CONSENSUS STATEMENT

International Society for Heart and Lung Transplantation Consensus Statement on the Referral and Selection of Pediatric Lung Transplant Candidates

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Children with advanced lung diseases are eligible to be considered as potential candidates for lung transplantation around the world. The timing of referral, evaluation, determination of candidacy, and listing of candidates poses challenges and ethical dilemmas for pediatric care providers. To address these challenges, the International Society for Heart and Lung Transplantation appointed an international panel of expert members to review the literature, to consider recent advances in the management of advanced pediatric lung diseases, and to generate the first consensus guidelines on the referral criteria and selection of pediatric lung transplant (LTx) candidates. This consensus document is meant to assist pediatric care providers throughout the world caring for children with advanced lung diseases to identify potential candidates for LTx, to optimize the timing of the referral of these patients to LTx centers, and to provide transplant centers with a framework for the evaluation and selection of children as candidates.

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KEYWORDS:
candidate; consensus; children; lung transplant; pediatrics; referral; selection

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1. BACKGROUND

Soon after the first lung transplant (LTx) was reported in a 15-year-old with familial pulmonary fibrosis by the University of Toronto in 1987,[1] pediatric LTx quickly grew as a field with the annual number of international transplants reaching 94 by 1995 and peaking at 136 in 2013.[2] Since the advent of CF transmembrane conductance regulator (CFTR) gene modulator therapy, the number of children with cystic fibrosis (CF) undergoing LTx has decreased, dramatically affecting the overall number of pediatric LTx.[2] Parallel to this decline in LTx performed for pediatric CF, there have been increasing numbers of LTx in children with interstitial lung disease (ILD) and pulmonary vascular disease (PVD).[3,4] Due to several changes occurring in the field of pediatric lung LTx, the referral and selection process has become more complex. Additionally, referral and selection in children require intimate knowledge of pediatric-specific conditions, disease progression in childhood, familial and caregiver factors, and a variety of other determinants that are unique in pediatrics compared to adults. These determinants are highlighted in this landmark document.

1.1. Goal of this consensus document

This document is intended to express a consensus of the membership of the International Society for Heart and Lung Transplantation (ISHLT) and to provide guidance for timely referral, assessment, optimization, and listing of children as potential LTx candidates. Previous ISHLT consensus documents on the selection of LTx candidates are primarily adult-focused and have provided limited pediatric-specific guidance on these key aspects.[5-8] As a result, the pediatric LTx community felt that for the first time, an independent document entirely focused on children was needed, which has been endorsed by leadership of the ISHLT.[9,10]

1.2. Use of this consensus document

These recommendations are intended to aid clinicians in the management of infants, children, and adolescents with advanced lung disease that is impacting survival and/or quality of life. Clinicians, patients, third-party payers, stakeholders, and courts should not view the recommendations contained in this consensus document as standards of care. Although evidence-based guidelines can summarize the best available evidence regarding the effects of an intervention in a given patient population, they cannot take into account all of the unique clinical circumstances that may arise when managing a patient. As such, their implementation is at the discretion of each treating physician who must work in concert with the patient and their family or caregivers through a process of shared decision-making. The application of these recommendations to patients must occur in the context of local center experience and expertise as well as national jurisdiction. If there is no experience or expertise locally, a transfer to a center with the appropriate expertise should be strongly considered.

2. METHODS

The consensus document was developed in accordance with the ISHLT Standards and Guidelines Committee document development policies. The Consensus Committee Members were selected to represent the diversity of the Society and were approved by the ISHLT Standards and Guidelines Committee. Each member was assigned to working groups in their area of expertise where they contributed to medical literature searches, development of content, voted on final consensus statements, and approved the final recommendations and manuscript. Meetings that occurred in the development of the consensus document were virtual and included sessions that involved the Consensus Committee or sessions that only included writing group members.

Medical literature searches were performed in mid-2024 and reviewed all pertinent articles. There was a final search for any updated literature before the submission of the consensus document for review. We focused the peer review on research since the inception of pediatric LTx. During review of the document, additional pertinent newly published articles were included, but a comprehensive review of literature was not repeated. A modified Delphi process was developed to reach consensus of the experts of the Consensus Committee Members. The Delphi process is a structured method for decision-making by the panel that was described by Delbecq et al[11] that is widely used in similar formats.

The Consensus Committee leadership developed the 3 scenarios used in the surveys, moderated the questionnaires, and managed the conduct of the study. Using a modified approach to the Delphi technique as

previously published, a total of 3 surveys were completed. Survey 1) included primarily open-ended questions about panelists' general approach to each section, allowing for text responses. This helped provide themes and content for subsequent surveys. Survey 2) consisted primarily of a series of statements based on panelists' responses to survey 1). The statements were developed by consolidating and clarifying the management options described in panelists' answers to the questions in survey 1). Panelists were asked to rate their agreement with each statement using a Likert scale ranging from −5 (*strongly* disagree) to +5 (*strongly agree*), with 0 indicating neutral sentiment. The opportunity to abstain from voting on a statement was provided throughout the process, in the event that a panelist felt they lacked the expertise to answer. Group discussion was led by their respective leaders, to allow thorough input, justification, and discussion from all voting members around answers provided in survey 2). Survey 3) was nearly identical to survey 2), except that panelists were provided with their own previous answers to survey 2), the mean and standard deviation of the group's previous answers, and whether consensus was achieved for each answer. For more complex statements, the opportunity for text-based comment was provided. All responses were anonymous, and only 1 adjudicator (N.A.) was provided access to a respondents' identifiable information, in the event that clarification was required around an answer, comment, or technical issue. Surveys were all administered via Qualtrics XM Survey Maker, a secure web-based application. Certain clarification to questions with ambiguity was provided, and the answers were discussed in group meetings. This additional information was intended to promote consensus by making participants aware of the overall group opinion; however, panelists were instructed throughout the process to vote according to personal sentiment and individual practice. The final aggregate results were circulated to all participants for review and comment. Full results for the final Survey 3 are provided in the Supporting Information Appendix.

