Midterm outcomes of the Potts shunt for pediatric pulmonary hypertension, with comparison to lung transplant

Timothy S. Lancaster, MD,[a] Shabana Shahanavaz, MBBS,[b] David T. Balzer, MD,[b] Stuart C. Sweet, MD,[c] R. Mark Grady, MD,[b] and Pirooz Eghtesady, MD, PhD[a]

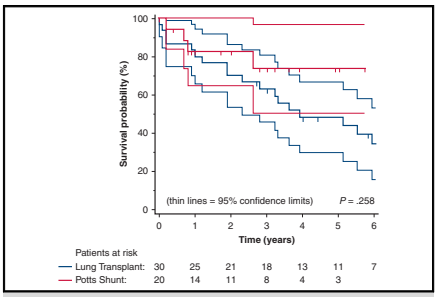
ABSTRACT

Objective: For children with severe pulmonary hypertension, addition of Potts shunt to a comprehensive palliation strategy might improve the outcomes afforded by medications and delay lung transplantation.

Methods: A prospective analysis was conducted of all children undergoing Potts shunt (first performed in 2013) or bilateral lung transplant for pulmonary hypertension from 1995 to present.

Results: A total of 23 children underwent Potts shunt (20 surgical, 3 transcatheter), and 31 children underwent lung transplant. All children with Potts shunt had suprasystemic right ventricle pressures despite maximal medical treatment. In the majority of patients, the Potts shunt was performed through a left thoracotomy approach (90%, 18/20), by direct anastomosis (65%, 13/20), and without the use of extracorporeal support (65%, 13/20). Perioperative outcomes after Potts shunt were superior to lung transplant including mechanical ventilation time (1.3 vs 10.2 days, $P = .019$), median hospital length of stay (9.8 vs 34 days, $P = .012$), and overall complication rate (35% [7/20] vs 81% [25/31], $P = .003$). Risk factors for operative mortality after Potts shunt (20%, 4/20; compared with 6%, 2/31 for lung transplant, $P = .195$) included preoperative extracorporeal membrane oxygenation and significant right ventricle dysfunction. In midterm follow-up (median 1.8, maximum 6.1 years), patients with Potts shunt had durable equalization of right ventricle/left ventricle pressures and improved functional status. There was no significant survival difference in patients with Potts shunt and patients with lung transplant ($P = .258$).

Conclusions: Potts shunt is an effective palliation for children with suprasystemic pulmonary hypertension that may become part of a strategy to maximize longevity and functional status for these challenging patients. (J Thorac Cardiovasc Surg 2021;161:1139-48)



Survival after Potts shunt versus lung transplant in children with pulmonary hypertension.

CENTRAL MESSAGE

Potts shunt is shown to be an effective palliation for children with suprasystemic pulmonary hypertension that can be used to maximize longevity and functional status for these challenging patients.

PERSPECTIVE

For children with severe pulmonary hypertension, addition of Potts shunt to a comprehensive palliation strategy might improve outcomes afforded by medications and delay need for lung transplantation. We report encouraging perioperative and midterm outcomes after Potts shunt, and discuss several considerations important for incorporating Potts shunt as a bridge to lung transplant.

See Commentaries on pages 1149, 1150, 1152, and 1153.

Pulmonary arterial hypertension (PAH) in children carries a poor prognosis with high rates of morbidity and mortality. Although survival has improved over the past 2 decades largely due to improvements in medical therapy, children with PAH continue to have 5-year survival of 50% to 70% depending on diagnostic category.[1-3] Those children

From the [a]Section of Pediatric Cardiothoracic Surgery, Washington University School of Medicine, St Louis Children's Hospital, St Louis, Mo; [b]Division of Pediatric Cardiology, Washington University School of Medicine, St Louis Children's Hospital, St Louis, Mo; and [c]Division of Pediatric Allergy, Immunology, and Pulmonary Medicine, Washington University School of Medicine, St Louis Children's Hospital, St Louis, Mo.

Read at the 100th Annual Meeting of The American Association for Thoracic Surgery: A Virtual Learning Experience, May 22-23, 2020.

Received for publication June 8, 2020; revisions received Oct 16, 2020; accepted for publication Oct 26, 2020; available ahead of print Dec 9, 2020.

Address for reprints: Pirooz Eghtesady, MD, PhD, Section of Pediatric Cardiothoracic Surgery, Washington University School of Medicine, St Louis Children's Hospital, Suite 6120, One Children's Place, St Louis, MO 63110 (E-mail: eghtesadyp@wustl.edu).

0022-5223/$36.00

https://doi.org/10.1016/j.jtcvs.2020.10.163

Abbreviations and Acronyms

BNP = brain natriuretic peptide
CPB = cardiopulmonary bypass
ECMO = extracorporeal membrane oxygenation
LPA = left pulmonary artery
LV = left ventricle
MRI = magnetic resonance imaging
PA = pulmonary artery
PAH = pulmonary arterial hypertension
RV = right ventricle
6MWD = 6-minute walk distance
WHO = World Health Organization

Scanning this QR code will take you to the table of contents to access supplementary information. To view the AATS Annual Meeting Webcast, see the URL next to the webcast thumbnail.

CONG

with medication-refractory disease and suprasystemic pulmonary artery (PA) pressures have especially high mortality.

Surgical management of patients with end-stage pulmonary hypertension typically has included only lung transplantation, which itself carries suboptimal long-term survival. Median survival for pediatric lung transplant recipients remains approximately 7 years.[4] Lung transplantation also carries the significant challenges of limited donor availability, potentially long waitlist times, and requirement for lifelong immunosuppression. In a prior review of our institutional experience, 27% of children with pulmonary hypertension listed for lung transplant died on the waiting list, and those patients more commonly had suprasystemic right heart pressures.[5]

The Potts shunt has been proposed and used over the past several years as an alternative surgical palliation for children with end-stage pulmonary hypertension.[6-8] Potts shunt is a surgical anastomosis between the left pulmonary artery (LPA) and descending aorta, first described as neonatal palliation to allow left-to-right shunting for cyanotic congenital heart disease.[9] The rationale for repurposing this technique to treatment of children with PAH comes from observations that patients with Eisenmenger syndrome have prolonged survival relative to those with primary pulmonary hypertension[10,11] and that patients with Eisenmenger syndrome with post-tricuspid shunts (ventricular septal defect, patent ductus arteriosus) have prolonged survival relative to those with pretricuspid (atrial level) shunts.[12]

