coronary and cerebral circulations of deoxygenated blood.⁷ Reverse Potts

physiology mimics the Eisenmenger physiology of a patent ductus arteriosus, which has a better natural history than IPAH due to preservation of RV function by ventricular unloading.^{11,12} Surgical and interventional approaches to create a reversed Potts shunt have been used with mixed results, with only a few institutions embracing this approach and having the requisite expertise to successfully manage the reverse Potts shunts.⁷⁻¹⁰ The largest series consisted of 24 pediatric patients with drug-refractory PAH in whom a Potts shunt was created (19 surgical and 5 via stenting of a persistent ductus arteriosus).⁸ Six patients (25%) experienced severe postoperative complications, and there were 3 early deaths (12.5%) related to low cardiac output. After a median follow-up of 2.1 years, the 21 survivors showed persistent improvement in functional capacities.^{8,9} Another study used a transcatheter approach to insert a stent between the aorta and pulmonary artery.¹³ Four of the 6 patients (67%) survived the procedure. These experiences have ignited an interest in novel approaches to slow the inevitable progression of disease. However, patient selection and indications remain poorly defined, as do a lack of reproducible technical approaches, timing, and management as a palliative tool for IPAH. Whether the classic reversed Potts shunt is the optimal design also remains in question, especially for older children and adults with systemic PAH and RV dysfunction.

These questions led us to design a novel unidirectional shunt as a pressure release valve to permit flow from the main pulmonary artery to the descending aorta, which opens only during periods of suprasystemic pulmonary arterial pressure (PAP).¹⁴ Unlike the classic reversed Potts shunt, the unidirectional-valved shunt (UVS) provides the physiologic benefit of unloading the RV while preventing “reversed” flow from the aorta to the pulmonary circulation when systemic blood pressure exceeds PAP and in early systole due to prolonged RV contraction and electromechanical dyssynchronization of the ventricles in patients with severe pulmonary hypertension (PH).¹⁴ (Figure 1 and Figure E1). This concept has been shown in an animal model¹⁵ and in one adult as a unidirectional flap valve.¹⁶ Previous series have cautioned against using the reverse Potts shunt in children with severely diminished RV function. However, given the limited treatment options, our team explored the use of the Potts shunt in older children and young adults, including those with significantly diminished RV function in the setting of systemic or suprasystemic PAP. The concern for the potential adverse impact of bidirectional shunting if the PAP intermittently becomes subsystemic informed our novel design in which we incorporated a valve in a shunt to prevent backflow into the pulmonary circulation. This unidirectional-valve design captures the physiologic benefit of RV-PA unloading while

