Pulmonary Circulation

WILEY

PVRi | Pulmonary Circulation Open Access

RESEARCH ARTICLE OPEN ACCESS

Approach to Lung Transplantation in Pulmonary Arterial Hypertension: A Delphi Consensus on Behalf of the Transplant Task Force of the Pulmonary Vascular Research Institute

Nicholas A. Kolaitis[1] | Hayley Barnes[2] | Deborah J. Levine[3] | Howard Castillo[4] | Selim M. Arcasoy[5] | Matthew Bacchetta[6] | Luke Benvenuto[5] | Erika Berman-Rosenzweig[5] | Marisa Cevasco[7] | Caitlin T. Demarest[6] | Celine Dewachter[8] | Michiel E. Erasmus[9] | Allan R. Glanville[10] | John Granton[11] | Shaf Keshavjee[11] | Vikramjit Khangoora[12] | Sheila Krishnan[13] | Olaf Mercier[14] | Andrea N. Miltiades[5] | David Montani[15] | Edward Murphy[13] | Ivan Robbins[6] | Franck F. Rahaghi[16] | Sahar A. Saddoughi[17] | Laurent Savale[15] | Marc A. Simon[1] | Jean-Luc Vachiery[18] | Corey E. Ventetuolo[19] | Helen M. Whitford[2] | Reda E. Girgis[13]

[1]University of California, San Francisco, San Francisco, California, USA | [2]Alfred Hospital, Melbourne, Australia | [3]Stanford University, Palo Alto, California, USA | [4]United Therapeutics Corporation, Silver Spring, Maryland, USA | [5]Columbia University Irving Medical Center, New York-Presbyterian Hospital, New York, New York, USA | [6]Vanderbilt University Medical Center, Nashville, Tennessee, USA | [7]University of Pennsylvania, Philadelphia, Pennsylvania, USA | [8]Universitaire de Bruxelles Erasme, Brussels, Belgium | [9]University Academic Center Groningen, Groningen, the Netherlands | [10]St. Vincent's Hospital, Sydney, Australia | [11]University of Toronto, Toronto, Canada | [12]Inova Fairfax Hospital, Falls Church, Virginia, USA | [13]Corewell Health, Grand Rapids, Michigan, USA | [14]Department of thoracic surgery and Heart-lung transplantation, Marie Lannelongue Hospital, Université Paris-Saclay, Le Plessis Robinson, France | [15]Department of Respiratory and Intensive Care Medicine, Pulmonary Hypertension National Referral Centre, Hôpital de Bicêtre, Université Paris–Saclay, AP-HP, INSERM UMR_S 999, Le Kremlin Bicêtre, France | [16]Cleveland Clinic Florida, Weston, Florida, USA | [17]Mayo Clinic, Rochester, Minnesota, USA | [18]Erasme Academic Hospital, Brussels, Belgium | [19]Brown University, Providence, Rhode Island, USA

Correspondence: Nicholas A. Kolaitis (nicholas.kolaitis@ucsf.edu)

Received: 7 January 2025 | **Revised:** 26 March 2025 | **Accepted:** 16 April 2025

Funding: The authors received no specific funding for this work.

ABSTRACT

Lung transplantation is indicated for selected patients with advanced pulmonary arterial hypertension (PAH). We used a modified Delphi process to develop recommendations on care of patients with PAH undergoing lung transplantation. This Delphi panel was recruited from the Pulmonary Vascular Research Institute's Innovative Drug Discovery Initiative - Lung Transplantation Workstream, consisting of clinical and research experts in PAH and lung transplantation. In this process, 29 panelists were given open-ended questions, querying topics related to lung transplantation in PAH. A steering group converted the responses into discrete statements. Panelists then rated agreement using a Likert scale in two further survey rounds: −5 (strongly disagree) to 5 (strongly agree). Consensus was defined as mean ≥ 2.5 or ≤ −2.5, with a standard deviation not crossing zero. Consensus was reached on 141 of 223 statements. Notable areas of consensus were for early discussions about transplantation, and agreement with previously published referral and listing criteria. There was agreement that lung transplantation could be offered in sick candidates, including those with concurrent renal or hepatic insufficiency. Bilateral lung transplantation was considered the procedure of choice for most patients, with rare indications for heart-lung transplantation. Consensus on bridging strategies included use of veno-arterial extracorporeal membrane oxygenation and preemptive awake cannulation in those with severe right ventricular dysfunction. Consensus was also achieved on intraoperative use of invasive hemodynamic

monitoring, and prolonged postoperative circulatory support guided by hemodynamic response and echocardiography. Patients with PAH undergoing transplantation require specialized management, which differs somewhat from other candidates.

1 | Introduction

Survival has improved in pulmonary arterial hypertension (PAH) due to advances in medical management and recognition of benefits of initial use of combination therapy [1–6]. Although treatment is now more effective, patients remain at risk of death [2, 3]. Lung transplantation is a potential therapeutic option for select patients, offering freedom from parenteral therapy, improved quality of life, and extended survival [7–12].

Annually, over 4500 lung transplants are performed [13]. Amongst indications for transplantation, PAH is unique as it predominantly involves the vascular compartment, and because the presence of elevated pulmonary artery pressure and right ventricular dysfunction are associated with increased perioperative risk [14]. Additionally, since PAH is rare, it also represents a small proportion of the candidate pool, accounting for less than 5% of global lung transplants.

Given the unique challenges facing patients with PAH when undergoing lung transplantation, we sought to create guidance on management. Although multiple professional societies have provided useful advice on the topic, few have provided comprehensive commentary on lung transplantation for PAH, nor made recommendations via a scientific approach to consensus building [7, 15–17]. We performed a modified Delphi study to develop an expert consensus on referral and listing criteria, as well as management of patients with PAH requiring lung transplantation.

2 | Methods

2.1 | Panel Selection

The Innovative Drug Discovery Initiative of the Pulmonary Vascular Research Institute assembles groups of experts on topics relevant to PAH. This modified Delphi project was derived from the Innovative Drug Discovery Initiative Lung Transplantation Workstream, consisting of 34 experts in PAH and/or lung transplantation. In addition, 2 members of the International Society for Heart and Lung Transplantation Pulmonary Vascular Disease Interdisciplinary Network were also invited to join as part of this collaborative effort. All members of the Workstream were invited, and 29 elected to participate (Table 1).

TABLE 1 | Make up of modified Delphi panel.