When used for PAH, the physiologic effect of the Potts shunt is to create an avenue for right-to-left shunting, providing a pop-off for the right ventricle (RV) and allowing PA pressures to reduce to systemic levels. Overall cardiac output can be improved by reducing both RV afterload and left ventricle (LV) compromise due to leftward bowing of the ventricular septum from a high-pressure RV. Progression to RV failure also may be delayed or avoided by reducing the chronic effects of high RV afterload. Atrial level shunts, in contrast, do not reduce RV afterload or improve LV geometry and can result in large, unregulated shunts producing significant cyanosis. Advantages of the Potts shunt include a more controllable and consistent shunt size, and desaturation of only the lower body because of its postductal position. Potts shunt also does not preclude future lung transplantation and can serve as a life-prolonging bridge to transplant, and in some patients can be performed by a transcatheter technique.

The only major experience to date of the Potts shunt applied to pulmonary hypertension is a recent multicenter European report from the groups in Paris and Rome, who reported the combined outcomes of 24 patients across 3 pediatric centers. In a median follow-up of 2.1 years, they reported 83% overall survival and improvements in symptoms, functional class, and 6-minute walk distance (6MWD).[7]

On the basis of these early encouraging results, we incorporated offering the Potts shunt as part of our pediatric Pulmonary Hypertension and Lung Transplant programs in 2013. In this report, we aimed to review the midterm outcomes of our experience with the Potts shunt for pediatric pulmonary hypertension and to compare perioperative and follow-up outcomes of the Potts shunt with our lung transplant experience for a comparable patient group. This report details the largest single-institution and first North American experience of the Potts shunt for pulmonary hypertension, as well as the largest reported cohort of lung transplants for pediatric pulmonary hypertension.

MATERIALS AND METHODS

This was a prospective, observational study of all pediatric patients (<21 years of age) who underwent Potts shunt or bilateral lung transplant for pulmonary hypertension from 1995 to present. Our first Potts shunt was performed in 2013. Infants undergoing lung transplant for pulmonary vein stenosis were excluded, because we have not offered Potts shunt to this patient group. The study was approved by the Institutional Review Board of Washington University School of Medicine, with a waiver of patient consent.

Indications for Potts Shunt

Patients who underwent Potts shunt were initially referred for evaluation through our pediatric Pulmonary Hypertension and Lung Transplant programs. All patients were managed with combination medical therapy including endothelin receptor antagonists, phosphodiesterase type-5 inhibitors, or prostanoids (intravenous, subcutaneous, or inhaled). Most patients were also managed with diuretics, and some with inotropes, before surgery.

Those who were offered Potts shunt had progressive suprasystemic pulmonary hypertension despite maximal medical therapy, as evidenced by worsening symptoms or by progressive right heart changes. Three patients underwent attempted salvage Potts shunt after presenting with pulmonary hypertensive crises or decompensated right heart failure requiring extracorporeal membrane oxygenation (ECMO). Initially, Potts shunt was offered only to patients who were not lung transplant candidates; however, offering was soon broadened to include patients who were transplant candidates but preferred to avoid or delay transplantation.

Clinical Evaluation and Follow-up

Preoperative evaluation of Potts shunt recipients included clinical assessment for symptoms of right heart failure, functional status (classified by World Health Organization [WHO] functional class), 6MWD, and brain natriuretic peptide (BNP) levels. Diagnostic evaluation included echocardiography and right heart catheterization for hemodynamic assessment, and chest computed tomography to evaluate the size and anatomy of aorta and pulmonary arteries. Later in the series, some patients underwent cardiac magnetic resonance imaging (MRI) for additional assessment of RV function, which also substituted for computed tomography scanning.

Postoperative follow-up included clinical evaluation and echocardiography at our institution or with referring cardiologists, at least annually. Eight patients had follow-up right heart catheterization as part of their ongoing care, although this was not obtained routinely.

Procedural Details

Surgical Potts shunt. The majority of the surgical Potts shunt procedures were performed as we have previously described,[13] which is through a left posterolateral thoracotomy at the fourth interspace. The descending aorta and LPA are carefully mobilized, and the shunt is constructed at the site of their nearest apposition, which is typically just proximal to the upper lobe PA branch. A direct anastomosis between the aorta and the LPA is usually performed, although a short polytetrafluoroethylene conduit may be used to address scenarios where the aorta and LPA cannot be directly anastomosed without excessive tension. We leave a silk ligature and snare loosely around the anastomosis and secure it to the anterior chest wall to facilitate localization and control of the shunt in the case of future lung transplant. The procedure is most often performed without extracorporeal support, although ECMO or cardiopulmonary bypass (CPB) may be used in selected scenarios. The shunt is sized to be 80% to 90% of the diameter of the descending aorta, with the goal of obtaining a 10% to 15% differential between upper and lower extremity oxygen saturations.

Late in the series, we have modified our preferred approach to involve placement of a valved conduit via a median sternotomy approach with use of CPB support (see "Discussion").

Transcatheter Potts shunt. Only patients with a patent ductus arteriosus were considered for transcatheter Potts shunt, which was performed by bare metal stenting of the ductus.[14,15] We have not offered transcatheter Potts shunt creation in the absence of an existing ductus arteriosus, as others have reported, out of concern for procedural complications.[16]

Lung transplant. All lung transplants were performed as bilateral sequential lung transplants via bilateral transverse thoracosternotomy or median sternotomy, using full CPB support.

Statistical Analysis

Statistical analyses were performed using SPSS Statistics, version 23.0 (IBM-SPSS Corp, Armonk, NY). Demographic, perioperative, and follow-up data were compared between the Potts shunt and lung transplant groups. Transcatheter Potts shunt recipients were included for long-term follow-up and survival outcomes, but were excluded from the perioperative outcomes analysis because our intent was to compare procedural details of surgical Potts shunt with lung transplant. Operative mortality was defined as death within 30 days of operation or before hospital discharge. Continuous variables were expressed as mean ± standard deviation or as median with interquartile range. Means were compared using the 2-sample t test, and medians were compared using the Mann–Whitney U statistic. Categorical variables were expressed as frequencies and percentages, and compared using Fisher exact test. Kaplan–Meier survival curves were constructed and compared with the log-rank test.