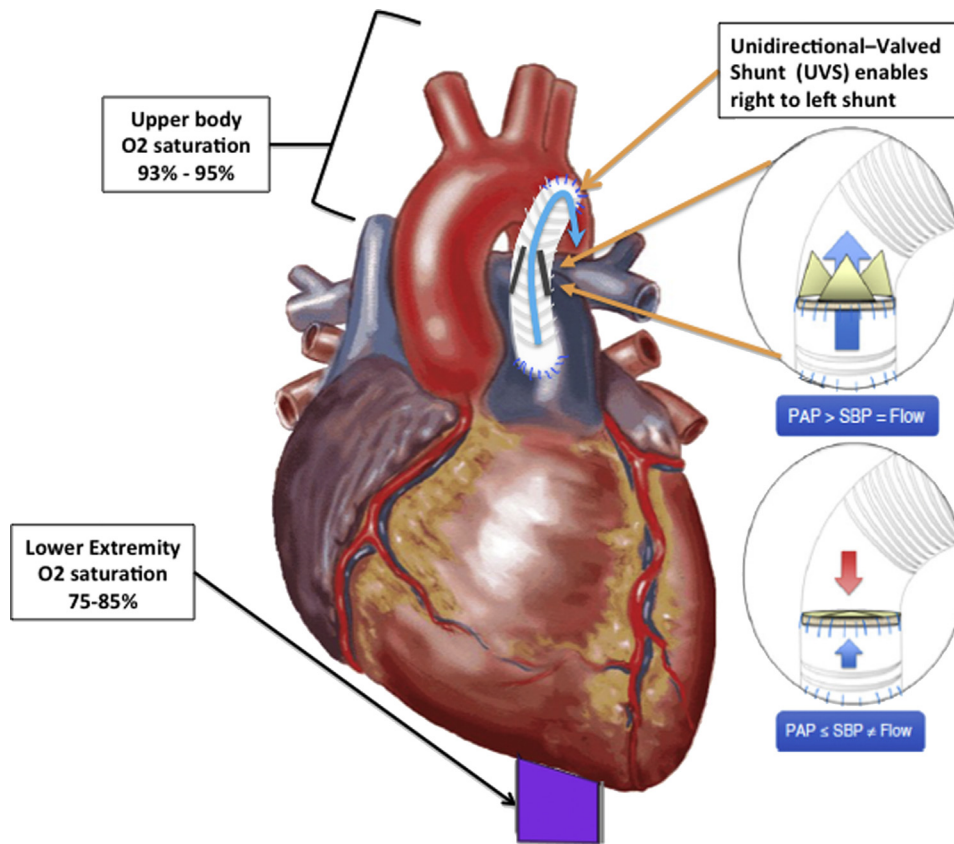


FIGURE 1. UVS design shows the site of surgical insertion of the valved conduit from the superior aspect of the main pulmonary artery to the descending aorta distal to left subclavian artery. O₂, Oxygen; UVS, unidirectional-valved shunt; PAP, systolic pulmonary artery pressure; SBP, systolic systemic arterial blood pressure.

preserving upper body oxygenation and preventing the negative impact of reversed left to right shunting on a vulnerable pulmonary arterial system. We report our first 5 UVS surgical cases to illustrate the unique physiology of the UVS shunt, discuss selection criteria, timing and applications, and review our approach and perioperative management.

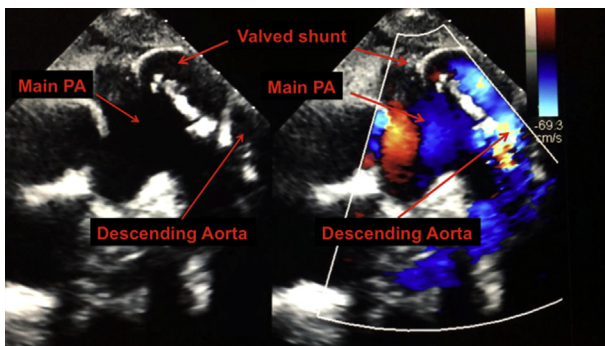


FIGURE 2. Transthoracic 2-dimensional echocardiogram of unidirectional-valved shunt (parasternal short axis view) showing the main pulmonary artery (*main PA*), unidirectional valved shunt, and descending aorta without (*left*) and with color Doppler (*right*); for an animated view, please see *Videos 1-3*).

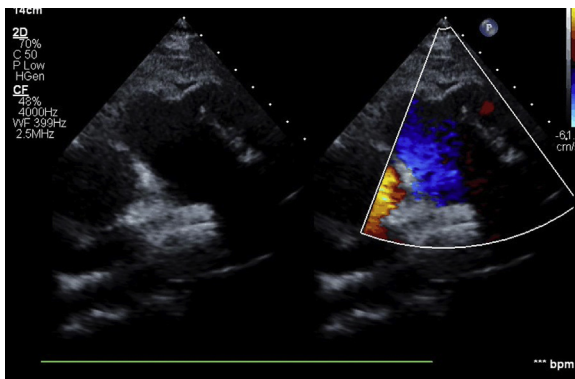
METHODS

A single-center retrospective review was performed of all IPAH cases of a novel UVS shunt done at Columbia University Medical Center–New York Presbyterian Hospital between November 1, 2016, and May 1, 2019, and followed up through August 1, 2019. As we originally described it, the UVS is a unidirectional (right to left) valved shunt created by sewing a 12-mm Contegra valved Conduit (Medtronic, Minneapolis, Minn) into a Fusion Bioline graft (Medtronic, Eatontown, NJ) (*Figure 1*¹⁴; *Figure 2* and *Videos 1-3* show the functional valve on transthoracic echocardiogram). In the immediate postoperative period the patients have arterial catheters in both the right radial artery and a femoral artery to monitor right arm and lower-extremity partial pressure of oxygen and arterial oxygen saturations. The patients also have oxygen saturation probes on the right upper extremity and a lower extremity to monitor for desaturation to the upper extremity, which reflects cerebral and coronary saturation. We also monitor cerebral and renal near-infrared spectroscopy. The use of exogenous blood products is minimized to reduce the risk of triggering PH crises and impact on future transplant eligibility.

Baseline preoperative, intraoperative, postoperative characteristics, and outcomes of these patients were included for review. This study was approved by the institutional review board at Columbia University (IRB#AAAK2059).