Clinical practice	
Pulmonologist	18
Cardiologist	3
Lung Transplant Surgeon	5
Anesthesiologist	2
Nurse Practitioner	1
Country	
North America	20
Europe	6
Australia	3
Years involved in transplant (median [IRQ])	16.5 years [7–25]
Years involved in PAH (median [IQR]	18 years [9–25]
Number of transplant performed at centers (median [IQR])	80 transplants/year [45–100]
Number of PH patients cared for at centers (median [IQR])	450 PH patients [300–1200]

2.2 | Modified Delphi Survey Methodology

The Delphi process is intended to create consensus in the absence of medical evidence [18]. It has been utilized in various aspects of pulmonary medicine, including PAH [4, 19–21]. In our study, we used a modified Delphi, with three survey rounds. Each round was moderated by a five-member steering committee: NAK, HB, HC, DL, RG.

In the first survey, participants were given 55 open-ended questions related to uncertainties in the field. The questions were developed by the steering committee after literature review and discussion with the larger panel. Topics in Survey 1 included soliciting commentary on unmet needs, timing of care decisions, candidate selection, supportive care while awaiting transplant, surgical considerations, and perioperative considerations (Table S1).

The steering committee then developed Survey 2, which was a series of 223 discrete statements derived from the responses to Survey 1. No new topics were introduced. Panelists then rated their agreement with these discrete statements using a Likert scale. The range of agreement was −5 to 5, with −5 representing strong disagreement, and 5 representing strong agreement [19, 20, 22, 23].

Survey 3 consisted of the same 223 discrete statements in Survey 2, along with the group mean and standard deviation from Survey 2. In this round, panelist re-scored each statement after considering the aggregated group response.

Upon return of Survey 3, each statement was assessed for consensus. Although there is no standard definition of consensus, we used previously accepted methodology [19, 20, 22, 23]. Consensus agreement was defined as a mean score greater than 2.5 with a standard deviation not crossing zero. Consensus disagreement was defined as a mean score less than −2.5 with a standard deviation not crossing zero.

Panelists were required to participate in at least two surveys and asked to abstain from answering any questions outside their content expertise. The consensus was conceived in October 2023, Survey 1 was distributed in November 2023, Survey 2 was distributed in December 2023, and Survey 3 was distributed in January 2024. This consensus was approved by the Monash University Institutional Review Board (Approval Number: 40697).

3 | Results

Of the 29 panelists, 20 (69%) responded to Survey 1, 28 (97%) responded to Survey 2, and 29 (100%) responded to Survey 3 (Figure 1). Consensus was achieved on 141 of 223 statements (63%), with 126 consensus agreements and 15 consensus disagreements (Table S2).

3.1 | Starting the Transplant Conversation

The panel reached consensus on need for increased awareness amongst PAH providers regarding proper timing of referral (mean agreement 4.15 ± 0.53), and that discussions about transplantation occur too late in the disease course (mean agreement 3.65 ± 0.75). Panelists felt that the transplant conversation should occur at diagnosis or shortly thereafter in high-risk patients by either the European Society of Cardiology/European Respiratory Society (ESC/ERS) 3-stratum model or the REVEAL 2.0 risk calculator. Panelists also felt that the transplant conversation should occur at the time of diagnosis for suspected or confirmed pulmonary veno-occlusive disease/pulmonary capillary hemangiomatous (PVOD/PCH) (Figure 2).

At follow up, panelists felt that the transplant conversation should occur in intermediate-high-risk patients despite appropriate therapy based on the ESC/ERS 4-stratum model or the REVEAL 2.0 risk calculator. Additionally, there was consensus that transplant conversations should occur if patients were deteriorating, starting triple therapy, starting parenteral prostacyclin, were pregnant or considering pregnancy, or had hypoxemia, hemoptysis or signs of end-organ dysfunction such as renal insufficiency or hepatic congestion.

3.2 | Referral for Lung Transplantation

There was unanimous consensus that close collaboration between PAH teams and lung transplant programs is important (mean agreement 5 ± 0). The panel agreed that referral should occur early, allowing for education about transplant and mitigation of barriers (mean agreement 3.96 ± 1.07). There was agreement that patients meeting indications for referral should be referred despite perceived potential contraindications, so transplant centers can determine candidacy (mean agreement 4.43 ± 0.74), and that a second opinion from a program with experience in transplantation for PAH should be obtained if a patient is denied. There was negative consensus for the statement suggesting that providers should wait to refer until the last therapeutic option has been introduced with insufficient effects (mean agreement -3.71 ± 2.17).

Panelists agreed that patients should be referred for transplantation if they were intermediate-high risk by the ESC/ERS 4-stratum model or had a REVEAL 2.0 score of ≥ 8 (intermediate-risk). Panelists also agreed that patients on appropriate therapy who remain World Health Organization Functional Class III-IV should be referred. Consensus was