RESULTS

Baseline Characteristics

In total, we performed Potts shunt in 23 children (20 surgical, 3 transcatheter). Over the course of the study period, lung transplant was performed in 31 children with a primary diagnosis of pulmonary hypertension. Table 1 illustrates clinical characteristics and hemodynamic data for the Potts shunt patient group at baseline (pre/perioperative) and latest follow-up. At baseline, in all children with Potts shunt and WHO functional class 3 or greater, 45% required supplemental oxygen and approximately 20% were mechanically ventilated before surgery. Three patients in the series were on preoperative ECMO, as detailed next. BNP levels were elevated at baseline reflecting some degree of RV dysfunction in most patients. All children were on combination medical therapy, most with all 3 major categories of pulmonary hypertension medications. Preoperative echocardiogram estimated suprasystemic RV pressures by tricuspid regurgitation jet in all patients, and 35% had at least moderate RV dysfunction. Preoperative catheterization demonstrated suprasystemic right-heart pressures, as well as markedly elevated and fixed pulmonary vascular resistance on maximal medical therapy. Cardiac MRI was obtained in 8 patients, and on average showed severe RV dilation, moderate RV dysfunction, and preserved LV function.

Procedural Data and Short-Term Outcomes

Table 2 illustrates perioperative data and immediate postoperative outcomes for the children undergoing Potts shunt compared with those who underwent lung transplant. In this analysis, preoperative characteristics of the transcatheter Potts patients were included, but their procedural and immediate postoperative outcomes were excluded to allow comparison between surgical Potts shunt and lung transplant. Mean patient age was 10.3 years (range, 6 weeks to 20 years) for Potts shunt recipients and 7.9 years (range, 7 months to 17 years) for lung transplant recipients. Approximately 40% of patients in each group had a prior diagnosis of congenital heart disease. Three children in the Potts shunt group were on preoperative ECMO after presenting with pulmonary hypertensive crisis or decompensated RV failure, and underwent Potts shunt as an attempted salvage therapy because they were not lung transplant candidates.

TABLE 1. Clinical and hemodynamic data for patients undergoing Potts shunt at baseline (preoperative/perioperative) and last follow-up (median follow-up 1.8 years, maximum 6.1 years)

	Preoperative/perioperative	Last follow-up	*P* value
N	23	18	
WHO functional class			
≥3	**100% (23/23)**	**17% (3/18)**	**<.001**
≥4	30% (7/23)	0% (0/18)	.232
6MWD (m)	412 ± 72	394 ± 96	.645
BNP (ng/L)	**799 ± 932**	**170 ± 218**	**.02**
Hemoglobin (g/dL)	**13.2 ± 2.9**	**15.7 ± 2.9**	**.026**
Medications			
Endothelin receptor antagonist	78% (18/23)	93% (14/15)	.371
Phosphodiesterase type-5 inhibitor	91% (21/23)	87% (13/15)	1.0
Prostanoids (IV/SQ/inhaled)	**91% (21/23)**	**53% (8/15)**	**.016**
Prostanoids (oral)	0% (0/23)	20% (3/15)	.053
Diuretic	52% (12/23)	80% (12/15)	.101
Echocardiogram			
Estimated RVP (mm Hg)	102 ± 21	91 ± 14	.115
SBP (mm Hg)	97 ± 12	105 ± 9	.052
≥ moderate RV dysfunction	35% (8/23)	22% (4/18)	.724
LV shortening fraction (%)	52 ± 12	47 ± 8	.206
Catheterization			
RVSP (mm Hg)	104 ± 20	90 ± 17	.104
LVSP (mm Hg)	82 ± 11	89 ± 13	.168
RVSP-LVSP differential (mm Hg)	**22 ± 16**	**1 ± 13**	**.003**
mPAP (mm Hg)	73 ± 16	69 ± 14	.61
PCWP (mm Hg)	13 ± 6	12 ± 8	.798
PVRi (WU*m^2)	21 ± 15	17 ± 3	.563
Cardiac index (L/min/m^2)	3.3 ± 1.1	3.2 ± 0.7	.856
Cardiac MRI (n = 8)			
RV EDVi (mL/m^2)	203 ± 77		
RV EF (%)	30 ± 12		
LV EF (%)	55 ± 8		
Postoperative oxygen saturation (%)			
Upper extremity	93 ± 4	94 ± 5	.354
Lower extremity	83 ± 7	84 ± 5	.633
Differential	10 ± 6	10 ± 5	.927

Statistically significant variables are shown in bold. *WHO*, World Health Organization; *BNP*, brain natriuretic peptide; *IV*, intravenous; *SQ*, subcutaneous; *RVP*, right ventricle pressure; *SBP*, systolic blood pressure; *RV*, right ventricle; *LV*, left ventricle; *RVSP*, right ventricle systolic pressure; *LVSP*, left ventricle systolic pressure; *mPAP*, mean pulmonary artery pressure; *PCWP*, pulmonary capillary wedge pressure; *PVRi*, indexed pulmonary vascular resistance; *MRI*, magnetic resonance imaging; *EDVi*, indexed end-diastolic volume; *EF*, ejection fraction.

The Potts shunt was performed via a left thoracotomy approach in 90% (18/20) and via median sternotomy in the remaining 10% (2/20). A direct anastomosis between LPA and aorta was performed in 65% (13/20), whereas a conduit was used in the remaining 35% (7/20). In 3 of these patients, a valved conduit was placed as the Potts shunt; 65% of Potts shunt recipients had the procedure completed without the use of extracorporeal support. In 5 patients, ECMO via neck cannulation was used for intraoperative support (including the 3 children who were on preoperative ECMO and the first 2 children in the series because of the novelty of the procedure). Full CPB was used in 2 other patients: One was an adolescent with a 4.0-cm aneurysmal LPA that could not be safely clamped without unloading, and another required simultaneous pulmonary valve replacement for moderate pulmonary regurgitation. All lung transplants were performed with full CPB support. Total operative time was markedly less for the Potts shunt recipients compared with lung transplant recipients.