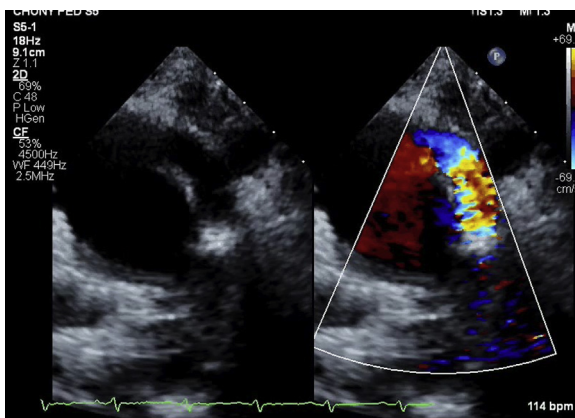
RESULTS

UVS surgery was performed in 5 patients with World Health Organization (WHO) functional class (FC) IV

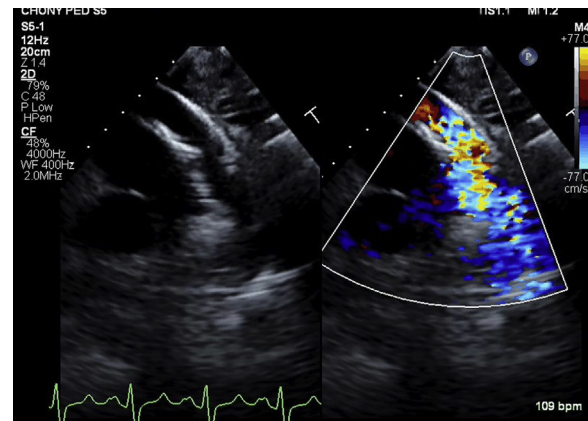


VIDEO 1. This transthoracic echocardiogram (parasternal short-axis view) illustrates the UVS shunt taking off from the main pulmonary artery. In the left panel, the valve can be seen and in the right panel the direction of flow can be seen with color Doppler imaging showing the integrity of the valved shunt. Video available at: [https://www.jtcvs.org/article/S0022-5223\(19\)32771-0/fulltext](https://www.jtcvs.org/article/S0022-5223(19)32771-0/fulltext).

symptoms (4 female; ages 12-22 years) and World Symposium on Pulmonary Hypertension (WSPH) group 1 PAH who were failing maximal medical therapy to provide durable long-term palliation and potential deferral of lung transplantation. One patient was not a transplant candidate, and 3 patients were listed for lung transplantation before UVS surgery. Baseline patient characteristics are shown in [Table 1](#) and [Table E1](#). All patients were receiving triple therapy for PH with a phosphodiesterase 5-inhibitor, endothelin receptor antagonist, and continuous intravenous or subcutaneously delivered prostanoid before UVS surgery. Four patients had IPAH, and 1 had IPAH following timely repair of transposition of the great vessels on day of life 3. Two



VIDEO 2. This transthoracic echocardiogram also demonstrates the UVS shunt on parasternal short-axis imaging without (*left*) and with color Doppler (*right*) in a second patient who underwent UVS surgery. Video available at: [https://www.jtcvs.org/article/S0022-5223\(19\)32771-0/fulltext](https://www.jtcvs.org/article/S0022-5223(19)32771-0/fulltext).



VIDEO 3. This transthoracic echocardiogram demonstrates the UVS shunt on parasternal short-axis imaging without (*left*) and with color doppler (*right*) in a third patient who underwent UVS surgery. Video available at: [https://www.jtcvs.org/article/S0022-5223\(19\)32771-0/fulltext](https://www.jtcvs.org/article/S0022-5223(19)32771-0/fulltext).

patients had IPAH associated with an atrial septal defect, and 2 had previously undergone atrial septostomy. The patients had suprasystemic PAP (PA mean 92 ± 16 mm Hg; pulmonary vascular resistance index 32 ± 9 U \cdot m 2 ; and pulmonary vascular resistance/systemic vascular resistance 0.7 [0.5-0.7]), severely depressed RV function by echocardiographic imaging, and elevated preoperative N-terminal pro b-type natriuretic peptide levels (range 1019-8064 pg/mL; n = 4). ([Table 2](#)) One patient was on inotropic support before surgery. None of the patients had a significant resting oxygen requirement preoperatively other than the elective use of nighttime oxygen in 2 patients. All had WHO FC IV symptoms and severe dyspnea with reduced exercise capacity, 2 with exertional chest pain and 1 with presyncope. Preoperative hemodynamics demonstrated severe PAH ([Table 2](#)).