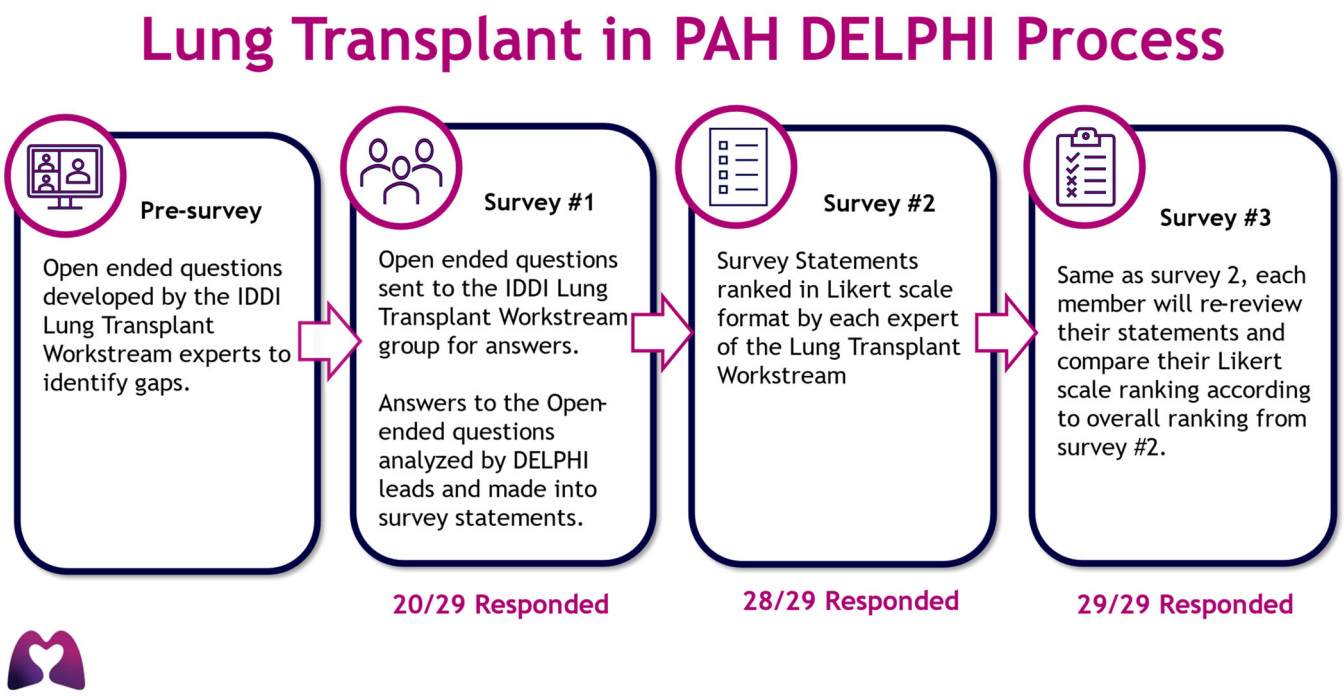

FIGURE 1 | Overview of the Delphi consensus process to develop recommendations on lung transplantation for patients with pulmonary arterial hypertension (PAH). Developed by the Pulmonary Vascular Research Institute Innovative Drug Delivery Initiative (IDDI) Lung Transplantation Workstream.

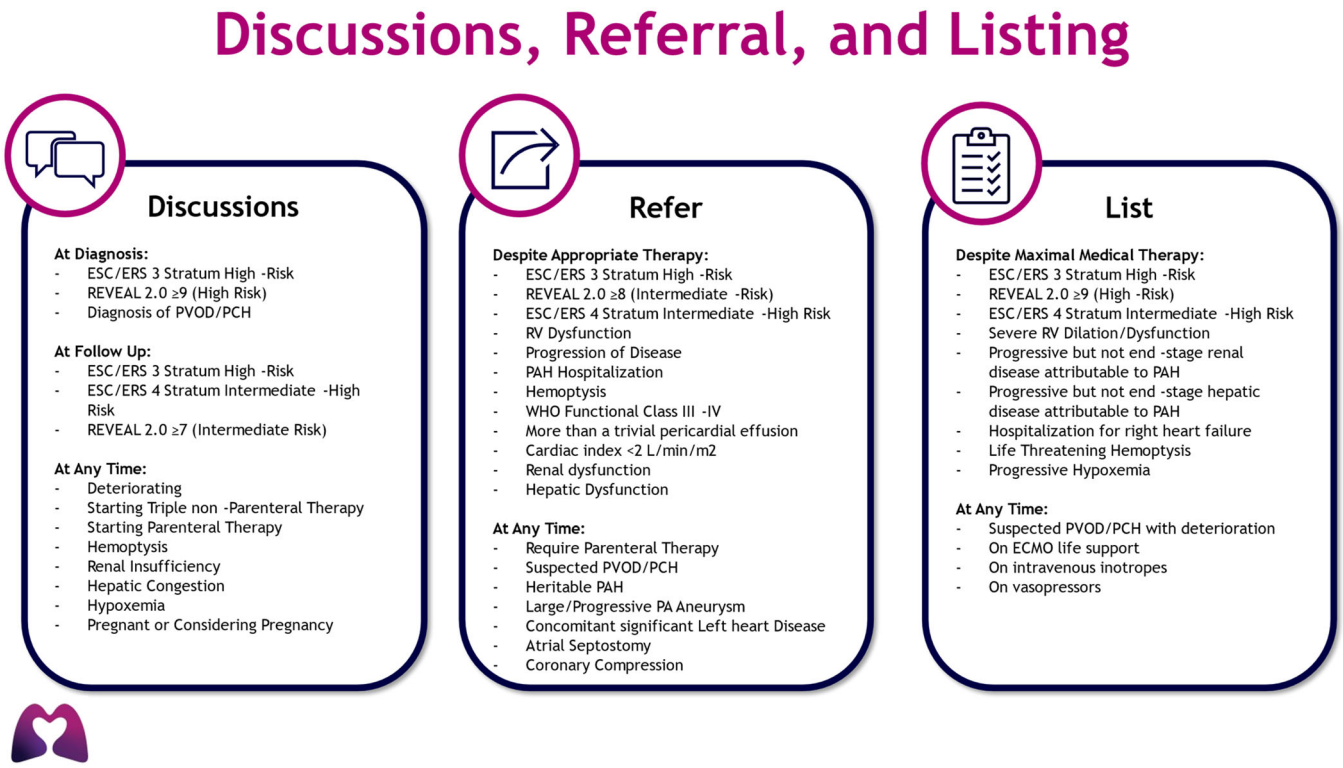

FIGURE 2 | When to start discussions about transplantation in patients with pulmonary arterial hypertension (PAH), when to refer patients with PAH for transplantation, and when to list patients with PAH for transplantation.

achieved that patients should be referred if, despite appropriate therapy, they have right ventricular dysfunction/dilation, disease progression, hospitalization for PAH, hemoptysis, the presence of pericardial effusion, cardiac index below 2 L/min/m^2, or the presence of renal or hepatic dysfunction. Additionally, regardless of therapy, consensus was achieved that patients should be referred if they require parenteral prostacyclin, have suspected or confirmed PVOD/PCH, hereditable PAH, a large or progressive pulmonary aneurysm, significant left heart disease, coronary compression, recurrent arrhythmias, or have undergone atrial septostomy (Figure 2).

3.3 | Contraindications to Transplantation Unique to PAH

Various potential contraindications to transplantation are unique to PAH. Panelists reached consensus that there should be an upper age limit for candidacy but did not reach consensus on the limit. There was negative consensus for the statement that transplantation in PAH should be reserved for patients under 60 (mean agreement -3.42 ± 1.68).

Panelists agreed that patients with drug-associated PAH would need a minimum of 6 months sobriety before they could be offered candidacy for transplant. Uncontrolled bleeding disorders were considered an absolute contraindication. Although thrombocytopenia is common in patients with PAH, providers did not agree on a level of thrombocytopenia representing absolute contraindication. They did reach a negative consensus against the idea that a platelet count under 100,000 is a contraindication.