Despite their preoperative morbidity, children generally recovered well from the Potts shunt; 30% were extubated in the operating room, and the duration of mechanical ventilation, hospital length of stay, and major complication rates were all substantially lower than for lung transplant. Of the 7 patients who had major complications after Potts shunt, 4 had prolonged ventilation (>7 days), 1 was reintubated, 2

TABLE 2. Perioperative characteristics and immediate postoperative outcomes for children undergoing Potts shunt compared with lung transplant

	Potts shunt	Lung transplant	*P*
N	23	31	
Age (y)	10.3 ± 6.2	7.9 ± 5.6	.14
Prior congenital heart disease diagnosis	43% (10/23)	39% (12/31)	1.0
Atrial septal defect	17% (4/23)	19% (6/31)	1.0
Ventricular septal defect	9% (2/23)	13% (4/31)	1.0
Patent ductus arteriosus	13% (3/23)	6% (2/31)	.64
Atrioventricular canal defect	4% (1/23)	3% (1/31)	1.000
D-transposition of great arteries	4% (1/23)	0% (0/31)	.426
Shone's complex	4% (1/23)	0% (0/31)	.426
Total anomalous pulmonary venous return	0% (0/23)	13% (4/31)	.128
Prior cardiovascular surgery	17% (4/23)	29% (9/31)	.358
Preoperative			
Mechanical ventilation	17% (4/23)	16% (5/31)	1.0
ECMO	13% (3/23)	3% (1/31)	.301
Intraoperative			
Extracorporeal support	**35% (7/20)**	**100% (31/31)**	**<.001**
CPB	29% (2/7)	100% (31/31)	
ECMO	71% (5/7)		
Operative time (min)	**192 ± 67**	**509 ± 245**	**<.001**
Postoperative			
Extubated in operating room	**30% (6/20)**	**0% (0/31)**	**.002**
Mechanical ventilation time (d, median)	**1.3 (0.25-5.8)**	**10.2 (4.4-20.1)**	**.019**
Hospital length of stay (d, median)	**9.8 (6.5-14.3)**	**34 (15-60)**	**.012**
Major complications	**35% (7/20)**	**81% (25/31)**	**.003**
Operative mortality	20% (4/20)	6% (2/31)	.195

Statistically significant variables are shown in bold. *ECMO*, Extracorporeal membrane oxygenation; *CPB*, cardiopulmonary bypass.

had reexplorations for postoperative bleeding, 1 had cardiac arrest, and 1 had distal spinal cord ischemia with lower-extremity weakness that was improving at last follow-up (likely related to hypotension during aortic clamping).

There were 4 early mortalities in the surgical Potts shunt group. This included all 3 patients who were on preoperative ECMO for whom Potts shunt was attempted as potential salvage therapy, and a fourth patient who had significant preoperative RV dysfunction and did not recover postoperatively. One patient in the transcatheter Potts shunt group also had early in-hospital mortality, a premature infant with interstitial lung disease and idiopathic PAH who died of progressive lung disease and hypoxia.

Survival Analysis and Late Outcomes

Median and maximum follow-up durations were 1.8 and 6.1 years for the Potts shunt group and 3.3 and 12.4 years for the lung transplant group, respectively. Kaplan–Meier survival analysis for the overall patient cohort showed that postoperative survival of the patients undergoing Potts shunt was not different than that of lung transplant recipients (Figure 1, *A*).

Given that there was early mortality for all 3 patients for whom Potts shunt was attempted after preoperative ECMO, our institutional practice has changed to no longer offer Potts for acutely decompensated children. Therefore, an additional survival analysis excluding patients on preoperative ECMO was performed to better characterize expected outcomes with our more current patient selection practices. In this analysis, survival after Potts shunt is certainly as good as lung transplant and shows a trend toward improved survival in longer-term follow-up (Figure 1, *B*).

Finally, we assessed clinical and hemodynamic outcomes for the Potts shunt recipients in late follow-up (Table 1). The 5 patients who had early mortality after Potts shunt were excluded from this analysis. There was a significant improvement in functional capacity, with only 17% (3/18) of patients in WHO functional class 3 at last follow-up, as well as a significant reduction in BNP levels. There was an expected polycythemia due to chronic lower body desaturation. 6MWDs did not change significantly, although these data were only available in half (9/18) of the postoperative patients. The majority (83% [15/18]) of patients reported subjective improvement in their overall functional status and absence of symptoms of right heart failure. Of the other 3, 1 reports no subjective improvement but has remained clinically stable throughout follow-up, whereas 2 had progressive