Operative, postoperative characteristics, and outcomes are shown in [Table 3](#). All patients underwent UVS shunt surgery via a median sternotomy on cardiopulmonary bypass, and bicaval cannulation was used for operative procedures that included atrial septal defect interventions. Cardiopulmonary bypass time ranged from 1 hour 47 minutes to 3 hours 13 minutes. The shunt was created out of a 12-mm valved Contegra conduit placed in a widened 10-mm Fusion Bioline graft to prevent dilation of the valve annulus. The proximal graft is beveled and sewn slightly off center from the anterior main pulmonary artery in a direction that allows for a natural travel to the aortic anastomosis. The distal anastomosis is established beyond the takeoff of the left carotid artery, at or beyond the level of the left subclavian artery depending upon anatomic considerations. Given this was our early experience, we took a cautious approach by leaving a small atrial communication (3-6 mm) to ensure adequate decompression of the right atrium

TABLE 1. Baseline patient demographic, clinical, and echocardiographic characteristics (n = 5)

Characteristics	
Age, y	15 (13-16)
Time from diagnosis to Potts shunt, y	12 (±3)
Sex	
Male	1 (20%)
Female	4 (80%)
Weight, kg	46 (44-53)
PH/cardiac diagnosis	
IPAH with atrial communication	4 (80%)
D-TGA with IPAH	1 (20%)
Preoperative atrial communication	
None	1 (20%)
PFO/septostomy	2 (40%)
Secundum ASD	2 (40%)
Pulmonary hypertension therapies before Potts shunt	
Sildenafil + endothelin antagonist + treprostinil	3 (60%)
Sildenafil + endothelin antagonist + epoprostenol	2 (40%)
Home oxygen use	2 (40%)
Symptoms	
Exertional dyspnea	4 (80%)
Exertional dyspnea + chest pain	1 (20%)
Echo RVSP, mm Hg	121 (±12)
Severe RV dysfunction	5 (100%)
Systolic septal flattening/bowing	5 (100%)
LVEF (%)	61 (±7)

Numbers represent means/medians (standard deviation/interquartile range) for continuous variables and numbers (%) for categorical variables. *PH*, Pulmonary hypertension; *IPAH*, idiopathic pulmonary arterial hypertension; *D-TGA*, dextro-transposition of the great arteries; *PFO*, patent foramen ovale; *ASD*, atrial septal defect; *RVSP*, right ventricular systolic pressure; *RV*, right ventricle; *LVEF*, left ventricle ejection fraction.

in the early postoperative period and to ensure there was a safety shunt in the event of UVS failure. The atrial communication can be closed percutaneously if the need arises. Four patients were placed on peripheral venovenous extracorporeal membrane oxygenation (ECMO) (3 intraoperatively and 1 postoperatively). We cannulate femoral vein to right internal jugular vein. We considered using a single dual lumen cannula in the right internal jugular vein but were concerned that positioning may be an issue for these patients who were quite fragile. ECMO eased the transition to the new physiology by providing an oxygenated right to left shunt through the atrial communication and the UVS shunt, enabling early extubation and mobilization (Table 3). All 4 patients on venovenous ECMO were extubated while on ECMO and ambulatory. All ECMO cannulas were inserted percutaneously and did not require a second general anesthesia for removal.

TABLE 2. Pre-UVS shunt hemodynamics and laboratory values (n = 5)

Values	
Time from catheterization to Potts shunt, d	186 (54-229)
RA mean, mm Hg	9 (6-9)
Systolic arterial blood pressure, mm Hg	103 (±7)
Systolic PA pressure, mm Hg	134 (±29)
Diastolic PA pressure, mm Hg	65 (±11)
PA mean, mm Hg	92 (±16)
PVRI, U*m ²	32 (±9)
PVR/SVR	0.7 (0.5-0.7)
PCWP, mm Hg	13 (11-14)
Cardiac index, L/min/m ²	2.6 (±0.8)
Mixed venous saturation, %	61 (±14)
Systemic arterial saturation, %	90 (±3)
NT-proBNP, pg/mL*	4013 (±2941)
Troponin, ng/mL	0.04 (±0.05)
Creatinine, mg/dL	0.6 (±0.1)

Numbers represent means/medians (SD/IQR) for continuous variables. *RA*, Right atrial; *PA*, pulmonary artery; *PVRI*, pulmonary vascular resistance indexed; *PVR*, pulmonary vascular resistance; *SVR*, systemic vascular resistance; *PCWP*, pulmonary capillary wedge pressure; *NT-proBNP*, N-terminal pro b-type natriuretic peptide. *N = 4.

All patients continued their PH medications with dose reduction in prostanoids in 3 of 5 patients. All were placed on aspirin and warfarin postoperatively with a target INR of 2.5-3.5.