Panelists felt that patients with mild renal or liver disease could be considered for transplantation, and that consultation with nephrology or hepatology is beneficial. There was consensus agreement that a Chronic Kidney Disease Epidemiology Collaboration (CKD-EPI) below 30 mL/min should prohibit lung transplantation alone, without concomitant kidney transplantation. There was also consensus agreement that patients with confirmed cirrhosis, esophageal varices, significant portal hypertension, bilirubin > 10, or Model for End Stage Liver Disease Score > 20 should not be offered lung transplantation alone, without concomitant liver transplantation.

There was consensus that critical illness is not a contraindication to lung transplantation in PAH, and that use of extracorporeal membrane oxygenation (ECMO) to complete evaluation is appropriate (mean agreement 4.24 ± 0.83). Notably, the panel felt patients on ECMO or mechanical ventilation should only be evaluated if awake enough to consent (mean agreement 3.00 ± 1.92).

3.4 | Listing Criteria

The panel reached consensus that patients with PAH should be listed for transplantation if intermediate-high-risk by the ERS/ESC 4-stratum model, or high-risk (9+ points) by REVEAL 2.0. Additionally, panelists agreed that patients should be listed for transplantation if they have severe right ventricular dilation/dysfunction, progressive but not end-stage renal or hepatic dysfunction, life-threatening hemoptysis, progressive hypoxemia, or PVOD/PCH. Notably, there was strong consensus that patients on ECMO (mean agreement 4.66 ± 0.61) or in those

dependent on intravenous inotropes or vasopressors could be considered for listing (mean agreement 4.36 ± 1.93) (Figure 2).

3.5 | Type of Surgery

Historically, patients with PAH have undergone lung transplantation alone or combined heart-lung transplantation. The panelists found consensus that single lung transplant should not be offered to patients with PAH (mean agreement 3.57 ± 2.03), and that no degree of right ventricular dysfunction should preclude lung transplantation alone (mean agreement 2.86 ± 2.09) (Figure 3).

Use of combined heart-lung transplant was considered appropriate in complex congenital heart disease not amenable to surgical repair, concomitant significant left heart disease, or significant coronary disease not amenable to revascularization. There was negative consensus against the idea that heart-lung transplantation should be performed when patients have an atrial septal defect amenable to surgical repair, an open patent ductus arteriosus amenable to surgical repair, or an atrial septostomy amenable to surgical closure. There was consensus that complex congenital heart disease or Potts shunt patients should undergo transplantation at centers with relevant expertise.

3.6 | Improving Allocation

Panelists felt that current allocation systems underestimate disease severity in PAH (mean agreement 4.32 ± 0.61), that health disparities and unequal access to care impact patients with PAH (mean agreement 4.23 ± 0.71), and that waiting list mortality remains unacceptably high. Although systolic pulmonary artery pressure has historically been included in the US Lung Allocation Score, and is in the Composite Allocation Score [24], the panel did not achieve consensus that it had utility in allocation calculations. Rather, the panel reached agreement that presence of right ventricular dysfunction, cardiac index, stroke volume index, right atrial pressure, and PAH risk prediction scores should be included in allocation models. The panel reached strong agreement that an exception system is needed for high-risk PAH patients (mean agreement 4.18 ± 1.02) and that high-urgency systems are necessary when not using priority-based allocation (mean agreement 4.46 ± 0.57).

3.7 | Interventional and Bridging Strategies

There was strong agreement that ECMO can be deployed as bridge to transplant for refractory right heart failure (mean agreement 4.68 ± 0.48) and refractory hypoxemia (mean agreement 4.07 ± 0.98) (Figure 4). Additionally, there was agreement that early prophylactic cannulation is advantageous for hospitalized patients at risk of deterioration (mean agreement 2.93 ± 1.69). Panelists felt that cannulation strategy depended on patient-specific characteristics, with peripheral veno-arterial ECMO having highest agreement (mean agreement 4.09 ± 0.81). Other potential bridging strategies with agreement included pulmonary artery to left atrial cannulation, reverse Potts shunts in pediatric PAH patients, and long term continuous intravenous ionotropic support. Panelists met negative agreement against using a right atrial to pulmonary artery catheter for bridging. There was no agreement on approaches to vasodilators in those bridged to transplant on ECMO, or on the benefit of prophylactic bronchial collateral embolization.

3.8 | Transplant Surgery

The panel agreed on multiple aspects surrounding transplant surgery itself (Figure 4). There was agreement that parenteral