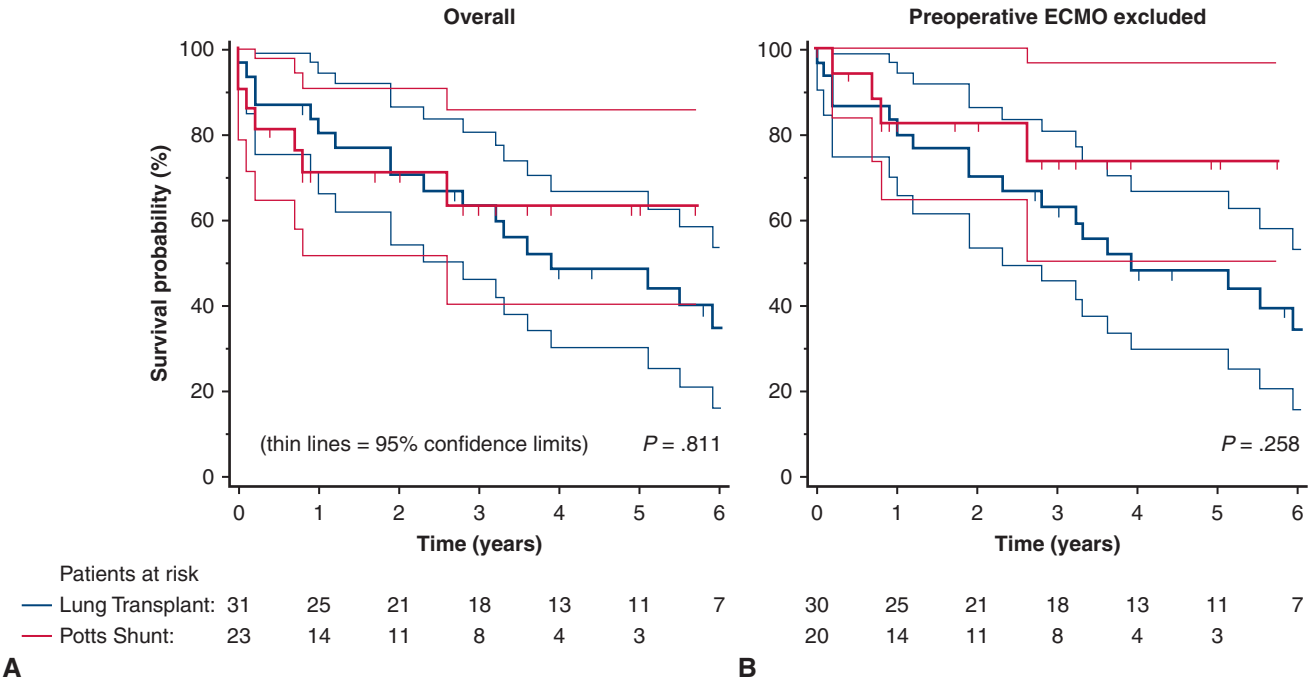


FIGURE 1. Kaplan–Meier survival analysis comparing children undergoing Potts shunt versus lung transplant for pulmonary hypertension: (A) overall patient cohort and (B) patients on preoperative ECMO excluded. *ECMO*, Extracorporeal membrane oxygenation.

RV dysfunction and were listed for lung transplant (1 underwent successful transplant at 6 months post-Potts, 1 died on the waiting list of a viral pneumonia and acute cardiorespiratory failure).

By last echocardiogram and catheterization, right-heart pressures were consistently reduced to systemic levels, with elimination of the differential between RV and LV systolic pressures. A consistent 10% oxygen saturation differential was seen throughout follow-up, indicating durable patency of the shunt.

DISCUSSION

In this report, we reviewed our experience to date of 23 children who have undergone Potts shunt for pulmonary hypertension and compared them with 31 children who underwent lung transplantation at our institution for the same diagnosis. Key findings of the present study were that (1) Potts shunt can be performed with overall low operative morbidity and markedly lower burdens of intraoperative extracorporeal support, operative time, postoperative mechanical ventilation, hospital length of stay, and overall complications compared with lung transplant; (2) Potts shunt led to improvements in functional status and symptom resolution in most children, and created a demonstrable and durable hemodynamic effect with equalization of left and right heart pressures and a stable extremity oxygen saturation differential that persisted through late follow-up; (3) early mortality after Potts shunt occurred in those children with acute preoperative decompensation requiring extracorporeal support and those with substantial preoperative RV dysfunction, indicating that timing of undertaking the Potts shunt and patient selection with attention to these characteristics are critical to optimal outcomes; and (4) to date, late survival after the Potts shunt has been at least equivalent to survival after lung transplant and showed a trend toward superior late survival after excluding those children in whom Potts was used as attempted rescue from preoperative ECMO (Figure 2, shows a pictorial summary of these key findings).

We have established that the Potts shunt is an effective alternative surgical palliation to lung transplant in pediatric patients with suprasystemic pulmonary hypertension. Although the Potts shunt has now, in our practice, become an established part of the management strategy for children with end-stage pulmonary hypertension, we expect continued evolution of the surgical technique to maximize its potential benefits. Two major considerations for the future management of these patients are (1) how best to perform the Potts shunt procedure so that the safety and simplicity of a potential future lung transplant can be maximized, which would make the use of Potts shunt as a true "bridge" to lung transplantation a more tenable proposition; and (2) how best to perform the procedure so that the physiologic benefit to the RV is maximized, thereby extending the time until transplant is required and maximizing overall longevity.

One distinct advantage of the Potts shunt is that it does not preclude future lung transplantation. Although the

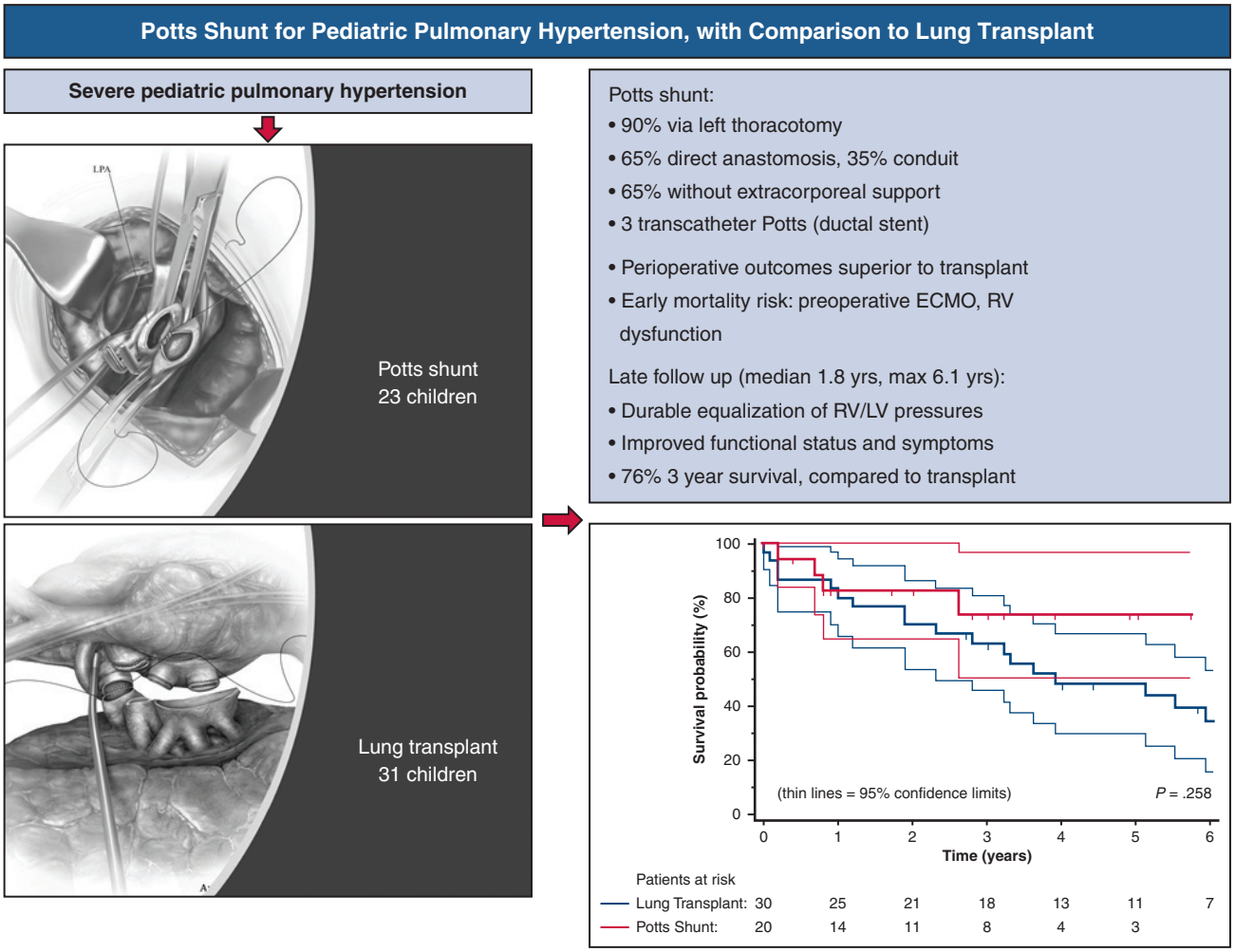
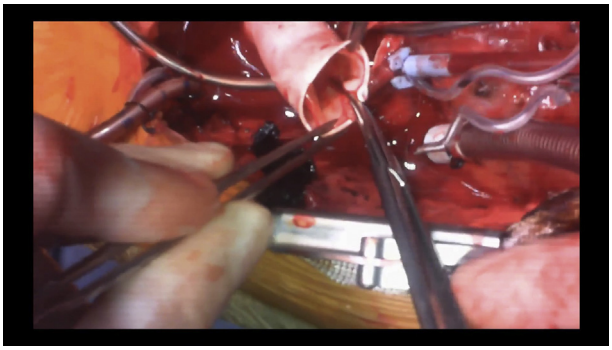