All 5 patients survived to hospital discharge (Table 3) and are alive at latest follow-up on August 1, 2019 (range 3-33 months; median 6 ± 11 months). Four of the 5 patients were discharged home on supplemental oxygen and all 4 were subsequently weaned off. There were no major early complications. No patient has had further presyncope or exertional chest pain since surgery. WHO FC has improved from WHO FC IV to WHO FC II-III in all patients. There have been improvements in N-terminal pro b-type natriuretic peptide levels (mean baseline vs postoperative 4013 vs 1107; n = 4) and estimated RV systolic pressure by transthoracic echocardiogram (baseline vs postoperative 121 ± 12 to 102 ± 8 mm Hg, P = .04; n = 5). The fractional area change of the right ventricle on TTE improved from 22% (preoperatively) to 35% at median follow-up of 5.8 months (n = 5). Four of the 5 patients have been compliant with anticoagulation; one who was noncompliant with warfarin had thrombosis of her shunt, which will be addressed percutaneously. All patients are alive and transplant-free at latest follow-up (range 3-33 months; median 6 ± 11 months). All 3 who were listed have been taken off the active lung transplant list.

TABLE 3. UVS operative, in-hospital characteristics, and outcomes

UVS shunt size, mm	12 (10-12)
ASD intervention	
None	2 (40%)
ASD closure	1 (20%)
ASD downsized	1 (20%)
ASD creation	1 (20%)
CPB time, min	149 (±29)
Postoperative venovenous ECMO	4 (80%)
ECMO in OR	3 (75%)
ECMO in ICU	1 (25%)
ECMO duration, d	5 (±5)
Intubation duration, d	3 (±2)
Extubated while on ECMO	4 (100%)
Resting preshunt SpO ₂ , % (right arm)	92 (±5)
Resting postshunt SpO ₂ , % (leg)	84 (±9)
Discharge PH therapy	
Sildenafil + endothelin antagonist + treprostinil	2 (40%)
Sildenafil + endothelin antagonist + epoprostenol	3 (60%)
Prostanoid dose decreased post operatively	3 (60%)
Length of stay, d	29 (±6)
Survival to hospital discharge	5 (100%)
Transplant-free survival	5 (100%)

Numbers represent means/medians (standard deviation/interquartile range) for continuous variables and numbers (%) for categorical variables. *UVS*, Unidirectional-valved shunt; *ASD*, atrial septal defect; *CPB*, cardiopulmonary bypass; *ECMO*, extracorporeal membrane oxygenation; *OR*, operating room; *ICU*, intensive care unit; *PH*, pulmonary hypertension, *SpO₂*, systemic arterial oxygen saturation.

DISCUSSION

We describe the use of a novel unidirectional-valved shunt for end-stage WSPH group 1 PAH. Similar to the

indications for atrial septostomy and reversed Potts shunt, this intervention should be reserved for adolescents and young adults with advanced WSPH group 1 PAH and clinical worsening despite maximal medical therapy as a bridge to lung transplantation.¹⁷ The rationale for waiting for medical failure relates to the invasive nature and risks of the intervention. However, some authors have suggested an intervention may be too late if the RV fails, precluding the use of shunt procedures.⁹ Despite apprehension about the failing RV, we successfully applied the UVS to patients with poor RV function by using venovenous ECMO as a bridge to recovery. Thus, adolescents and young adults with IPAH who are on maximal therapy, including parenteral prostanoids, have relatively preserved gas exchange without substantial oxygen requirement, and who show signs of clinical worsening should be considered for a shunt.¹⁷ Other critical factors to consider include lung transplant eligibility, comorbidities precluding transplantation, and team expertise in performing and managing shunt physiology (Table E2).

Shunt Design Selection (Reversed Potts, UVS, Versus Transcatheter)

There are several considerations in shunt design selection of the reversed Potts versus UVS shunt. The use of a classic reversed Potts shunt with direct anastomosis of the left PA to descending aorta has certain advantages and disadvantages. Although the approach through a left thoracotomy is appealing in its simplicity, there are marked challenges in the older patient. Not infrequently there are extensive aortopulmonary collateral vessels to dissect and control during the exposure. Tissue pliability is diminished, compromising a primary anastomosis of the left PA and aorta. Sizing a graft presents difficulties to ensure an appropriate lay of the graft, especially in dynamic ventilation, which could compromise the integrity of the anastomosis

TABLE 4. Early observed postoperative physiologic patterns in the postoperative UVS shunt and management strategies