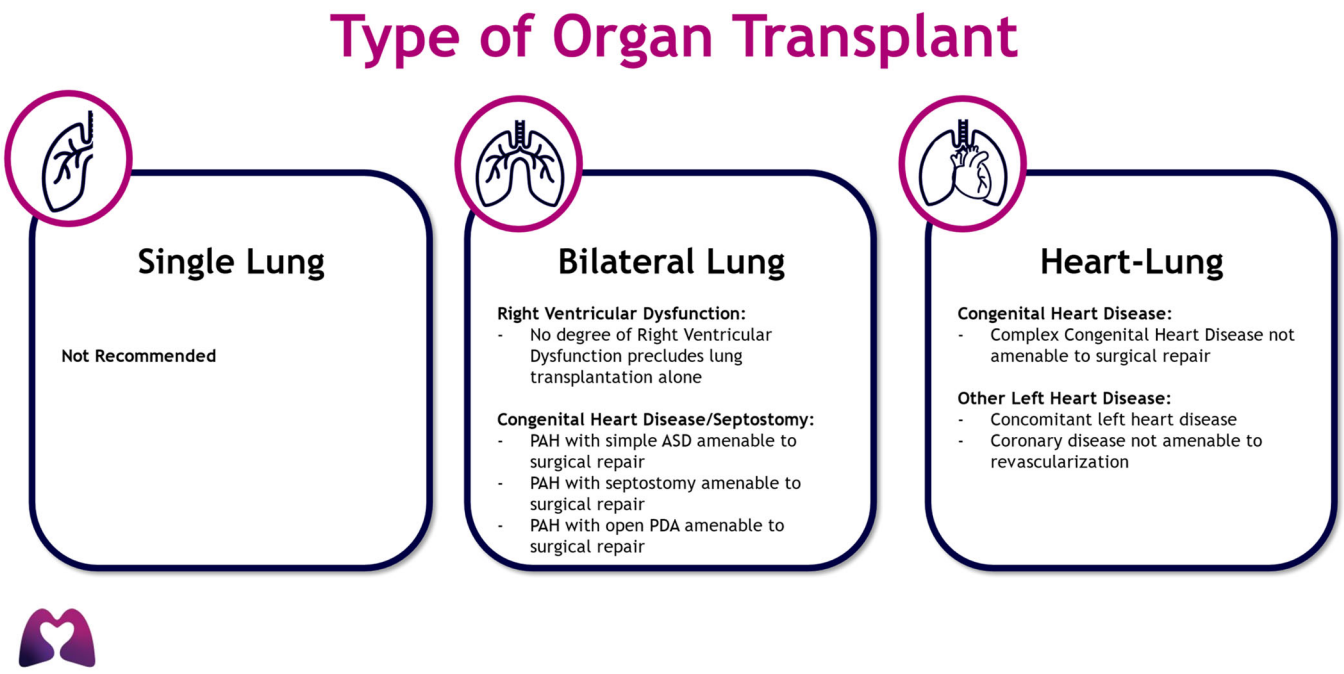

FIGURE 3 | Type of organ transplant recommended for patients with pulmonary arterial hypertension.

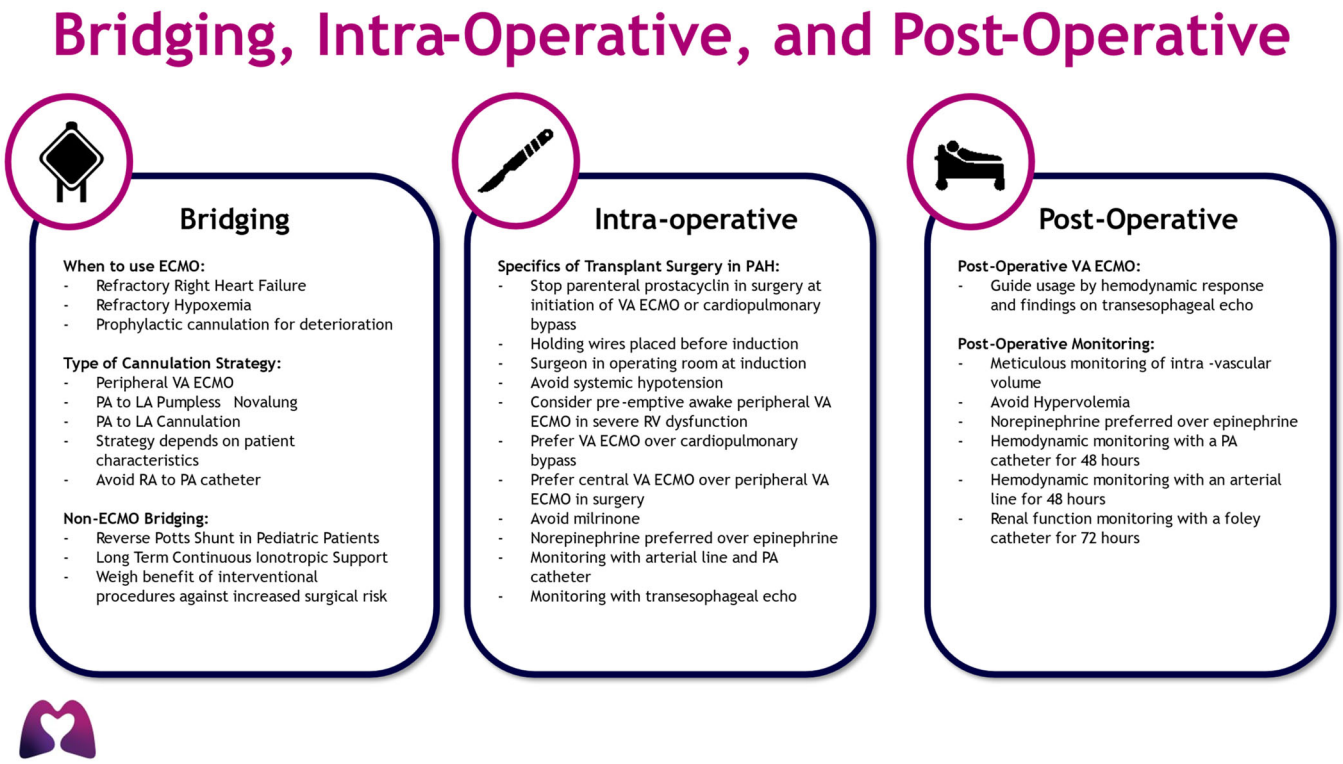

FIGURE 4 | Recommendations regarding bridging, intra-operative management, and postoperative management for patients with pulmonary arterial hypertension requiring lung transplantation.

prostacyclin analogs should be stopped intraoperatively after initiating either veno-arterial ECMO or cardiopulmonary bypass (mean agreement 3.87 ± 1.69) and a strong negative consensus against using parenteral prostacyclin analog therapy postoperatively (mean disagreement -4.00 ± 1.60). Regarding induction of anesthesia, there was agreement that holding wires for vascular cannulation should be placed in advance of induction, that surgeons should be in the room at induction, that hypotension should be avoided, and that pre-emptive awake veno-arterial ECMO should be considered in severe right ventricular dysfunction. Intraoperatively, there was agreement that central veno-arterial ECMO is preferred to cardiopulmonary bypass and peripheral ECMO strategies, that milrinone should be avoided, and that norepinephrine should be preferred. There was also agreement that patients should be monitored intraoperatively with an arterial line and pulmonary artery catheter (mean agreement 4.38 ± 0.74), as well as with continuous transesophageal echocardiography (mean agreement 3.35 ± 1.19).

3.9 | Postoperative Management

Although there is increasing use of pre-emptive veno-arterial ECMO in patients with PAH to minimize risks of primary graft dysfunction, the panel did not reach consensus on its routine use for postoperative recovery. Rather, the panel felt postoperative ECMO should be guided by hemodynamic response and transesophageal echocardiography findings (Figure 4). The panel reached consensus on multiple methods to monitor recovery postoperatively, including pulmonary artery catheters in the first 48 h (mean agreement 2.67 ± 1.53), arterial lines in the first 48 h (mean agreement 3.86 ± 1.24), and foley catheters in the first 72 h (mean agreement 3.36 ± 2.26). Norepinephrine was preferred by the panel over epinephrine in the early postoperative period (mean agreement 3.26 ± 1.24). The panel did not achieve any consensus on the postoperative use of nitric oxide, length of mechanical ventilation, length of sedation, or extubation strategies.