FIGURE 2. Our experience with the Potts shunt for severe pediatric pulmonary hypertension in comparison with lung transplant is shown. *ECMO*, Extracorporeal membrane oxygenation; *LV*, left ventricle; *RV*, right ventricle.

world experience with lung transplant after Potts shunt is admittedly small, we have performed 2 such procedures, 1 after direct anastomosis and 1 after a conduit Potts. Localization, control, and takedown of the Potts shunt are important considerations at the time of lung transplant that unavoidably add complexity and risk compared with a primary lung transplant. Especially if the Potts shunt has been performed through a left thoracotomy, there can be adhesions in the left pleural space and in the left pulmonary hilum from the previous dissection. Control of the shunt is necessary before initiating CPB to prevent substantial arterial runoff into the pulmonary circulation, but localization and encircling of the shunt can be hazardous especially in the case of a direct anastomosis Potts. To address these challenges, we have left a loose silk ligature with a tourniquet around the Potts anastomosis or conduit and secured it to the anterior chest wall at the time of the Potts operation, which facilitates identification and enables control of the shunt when CPB is initiated. Even with this in place, shunt takedown at the time of recipient pneumonectomy can still present a challenge. Possible methods for takedown of the shunt include external ligation with suture or vascular staplers (which we have used), or internal closure through the PA under brief circulatory arrest, all of which were described as early as the 1960s.[17-19] Others have reported transcatheter occlusion of the Potts shunt immediately before lung transplant by placement of a covered descending aortic stent graft.[20] The latter, however, can only be performed in older children not anticipated to have further growth. The median sternotomy approach with placement of a conduit Potts shunt, as reported here and by others,[21] would certainly allow easier access and control, although the lack of growth potential of a conduit Potts shunt also limits that approach to older children.

Efforts to maximize RV protection and prolong tolerance of Potts shunt physiology center around timing of shunt



VIDEO 1. Surgical video detailing technical steps of valved Potts shunt placement via a median sternotomy approach. Video available at: https://www.jtcvs.org/article/S0022-5223(20)33316-X/fulltext.

placement and possibly consideration of using a valved communication. A healthy RV can be remarkably tolerant of systemic arterial pressures, as has been demonstrated for patients with Eisenmenger syndrome and post-tricuspid shunts, who have documented survival into the seventh decade and exhibit RV physiology comparable to fetal life with maintained RV wall thickness, flat ventricular septum, and preserved RV function despite lifelong exposure to systemic pressures.[11,12] Prolonged exposure to suprasystemic pressures is poorly tolerated, however, and intervention should be pursued early enough to prevent significant RV deterioration because it is unlikely for an already dysfunctional RV to substantially improve with Potts shunt given continued exposure to systemic arterial pressures.[22] The concept of using a unidirectional valved conduit as the Potts shunt might help to maximize RV protection even further, as it is likely that a nonvalved Potts shunt allows some dynamic left-to-right shunting during diastole when PA diastolic pressures drop below aortic diastolic pressures. A unidirectional valved conduit in theory allows only right-to-left shunting and may prevent this diastolic runoff into the pulmonary circulation. The valved conduit Potts shunt was first evaluated in animal studies[23] and first used in patients by the group from Marie Lannelongue.[7] The Columbia group has also recently reported success with this approach in their initial experience of 5 patients.[21] We have similarly had success with this approach, using a bovine jugular vein conduit (Contegra, Medtronic, Minneapolis, Minn) supported inside of a polytetrafluoroethylene graft to prevent future dilation and valvar incompetence (Video 1).

CONCLUSIONS

In children with end-stage pulmonary hypertension, the Potts shunt yielded durable alleviation of suprasystemic RV pressures and improved functional capacity, was associated with no difference in survival when compared with lung transplant, and was shown to be a viable component of a developing palliation strategy that may help delay lung transplant and extend longevity and quality of life for these patients. Limitations of the study include the relatively small sample size (although the largest of this patient population reported to date), relatively short follow-up period, and the possibility of an era effect in the management of the early lung transplant recipients. Ongoing work in this area will focus on ways to enhance patient selection and further decrease morbidity, which will help to yield the maximal benefit from a staged palliation strategy that incorporates Potts shunt and lung transplantation.