Physiologic state	Clinical pattern	Clinical data example	Intervention
Rest	Mild desaturation with small or insignificant gradient between the right arm and leg O ₂ saturation differential U/L (<5%)	HR 100 bpm SBP 100/65 PAP 100/65 RA sat. 96% LE sat. 92%	None
Exertion/exercise	Differential cyanosis increases Right arm saturation >> lower extremity saturation (target U/L: 5%-20% differential saturations)	HR 124 bpm SBP 109/74 PAP 115/75 RA sat. 94% LE sat. 82%	Normal response; confirms shunt patency
Pulmonary hypertensive crisis with systemic arterial hypotension	Triggered by hypoxia, or hypovolemia and net systemic vasodilation Large differential O ₂ saturation followed by pre- and postshunt desaturation	HR 120 SBP 100/65 PAP 120/80 RA sat. 85% LE sat. 75%	Raise SBP; Increase O ₂ supplementation temporarily if needed
Systemic arterial hypertensive crisis	Triggered by agitation and or pain; narrowing of shunt differential from increase in SBP	HR 140 SBP 130/75 PAP 110/70 RA sat. 96% LE sat. 94%	Treat with anxiolytic/pain therapy as needed

O₂, Oxygen; U/L, upper/lower extremity; HR, heart rate; SBP, systolic blood pressure; PAP, pulmonary arterial pressure; RA, right arm; sat., saturation; LE, lower extremity.

and the stability of flow through it. Bidirectional flow may have deleterious effects on pulmonary blood flow and pressure, increasing the risk of worsening pulmonary function, edema, and hemorrhage. Assuming the patient eventually undergoes lung transplant, the surgeon would be in the unenviable position of dissecting through a reoperative field where there is a direct anastomosis between the left PA and aorta in a blind area where the risk of evulsion could be devastating. Another challenge is the difficulty of controlling the amount of bidirectional shunting in dynamic pulmonary and systemic circulations and the impact on the fragile pulmonary circulation when the systemic pressure rises above the PAP, see Figure 2.

The UVS shunt, likewise, has advantages and disadvantages. The primary disadvantage to a previous midline sternotomy is the substernal dissection at the time of transplant. There is also the concern of being able to ligate the shunt, but the standard exposure used in transplantation provides adequate exposure to the shunt given the aortic anastomosis is near the take-off of the left subclavian artery. The UVS's distinct advantage is the ability to selectively shunt during suprasystemic PAPs including during exertion and PH crises while preventing systemic to pulmonary shunting at times when not required (Figures 1 and 2). In our experience, the suprasystemic PAP often decreases to systemic levels after implantation of a UVS shunt with commensurate reduction in PA diastolic pressure over time creating a potential for intermittent left to right shunting even in patients with severe PAH. In 1 patient with pre- and postoperative hemodynamics, the PA diastolic pressure decreased to subsystemic levels from 65 to 48 mm Hg over 6 months. The essential point is that systemic pressures will have transient points of greater pressures than the PAPs during the cardiac cycle, and the amount and frequency of these episodes will likely change over the course of the patient's life. The UVS simply acts as a "back stop" to prevent the deleterious effects of left-to-right shunting on a PA-RV system that grows less tolerant with age and potentially broadens the application into adulthood.

A transcatheter approach is limited to a few centers; the optimal approach and application is typically for young patients with an existing patent ductus arteriosus that is stentable.^{13,18,19}

RV Function

Preserved RV function is considered a prerequisite for the reversed Potts shunt because of concerns that a compromised RV is unlikely to improve by reducing the systolic PAP from suprasystemic to systemic pressure, an opinion supported by a simulation study suggesting that RV afterload changes little after Potts shunt,²⁰ hence raising concerns that the compromised RV will suffer further from

exposure to systemic pressures from the aortic runoff with a bidirectional shunt. This will result in pressure and volume shifts into the pulmonary circulation during periods when the systemic pressure is greater than PAP, resulting in a greater resting mean PAP. The UVS design reduces this risk by eliminating bidirectional flow, providing a "pop-off" to prevent syncope and potentially sudden death while preserving well-oxygenated coronary and cerebral perfusion without run off into the pulmonary circulation during diastole. We observed improved RV systolic function over time associated with the reduction in systolic PAP to systemic levels, despite poor preoperative right ventricular function. However, any benefit from invasive surgery should be weighed with the risk of poor baseline RV function when palliating a patient to defer lung transplantation.