4 | Discussion

In this modified Delphi, a panel of experts developed consensus on multiple aspects of management in patients with PAH requiring lung transplantation.

Although the ISHLT, the ESC/ERS, and the World Symposium on Pulmonary Hypertension (WSPH) have all created guidelines on referral and listing, there is limited guidance on when PAH providers should start discussing transplantation [7, 15, 16, 25]. By comparison, other disease-specific societies, such as the Cystic Fibrosis Foundation, provide clear guidance on timing of discussions [26]. Our panelists found consensus for early discussions of lung transplantation in patients at significant risk of progression throughout their lifetime, either due to diagnosis or clinical profile. While discussing transplantation early in the course of disease may be uncomfortable, it has the benefit of demystifying transplantation. If adopted by PAH providers and recommended by professional societies, these early discussions may facilitate easier conversations when referral is warranted.

Our panelists agreed with existing referral and listing indications outlined in prior ISHLT, ESC/ERS, and WSPH guidelines

[7, 15, 16, 25]. Although these guidelines generally advocated for early referral, our study provides more granular parameters prompting referral. These PAH-specific parameters are associated with increased risk that may not be accounted for in existing risk assessment tools [27–29]. Although risk assessment is becoming more refined, the current tools are designed to assess risk of mortality, not post-transplant outcomes or survival to transplantation [15, 27]. Should risk assessment tools be refined to predict outcomes relevant in transplantation, they may better inform listing and referral practices. Better risk assessment relevant to transplantation for PAH may also inform allocation modeling; patients with PAH have inequitable access to transplantation in some allocation systems due to the models containing parameters not relevant to disease severity in PAH [30–34]. Our panelists also provided some guidance for policy makers as to which parameters may be better to include in future allocation models, including right ventricular dysfunction, cardiac index, stroke volume index, right atrial pressure, and PAH risk prediction scores. Our study also provides clearer guidance to the referring clinician. Notably, that patients should be referred despite perceived potential contraindications, and that patients denied at one center should be offered to seek evaluation at a second center. Transplant is evolving, and centers are becoming more skilled with complex cases. Patients should be afforded every opportunity to seek transplantation, especially considering that transplant is appropriate even in the setting of renal or hepatic impairment, or critical illness.

The 2024 WSPH proceedings listed bilateral lung transplantation as the method of choice in pulmonary hypertension over combined heart-lung transplantation, and the 2021 ISHLT Consensus Document on the Selection of Lung Transplant Candidates favored bilateral lung transplantation alone, even in the presence of right ventricular failure [7, 16, 25]. The 2022 ESC/ERS Guidelines commented that most patients receive bilateral lung transplantation alone, but remained agnostic in their recommendations [15]. Despite the fact that lung transplantation alone is sufficient for most patients, some centers continue to perform heart-lung transplantation at high rates [8]. Our findings further reinforce the ISHLT and WSPH statements, by showing in expert consensus that no degree of right ventricular dysfunction should preclude lung transplantation alone.

The most comprehensive document on bridging and surgical management in patients with PAH undergoing lung transplantation is the 2022 ISHLT Consensus Statement on the Perioperative Management of Patients with Pulmonary Hypertension and Right Heart Failure Undergoing Surgery [35]. Although this is a consensus statement, it is more of a scoping review, and was not developed via scientific consensus building approaches. Our consensus study adds to the literature by providing guidance based on consensus opinion regarding bridging and management in the intraoperative and postoperative periods. It is notable that we found agreement on the use of ECMO as a bridge to transplantation, and the appropriateness of evaluating awake patients while on ECMO. Patients bridged to transplant on ECMO can achieve excellent outcomes and have similar improvements in quality of life post-transplant as those who were not bridged to transplant on ECMO [36, 37]. Despite proving clear guidance on use of ECMO as a bridge to transplant, one nuance not addressed in our consensus was the patient who previously expressed willingness to consider transplantation or who was listed for transplantation but is now temporarily unable to consent due to critical illness or sedation. In this scenario, patients who meet criteria for transplantation may continue to be evaluated at the discretion of the transplant center, and the temporary critical illness should be taken into the context of their prior willingness to consider transplantation.

Although we found many areas of consensus, some statements found neither consensus agreement nor consensus disagreement. These represent areas warranting further research, to help determine best practices. Notable statements without consensus include how to approach Potts Shunts in adults, the utility of atrial septostomy, the utility of right ventricular assist devices, the utility of central ECMO in bridging, when to deploy bronchial artery collateral embolization, what to do with vasodilators in patients on ECMO, and how to use inhaled nitric oxide postoperatively. Another important area where we found lack of consensus is regarding the use of pre-emptive veno-arterial ECMO postoperatively, which is commonly used to help mitigate risk of primary graft dysfunction [38]. PAH is a notable risk factor for primary graft dysfunction, and primary graft dysfunction is associated with increased mortality and worse quality of life [39–41]. Pre-emptive use of veno-arterial ECMO postoperatively has the advantage of reduced reperfusion pressure as well as controlled loading of the left heart [42, 43]. That we did not find consensus for this practice highlights the need for further research to inform best practice for recovery after transplantation.

Our study has limitations. It is possible that there was bias in panelist selection. We mostly sampled providers from large volume transplant and PAH programs, so it is possible our recommendations do not reflect practice in smaller centers or resource-poor settings. Additionally, although we tried to be as broad as possible, we did limit our Delphi to PAH, so these recommendations do not apply to transplantation for other forms of pulmonary hypertension including pulmonary hypertension related to lung disease or chronic thromboembolic pulmonary hypertension.

Despite the limitations, our study has notable strengths. It is the only document to make consensus recommendations on lung transplantation for PAH with a rigorous approach to consensus building. Further, our panelists represent a variety of clinical specialists from three separate continents (North America, Europe, Australia).

Lung transplantation is a therapeutic option for patients with advanced PAH. It is highly successful and can both extend a patient's life and improve quality of life. The complexities of transplantation are unique for this disease state and require special attention. Our Delphi provides consensus recommendations on the care of these patients.

Author Contributions

Nicholas A. Kolaitis wrote the first draft of the manuscript, created the survey response forms, and compiled survey results. Nicholas A. Kolaitis, Hayley Barnes, Deborah J. Levine, Howard Castillo, Reda E. Girgis made substantial contributions to the conception and design of the work. These authors designed survey 2 based on feedback survey 1.