Webcast

You can watch a Webcast of this AATS meeting presentation by going to: https://aats.blob.core.windows.net/media/20AM/Presentations/Mid-term%20Outcomes%20of%20the%20Potts%20Shunt.mp4.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

1. Benza RL, Miller DP, Barst RJ, Badesch DB, Frost AE, McGoon MD. An evaluation of long-term survival from time of diagnosis in pulmonary arterial hypertension from the REVEAL registry. *Chest*. 2012;142:448-56.
2. Haworth SG, Hislop AA. Treatment and survival in children with pulmonary arterial hypertension: the UK pulmonary hypertension service for children 2001-2006. *Heart*. 2009;95:312-7.
3. Zijlstra WMH, Douwes JM, Rosenzweig EB, Schokker S, Krishnan U, Roofthooft MTR, et al. Survival differences in pediatric pulmonary arterial hypertension: clues to a better understanding of outcome and optimal treatment strategies. *J Am Coll Cardiol*. 2014;63:2159-69.
4. Rossano JW, Singh TP, Cherikh WS, Chambers DC, Harhay MO, Hayes D Jr, et al. The international thoracic organ transplant registry of the International Society for Heart and Lung Transplantation: twenty-second pediatric lung and heart-lung transplantation report-2019; focus theme: donor and recipient size match. *J Heart Lung Transplant*. 2019;38:1015-27.
5. Goldstein BS, Sweet SC, Mao J, Huddleston CB, Grady RM. Lung transplantation in children with idiopathic pulmonary arterial hypertension: an 18-year experience. *J Heart Lung Transplant*. 2011;30:1148-52.
6. Blanc J, Vouhe P, Bonnet D. Potts shunt in patients with pulmonary hypertension. *N Engl J Med*. 2004;350:623.
7. Baruteau AE, Belli E, Boudjemline Y, Laux D, Lévy M, Simonneau G, et al. Palliative Potts shunt for the treatment of children with drug-refractory pulmonary arterial hypertension: updated data from the first 24 patients. *Eur J Cardiothorac Surg*. 2015;47:e105-10.
8. Grady RM, Eghtesady P. Potts shunt and pediatric pulmonary hypertension: what we have learned. *Ann Thorac Surg*. 2016;101:1539-43.

9. Potts WJ, Smith S, Gibson S. Anastomosis of the aorta to a pulmonary artery; certain types in congenital heart disease. *J Am Med Assoc*. 1946;132:627-31.
10. Hopkins WE, Ochoa LL, Richardson GW, Trulock EP. Comparison of the hemodynamics and survival of adults with severe primary pulmonary hypertension or Eisenmenger syndrome. *J Heart Lung Transplant*. 1996;15:100-5.
11. Hopkins WE, Waggoner AD. Severe pulmonary hypertension without right ventricular failure: the unique hearts of patients with Eisenmenger syndrome. *Am J Cardiol*. 2002;89:34-8.
12. Kempny A, Hjortshoj CS, Gu H, Li W, Opotowsky AR, Landzberg MJ, et al. Predictors of death in contemporary adult patients with Eisenmenger syndrome: a multicenter study. *Circulation*. 2017;135:1432-40.
13. Eghtesady P. Potts shunt for children with severe pulmonary hypertension. *Oper Tech Thorac Cardiovasc Surg*. 2015;20:293-305.
14. Boudjemline Y, Patel M, Malekzadeh-Milani S, Szezepanski I, Levy M, Bonnet D. Patent ductus arteriosus stenting (transcatheter Potts shunt) for palliation of suprasystemic pulmonary arterial hypertension: a case series. *Circ Cardiovasc Interv*. 2013;6:e18-20.
15. Latus H, Apitz C, Moysich A, Kerst G, Jux C, Bauer J, et al. Creation of a functional Potts shunt by stenting the persistent arterial duct in newborns and infants with suprasystemic pulmonary hypertension of various etiologies. *J Heart Lung Transplant*. 2014;33:542-6.
16. Esch JJ, Shah PB, Cockrill BA, Farber HW, Landzberg MJ, Mehra MR, et al. Transcatheter Potts shunt creation in patients with severe pulmonary arterial hypertension: initial clinical experience. *J Heart Lung Transplant*. 2013;32:381-7.
17. Kirklin JW, Devloo RA. Hypothermic perfusion and circulatory arrest for surgical correction of tetralogy of Fallot with previously constructed Potts' anastomosis. *Dis Chest*. 1961;39:87-91.
18. Sirak HD, Britt CI. A technic for taking down the Potts' anastomosis. *Circulation*. 1962;25:110-3.
19. Leand PM, Bender HW Jr, Martz MN, Crisler C, Agnew HD, Gott VL. A simple method for closure of the Potts anastomosis with a mechanical stapler. *J Thorac Cardiovasc Surg*. 1971;62:285-9.
20. Paradela M, Mercier O, Baruteau A, Fadel E. Endovascular closure of Potts shunt before double lung transplantation for idiopathic pulmonary arterial hypertension. *J Thorac Cardiovasc Surg*. 2013;146:e5-7.
21. Rosenzweig EB, Ankola A, Krishnan U, Middlesworth W, Bacha E, Bacchetta M. A novel unidirectional-valved shunt approach for end-stage pulmonary arterial hypertension: early experience in adolescents and adults. *J Thorac Cardiovasc Surg*. November 14, 2019 [Epub ahead of print].
22. Delhaas T, Koeken Y, Latus H, Apitz C, Schranz D. Potts shunt to be preferred above atrial septostomy in pediatric pulmonary arterial hypertension patients: a modeling study. *Front Physiol*. 2018;9:1252.
23. Bui MT, Grollmus O, Ly M, Mandache A, Fadel E, Decante B, et al. Surgical palliation of primary pulmonary arterial hypertension by a unidirectional valved Potts anastomosis in an animal model. *J Thorac Cardiovasc Surg*. 2011;142:1223-8.

Key Words: Eisenmenger syndrome, lung transplant, Potts shunt, pulmonary hypertension

Discussion

Presenter: Dr Timothy S. Lancaster

Dr Emre Belli *(Paris, France)*. To transform the idiopathic primary pulmonary hypertension to a ductus arteriosus-related Eisenmenger syndrome type is one of the rare contributions in the new era to our surgical armamentarium, and it is marginal, too, because the patients are rare and managed far from our surgical environment. I have a few quick questions—first of all, technical ones.