Intraoperative Considerations

Intraoperative considerations include having an experienced cardiac anesthesia team for safe induction of patients with PAH and quick access to ECMO or bypass in the event of a PH crisis during induction. For patients with a patent foramen ovale, atrial septostomy, or even an atrial septal defect, there should be a preoperative discussion about atrial shunt creation, downsize, or closure. For large atrial septal defects, closure or downsizing (3-6 mm) should be performed at the time of surgery. Although there is the theoretical risk of worsening global cyanosis in the presence of an atrial communication, the UVS or classic reversed Potts shunt should decompress the RV and right atrium, which will ultimately decrease right to left shunting at the atrial level. This may ease the transition of circulation in the immediate postoperative period. The surgical team should be prepared to place the patient on ECMO intraoperatively or postoperatively if needed for hemodynamic instability or cyanosis. In the presence of a patent UVS shunt or classic reverse Potts shunt with or without an atrial communication, venovenous ECMO will maintain adequate saturations in the upper and lower body, while medication is adjusted and the patient acclimates to the new shunt physiology.

We chose this approach to ensure adequate oxygenation to facilitate extubation while on ECMO, which enabled us to monitor the physiology and shunting while awake and active an impossible task in sedated and intubated patients. Early extubation also reduces the risk of ventilator-associated complications, which are poorly tolerated in patients with PAH. Venovenous ECMO was slowly weaned until the patient was off vasopressors and oxygen supplementation was not at maximal settings with >90% saturations in the right arm. For hemodynamic instability,

venoarterial ECMO could be used as a bridge to recovery; however, it should be used cautiously, as the increase in afterload could work counter to the intended purpose of the shunt, hence our preference for venovenous ECMO.

Postoperative Management

A multidisciplinary team with expertise in pulmonary hypertension, congenital heart disease, cardiothoracic surgery, and ECMO should manage patients undergoing UVS or reversed Potts shunt. Intensive care unit management should include continuous simultaneous monitoring of right arm and pedal saturation/partial pressure of oxygen to determine shunting patterns. A pulmonary arterial catheter is used for continuous monitoring and guidance of inotropic and vasopressor support in the early postoperative period. For patients on intravenous prostanoid therapy, we reduce the dose by 10% at the time of UVS shunt placement, with additional down-titration as needed because the intravenous prostanoid is delivered more directly into the systemic circulation, potentially leading to severe systemic hypotension. Patients will slowly wean from oxygen and the expectation is that they will have differential cyanosis with preserved right arm saturation and lower postshunt saturations (leg); the differential saturations will increase with exertion. We observed 4 categories of physiologic responses in the early postoperative period with shunting driven by pulmonary vascular resistance and systemic vascular resistance (Table 4). It is essential to understand these different patterns and respond accordingly. Anticoagulation is initiated to prevent shunt thrombosis. Patients should be anticoagulated with heparin and then transitioned to warfarin and low-dose aspirin when stable. Patients should receive subacute bacterial endocarditis prophylaxis.

There are several limitations to this retrospective report. There is limited long-term follow-up of this approach, and the integrity of the Contegra conduit will need to be monitored over time. Potential additional drawbacks of using the Contegra valve conduit include the risk of endocarditis and the risk of thrombosis. These patients require anticoagulation, so if anticoagulation is contraindicated, the UVS would not be the preferred approach. In the event of conduit stenosis or valve thrombosis or incompetence, we envision catheter-based interventions. Finally, if the patient undergoes lung transplantation in the future, the conduit will need to be ligated.

In summary, the UVS and classic reversed Potts shunts are evolving into durable palliative approaches for the management of medication-refractory IPAH. Patient selection is critical and considered within the context of both clinical and psychosocial concerns such as lung transplant candidacy (Table E2). A unidirectional-valved shunt is a viable option for an adolescent or young adult who is not transplant eligible to provide durable

palliative support, or for a patient who wishes to prolong the time until lung transplantation. Whether the UVS approach is preferred over the classic reverse Potts shunt remains to be seen. Venovenous ECMO as a transition bridge in this approach can be very useful. If the patient is transplant eligible, we recommend having the patient evaluated and listed before undergoing this procedure in the event the procedure fails. The perioperative management is critical and requires multidisciplinary planning and expertise in PH, congenital heart disease, cardiothoracic surgery, and ECMO with an understanding of the postoperative physiologic patterns. With advances in shunt approaches, there may be a lower threshold for performing these procedures in the future, which convert IPAH physiology to Eisenmenger-like physiology, preserving RV function potentially for decades and deferring lung transplantation.

CONCLUSIONS

Our UVS shunt design and reversed Potts shunt will likely find their place in the armamentarium of treatments for end-stage group 1 PAH in older adolescents and young adults with suprasystemic PAP. Further, the successful use of a unidirectional-valved shunt could broaden the application of use in patients with systemic or near-systemic PAP including adults with IPAH. More work is required to determine the preferred approach and timing of shunt therapy to prevent RV failure, preserve upper body oxygenation, reduce the need for intravenous prostanoid therapy, and defer lung transplantation. Clinical experience and simulation models will inform future treatment algorithms for PAH management.