Nicholas A. Kolaitis, Hayley Barnes, Deborah J. Levine, Howard Castillo, Reda E. Girgis, Selim M. Arcasoy, Matthew Bacchetta, Luke Benvenuto, Erika Berman-Rosenzweig, Marisa Cevasco, Caitlin T. Demarest, Celine Dewachter, Michiel E. Erasmus, Allan R. Glanville, John Granton, Shaf Keshavjee,Vikramjit Khangoora, Sheila Krishnan, Olaf Mercier, Andrea N. Miltiades, David Montani, Edward Murphy, Ivan Robbins, Selim M. Arcasoy, Savale Laurent, Marc A. Simon, Jean-Luc Vachiery, Corey E. Ventetuolo, Helen M. Whitford made substantial contributions to the acquisition, analysis, or interpretation of data for the work. These authors participated in the survey rounds. They also revised the manuscript for important intellectual content. Franck F. Rahaghi made contributions to conception and design of the work.

Acknowledgments

The authors would like to thank the Pulmonary Vascular Research Institute for instituting the Innovative Drug Discovery Initiative Program. The authors received no specific funding for this work.

Ethics Statement

This study underwent IRB review and approval by the Monash University Institutional Review Board (Approval Number: 40697).

Conflicts of Interest

The authors declare no conflicts of interest.

References

1. K. Y. Chang, S. Duval, D. B. Badesch, et al., "Mortality in Pulmonary Arterial Hypertension in the Modern Era: Early Insights From the Pulmonary Hypertension Association Registry," *Journal of the American Heart Association* 11, no. 9 (2022): e024969.

2. N. Galiè, R. N. Channick, R. P. Frantz, et al., "Risk Stratification and Medical Therapy of Pulmonary Arterial Hypertension," *European Respiratory Journal* 53, no. 1 (2019): 1801889.

3. M. M. Hoeper, D. B. Badesch, H. A. Ghofrani, et al., "Phase 3 Trial of Sotatercept for Treatment of Pulmonary Arterial Hypertension," *New England Journal of Medicine* 388 (2023): 1478–1490.

4. V. V. McLaughlin, R. Channick, T. De Marco, et al., "Results of an Expert Consensus Survey on the Treatment of Pulmonary Arterial Hypertension With Oral Prostacyclin Pathway Agents," *Chest* 157, no. 4 (2020): 955–965.

5. G. Kovacs, S. Bartolome, C. P. Denton, et al., "Definition, Classification and Diagnosis of Pulmonary Hypertension," *European Respiratory Journal* 64, no. 4 (2024): 2401324.

6. O. Sitbon, R. Channick, K. M. Chin, et al., "Selexipag for the Treatment of Pulmonary Arterial Hypertension," *New England Journal of Medicine* 373, no. 26 (2015): 2522–2533.

7. L. E. Leard, A. M. Holm, M. Valapour, et al., "Consensus Document for the Selection of Lung Transplant Candidates: An Update From the International Society for Heart and Lung Transplantation," *Journal of Heart and Lung Transplantation* 40, no. 11 (2021): 1349–1379.

8. M. Valapour, C. J. Lehr, M. A. Skeans, et al., "OPTN/SRTR 2020 Annual Data Report: Lung," *American Journal of Transplantation* 22, no. S2 (2022): 438–518.

9. M. de Perrot, J. T. Granton, K. McRae, et al., "Outcome of Patients With Pulmonary Arterial Hypertension Referred for Lung Transplantation: A 14-year Single-Center Experience," *Journal of Thoracic and Cardiovascular Surgery* 143, no. 4 (2012): 910–918.

10. N. Kolaitis and J. Singer, "Defining Success in Lung Transplantation: From Survival to Quality of Life," *Seminars in Respiratory and Critical Care Medicine* 39, no. 2 (2018): 255–268.

11. J. P. Singer, P. P. Katz, A. Soong, et al., "Effect of Lung Transplantation on Health-Related Quality of Life in the Era of the Lung Allocation Score: A U.S. Prospective Cohort Study," *American Journal of Transplantation* 17, no. 5 (2017): 1334–1345.

12. L. G. Singer, N. A. Chowdhury, M. E. Faughnan, et al., "Effects of Recipient Age and Diagnosis on Health-Related Quality-of-Life Benefit of Lung Transplantation," *American Journal of Respiratory and Critical Care Medicine* 192, no. 8 (2015): 965–973.

13. H. S. Kulkarni, W. S. Cherikh, D. C. Chambers, et al., "Bronchiolitis Obliterans Syndrome-Free Survival After Lung Transplantation: An International Society for Heart and Lung Transplantation Thoracic Transplant Registry Analysis," *Journal of Heart and Lung Transplantation* 38, no. 1 (2019): 5–16.

14. N. A. Kolaitis, "Lung Transplantation for Pulmonary Arterial Hypertension," *Chest* 164, no. 4 (2023): 992–1006.

15. M. Humbert, G. Kovacs, M. M. Hoeper, et al., "2022 ESC/ERS Guidelines for the Diagnosis and Treatment of Pulmonary Hypertension," *European Respiratory Journal* 61, no. 1 (2023): 2200879.

16. M. M. Hoeper, R. L. Benza, P. Corris, et al., "Intensive Care, Right Ventricular Support and Lung Transplantation in Patients With Pulmonary Hypertension," *European Respiratory Journal* 53, no. 1 (2019): 1801906.

17. T. Fuehner, C. Kuehn, J. Hadem, et al., "Extracorporeal Membrane Oxygenation in Awake Patients as Bridge to Lung Transplantation," *American Journal of Respiratory and Critical Care Medicine* 185, no. 7 (2012): 763–768.

18. A. L. Delbecq, A. H. Van de Ven, and D. H. Gustafson, *Group Techniques for Program Planning: A Guide to Nominal Group and Delphi Processes* (Scott, Foresman, 1975).

19. F. F. Rahaghi, N. A. Kolaitis, A. Adegunsoye, et al., "Screening Strategies for Pulmonary Hypertension in Patients With Interstitial Lung Disease," *Chest* 162, no. 1 (2022): 145–155.

20. F. F. Rahaghi, V. P. Balasubramanian, R. C. Bourge, et al., "Delphi Consensus Recommendation for Optimization of Pulmonary Hypertension Therapy Focusing on Switching From a Phosphodiesterase 5 Inhibitor to Riociguat," *Pulmonary Circulation* 12, no. 2 (2022): e12055.