You had the 3 patients under ECMO before surgery where you have judged that it can be considered a contraindication for this procedure. However, 4 other patients underwent the Potts procedure with cardiopulmonary support. What was the reason or rationale for this support?

Dr Timothy S. Lancaster *(St Louis, Mo)*. You're right. Three of the patients who underwent the Potts on mechanical circulatory support were the 3 who were on preoperative ECMO. There were 4 others, 2 patients with ECMO. Those were the first 2 patients in the series, in whom ECMO was used for additional support because of the unfamiliarity with the procedure, and those patients tolerated that well.

CPB was used in the 2 remaining patients. One was an older teenager with an extremely dilated PA (~4 cm), which we thought could not be clamped safely without decompression of the right side, so that patient was cannulated femoral vein to descending aorta for partial support. The last one was done on full CPB; that was a patient postarterial switch who had pulmonary hypertension, but also severe pulmonary insufficiency, and we wanted to do pulmonary valve replacement at the same time as the Potts shunt. That was done through a redo sternotomy approach—pulmonary valve replacement and a Potts shunt.

Dr Belli. So 7 patients had no direct anastomosis, but with interposition of a conduit. The spirit of this procedure in particular when you do a through thoracotomy without support is to crossclamp both vessels simultaneously, allowing to adjust right to left pressure ratio. How did you manage this situation? Because if you crossclamp first the PA to the aorta, you can be in trouble to diminish cardiac output. How did you manage technically?

Dr Lancaster. In all the patients, we certainly perform a test clamp of the PA before initiating the anastomosis. We use side-biting clamps for partial occlusion in almost all scenarios. In the patients in whom we use a conduit, we do clamp just 1 vessel at a time and with the combination of test clamping and an experienced anesthesia team, now accustomed to taking care of these patients with supersystemic PA pressures, our patients have done okay with that.

Dr Belli. Moving on to the spirit and philosophy of this procedure: More than one-third of your patients had prior congenital heart surgery. I didn't see the details on the slides. What etiologic type of pulmonary hypertension did you consider?

Dr Lancaster. Three or 4 of them had received repairs of atrial septal defects and ventricular septal defects. There was the 1 patient I alluded to who had a previous arterial switch operation and 1 patient who had an AV canal repair. In all scenarios, the patients did not have residual congenital heart defects that we thought explained their pulmonary hypertension, as in Eisenmenger syndrome. It was believed to

be idiopathic PAH coincidental with preexisting congenital heart disease.

Dr Belli. I wasn't expecting to have a Shone-like hypoplastic—

Dr Lancaster. Yes, there was 1 patient (the first patient) who had a Shone complex.

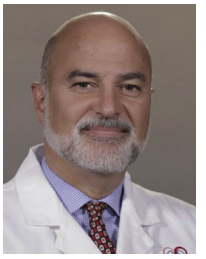
Dr Belli. There is 1 other issue: The pulmonary vascular resistance in some patients as we observe is not stable. That means there is a pulmonary reactivity, and we observed in a couple of patients that the shunt is working or not according to the patient's status. Would you comment on this? Can we have an additional technique to do this type of procedure—avoiding left-to-right but allowing a right-to-left shunt?

Dr Lancaster. Certainly. I know that you've written on that topic. I would say that in the follow-up we've obtained on our patients so far, we haven't seen the problem that you allude to, which would be pulmonary overcirculation due to left-to-right shunting through the Potts shunt. But a concept that has been used in a small number of patients is to use a 1-way valved conduit as the Potts shunt to allow right-to-left shunting and prevent future left-to-right shunting in the case that the PA pressures do become sub-systemic, as you said. I know that you've done that in animals and in a couple humans. We also have used valved conduit in 2 patients so far. The most recent was the postarterial switch patient who had pulmonary valve replacement at the same time that we used a valved conduit for the Potts shunt. In that scenario, we used a bovine jugular vein valved conduit inside a polytetrafluoroethylene graft (essentially a supported Contegra graft) as the conduit. That patient is only a few months out, but he's done very well so far.

Dr Belli. Of course, there is a debate going on—Potts versus transplantation. And nowadays, according to your experience, which type of patient you schedule is more likely for Potts procedure: pathology status, age, and so forth? What are the indications where primarily you would go on Potts rather than listing for a transplant?

Dr Lancaster. At this point, we are often getting referred patients who already have an interest in Potts shunt. So that's definitely 1 factor—the family's preference for or against Potts shunt, or for or against lung transplant. I'd say that in patients who seem to be appropriate candidates (who, as evident from the data that we shared, would be patients with supersystemic pulmonary hypertension, but preserved RV function), I think we would recommend Potts and certainly offer Potts shunt if that's what the family prefers. To better gauge RV function, we are starting to do more cardiac MRI preoperatively to get a better objective assessment of RV ejection fraction and function.

Dr Belli. Thank you.



Dr Emile A. Bacha *(New York, NY).* As you probably know, we published our experience in the *Journal* in 2019 with a valved unidirectional shunt placed via sternotomy on bypass in every single case. We do that because we want to stay away from the pleural spaces for the time when the lung transplant will occur. We think that the valve provides a lot of benefits, as you just discussed. We do those all on CPB and use perioperative ECMO. We've done 6 or 7 cases (5 were described in the article, and in the meantime we've done 2 more). I've been impressed after the initial rocky postoperative course (which is difficult) how well these patients become from a symptomatic standpoint. I mean, they really start to be able to go to school, walk around, and so forth. It is impressive how their RV function improves almost immediately. It goes from very bad RV function on the preoperative TEE to much better function on the intraoperative postoperative TEE. I'm a big fan of this procedure, but I would strongly recommend using the valved conduits.

Dr Lancaster. After our most recent experience with the valved conduit, I think we will likely transition to that being the primary approach, probably through the front on bypass, like you said. We have performed 2 lung transplants after Potts shunt. One was a patient of our own in the series, and another had a Potts shunt completed at an outside facility, and the adhesions in the left chest and the potential hazard of taking down the shunt should not be underestimated. So I think that staying out of pleural spaces is important for future lung transplant.