Conflict of Interest Statement

All authors have nothing to disclose with regard to commercial support.

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Key Words: idiopathic pulmonary arterial hypertension, reverse Potts shunt, pulmonary hypertension, unidirectional-valved shunt, ECMO, atrial septostomy, treatment, differential cyanosis

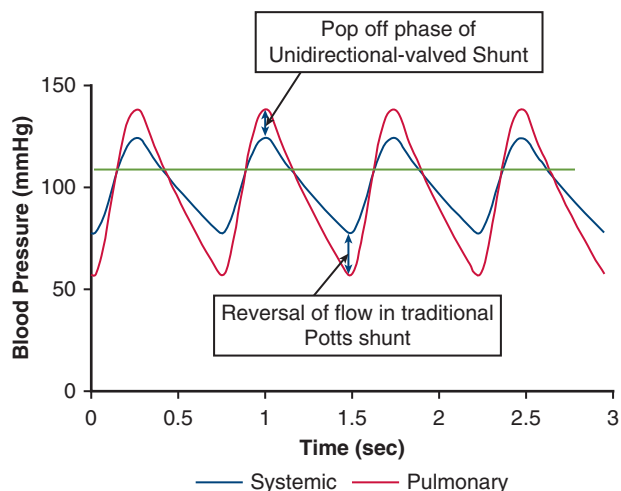


FIGURE E1. Pulmonary and systemic arterial pressures in a patient with idiopathic pulmonary arterial hypertension demonstrating potential for reversal of flow in the traditional reversed Potts shunt. The *green line* represents opening and closing of the shunt valve, the *blue line* represents systemic systolic blood pressure, and the *red line* represents the systolic pulmonary arterial pressure.

TABLE E1. Individual baseline UVS shunt patient characteristics

Patient	Age at surgery, y	Weight, kg	Diagnosis	WHO FC	Oral PH therapy	Prostanoid type and dose	Initial symptoms	Oxygen therapy	BNP, pg/mL
1	22	53	IPAH + BAS	IV	Sildenafil + ambrisentan	Treprostinil 22 ng/kg/min	DOE Presyncope	No	3410
2	15	46	IPAH + ASD	IV	Sildenafil + bosentan	Epoprostenol 98 ng/kg/min	DOE	Yes	3560
3	13	44	IPAH + ASD	IV	Sildenafil + ambrisentan	Treprostinil 242 ng/kg/min	DOE	Yes	N/A
4	12	22	IPAH + BAS	IV	Sildenafil + bosentan	Epoprostenol 104 ng/kg/min	DOE Chest pain	No	1019
5	16	56	D-TGA s/p ASO IPAH	IV	Sildenafil + Macitentan	Treprostinil 212 ng/kg/min	DOE	No	8064

WHO, World Health Organization; FC, functional class; PH, pulmonary hypertension; BNP, b-type natriuretic peptide; IPAH, idiopathic pulmonary arterial hypertension; BAS, balloon atrial septostomy; DOE, dyspnea on exertion; ASD, atrial septal defect; N/A, not available; D-TGA, dextro-transposition of the great arteries; ASO, arterial switch operation.

TABLE E2. Clinical considerations for UVS candidacy

Clinical considerations	Favorable	Less favorable
Age	Adolescent/young adult	Older adult
WHO functional class	III/IV	I/II (risk may outweigh benefit)
RV systolic function	Preserved or diminished but able to generate systemic–suprasystemic PA pressure	Poor RV function unable to generate a systemic PA pressure
LV systolic function	Preserved	LV systolic dysfunction
Anticoagulation	No contraindications	Contraindications
Lung transplant feasibility	Long wait time or contraindication Young age	Short wait/older age
Comorbidities	Absence of end-organ dysfunction	End-organ dysfunction
Parenchymal Lung disease	Absent	Present
Oxygen supplementation	Minimal or due to atrial shunting	Chronic O ₂ dependence
Team expertise	PH/CHD/CTs/ECMO teams	Lack of multidisciplinary team

WHO, World Health Organization; *RV*, right ventricle; *PA*, pulmonary arterial; *LV*, left ventricle; *O₂*, oxygen; *PH*, pulmonary hypertension; *CHD*, congenital heart disease; *CT*, cardiothoracic surgery; *ECMO*, extracorporeal membrane oxygenation.

CONG