21. M. Wilson, R. H. Anguiano, R. L. A. Awdish, et al., "An Expert Panel Delphi Consensus Statement on the Use of Palliative Care in the Management of Patients With Pulmonary Arterial Hypertension," *Pulmonary Circulation* 12, no. 1 (2022): e12003.

22. F. F. Rahaghi, R. P. Allen, V. P. Balasubramanian, et al., "An Expert Panel Delphi Consensus Statement on Patient Selection and Management for Transitioning Between Oral and Inhaled Treprostinil," *Pulmonary Pharmacology & Therapeutics* 66 (2021): 101979.

23. F. F. Rahaghi, J. P. Feldman, R. P. Allen, et al., "Recommendations for the Use of Oral Treprostinil in Clinical Practice: A Delphi Consensus Project Pulmonary Circulation," *Pulmonary Circulation* 7, no. 1 (2017): 167–174.

24. O. Committee Establish Continuous Distribution of Lungs. 2023.

25. L. Savale, A. Benazzo, P. Corris, et al., "Transplantation, Bridging, and Support Technologies in Pulmonary Hypertension," *European Respiratory Journal* 64 (2024): 2401193.

26. K. J. Ramos, P. J. Smith, E. F. McKone, et al., "Lung Transplant Referral for Individuals With Cystic Fibrosis: Cystic Fibrosis Foundation Consensus Guidelines," *Journal of Cystic Fibrosis* 18, no. 3 (2019): 321–333.

27. R. L. Benza, M. K. Kanwar, A. Raina, et al., "Development and Validation of an Abridged Version of the REVEAL 2.0 Risk Score Calculator, REVEAL Lite 2, for Use in Patients With Pulmonary Arterial Hypertension," *Chest* 159, no. 1 (2021): 337–346.

28. R. L. Benza, D. P. Miller, A. Frost, R. J. Barst, A. M. Krichman, and M. D. McGoon, "Analysis of the Lung Allocation Score Estimation of Risk of Death in Patients With Pulmonary Arterial Hypertension Using Data From the REVEAL Registry," *TRANSPLANTATION* 90, no. 3 (2010): 298–305.

29. A. Boucly, A. Beurnier, S. Turquier, et al., "Risk Stratification Refinements With Inclusion of Haemodynamic Variables at Follow-Up in Patients With Pulmonary Arterial Hypertension," *European Respiratory Journal* 64, no. 3 (2024): 2400197.

30. H. Chen, S. C. Shiboski, J. A. Golden, et al., "Impact of the Lung Allocation Score on Lung Transplantation for Pulmonary Arterial Hypertension," *American Journal of Respiratory and Critical Care Medicine* 180, no. 5 (2009): 468–474.

31. K. M. Wille, L. B. Edwards, L. R. Callahan, A. R. McKoy, and K. M. Chan, "Characteristics of Lung Allocation Score Exception Requests Submitted to The National Lung Review Board," *Journal of Heart and Lung Transplantation* 36, no. 7 (2017): 812–814.

32. N. A. Kolaitis, H. Chen, D. R. Calabrese, et al., "The Lung Allocation Score Remains Inequitable for Patients With PAH, Even After the 2015 Revision," *American Journal of Respiratory and Critical Care Medicine* 207, no. 3 (February 2023): 300–311.

33. T. M. Egan and L. B. Edwards, "Effect of the Lung Allocation Score on Lung Transplantation in the United States," *Journal of Heart and Lung Transplantation* 35, no. 4 (2016): 433–439.

34. T. M. Egan, S. Murray, R. T. Bustami, et al., "Development of the New Lung Allocation System in the United States," *American Journal of Transplantation* 6, no. 5 Pt 2 (2006): 1212–1227.

35. D. P. McGlothlin, J. Granton, W. Klepetko, et al., "ISHLT Consensus Statement: Perioperative Management of Patients With Pulmonary Hypertension and Right Heart Failure Undergoing Surgery," *Journal of Heart and Lung Transplantation* 41, no. 9 (2022): 1135–1194.

36. J. Kukreja, S. Tsou, J. Chen, et al., "Risk Factors and Outcomes of Extracorporeal Membrane Oxygenation as a Bridge to Lung Transplantation," *Seminars in Thoracic and Cardiovascular Surgery* 32, no. 4 (2020): 772–785.

37. N. A. Kolaitis, A. Soong, P. Shrestha, et al., "Improvement in Patient-Reported Outcomes After Lung Transplantation Is not Impacted by the Use of Extracorporeal Membrane Oxygenation as a Bridge to Transplantation," *Journal of Thoracic and Cardiovascular Surgery* 156, no. 1 (2018): 440–448.e2.

38. M. Otto, D. McGiffin, H. Whitford, et al., "Survival and Left Ventricular Dysfunction Post Lung Transplantation for Pulmonary Arterial Hypertension," *Journal of Critical Care* 72 (2022): 154120.

39. J. M. Diamond, J. C. Lee, S. M. Kawut, et al., "Clinical Risk Factors for Primary Graft Dysfunction After Lung Transplantation," *American Journal of Respiratory and Critical Care Medicine* 187, no. 5 (2013): 527–534.

40. M. K. Porteous, J. C. Lee, D. J. Lederer, et al., "Clinical Risk Factors and Prognostic Model for Primary Graft Dysfunction After Lung Transplantation in Patients With Pulmonary Hypertension," *Annals of the American Thoracic Society* 14, no. 10 (2017): 1514–1522.

41. N. A. Kolaitis, Y. Gao, A. Soong, et al., "Primary Graft Dysfunction Attenuates Improvements in Health-Related Quality of Life After Lung Transplantation, but not Disability or Depression," *American Journal of Transplantation* 21, no. 2 (2021): 815–824.

42. A. Avriel, A. H. Klement, S. R. Johnson, M. de Perrot, and J. Granton, "Impact of Left Ventricular Diastolic Dysfunction on Lung Transplantation Outcome in Patients With Pulmonary Arterial Hypertension," *American Journal of Transplantation* 17, no. 10 (2017): 2705–2711.

43. S. Gupta, F. Torres, S. Bollineni, M. Mohanka, and V. Kaza, "Left Ventricular Dysfunction After Lung Transplantation for Pulmonary Arterial Hypertension," *Transplantation Proceedings* 47, no. 9 (2015): 2732–2736.

Supporting Information

Additional supporting information can be found online in the Supporting Information section.