Based on Likert scale from −5 (*strongly disagree*) to 5 (*strongly agree*), panelists were considered to have reached a consensus if the mean Likert scale score was ≤−2.5 (consensus against) or ≥+2.5 (consensus for), reflecting a 75% consensus level. Consensus was only reached if standard deviation (SD) did not cross 0, so as to avoid a bimodal distribution.

3. TIMING OF REFERRAL, EVALUATION, AND LISTING

3.1. Referral

Referring children for consideration of LTx is a complex and multifactorial process, which should ideally be initiated before there is an urgent need for transplantation. Panelists unanimously and strongly agreed that, as a general rule, pediatric LTx candidates should be referred early; the definition of "early" is dependent on the candidate's underlying diagnosis. This recommendation is consistent with the most recent ISHLT Consensus Document on LTx candidate referral and selection, stating that children should be referred before meeting criteria for active waitlisting.[8] This is especially necessary for children with advanced lung diseases given the following factors: (1) the complexity of the referral and listing processes in pediatrics, (2) the risk of more rapid decompensation in children compared to adults, (3) the fact that children and their families may need to travel long distances to be evaluated at a center capable of transplanting children, and (4) the challenges in acquiring suitable-sized organs, particularly for younger children, which may lead to longer wait times than for adults.[3,12-15] Early referral in this particularly vulnerable population aims to maximize the chances of successful transplant before decompensation or waitlist mortality. In addition, earlier referral establishes a working relationship with the care team and provides an opportunity for the patient and family to be introduced to the idea of and be educated around LTx, its requirements, and anticipated patient-specific outcomes. It is worth noting that timing of referral is also dependent on how effectively a jurisdiction's organ allocation system can provide access to deceased donor organs for children and the location-specific logistics of the referral process.

Panelists unanimously agreed that pediatric candidates for LTx should be referred if the candidate meets one of the following: (1) They meet disease-specific criteria, (2) there is a barrier that must be addressed to improve candidacy, (3) discussion around candidacy would affect therapeutic options such as bridging strategies, or (4) they require multiorgan transplantation (MOT) (Table 1). Disease-specific criteria for referral will be covered in subsequent sections.

A referral for pediatric LTx should comprise complete medical and surgical histories, including but not limited to primary diagnosis leading to transplant consideration, all other diagnoses, summary of patient's course and trajectory, hospitalizations, pulmonary function results, radiologic images and reports (including chest computed

Table 1 General Criteria for Referral of Children for Pediatric Lung Transplant

Referral for lung transplant in the pediatric candidate with advanced lung disease should occur if ONE of the 4 following criteria is met	1. Disease-specific criteria for referral are met.[a]
	2. There is a barrier that must be addressed to improve candidacy. These may include but are not limited to malnutrition, incomplete essential vaccinations, persistent poor adherence, poor psychosocial situation, or other modifiable risk factors that have been previously established.[8]
	3. Discussion around candidacy will affect or influence therapeutic options such as bridging strategies, which may include extracorporeal lung support (ECLS).
	4. There is a requirement for multiorgan transplantation due to multiorgan dysfunction.

[a]Refer to disease-specific criteria in the appropriate subsequent section (i.e., obstructive lung diseases, restrictive lung diseases, pulmonary vascular diseases [pulmonary hypertension], etc.).

tomography and echocardiography), history of surgeries and procedures (especially involving the chest or thorax), growth and development, vaccinations, medications, history of infection (namely respiratory cultures), social and family histories, and evaluations from allied health professionals. If available, cardiac catheterization reports or cardiac magnetic resonance imaging results should be included. Vaccination represents a major preventative measure against infectious disease risk factors in this population; therefore, history of primary vaccinations—particularly live vaccines—as advised by national or institutional vaccination guidelines is important to assess before transplantation.[8,16] This topic is covered in greater detail in a subsequent section on general pediatric considerations. A summary of suggested information that should be provided at the time of referral is shown in Table 2, which may vary based on institutional requirements.

REFERRAL: Pediatric lung transplant candidates should be referred early as a general rule. The definition of early referral depends on the underlying diagnosis.
Average response = +4.96
Range = +4 to +5
SD = 0.20

REFERRAL: Pediatric candidates for lung transplant should be referred: if they meet disease-specific criteria, if there is a barrier that must be addressed to improve candidacy, if discussion around candidacy would affect therapeutic options, or if they require multiorgan transplantation.
Average response = +4.96
Range = +4 to +5
SD = 0.20

3.2. Risk factors

Factors that place pediatric candidates at increased risk of poor outcomes are essentially the same as for adults, with 1 major difference being that all infants requiring LTx require intubation and mechanical ventilation. Many of the absolute contraindications, risk factors conferring substantially increased risk, and general risk factors previously suggested are applicable to the pediatric population.[8] However, panelists agreed that the following risk factors are unique or particularly important in children: caregiver unwillingness to pursue transplantation, unreliable support system or caregiving plan, persistent poor adherence despite interventions, uncontrolled aspiration into the lower airway, significant aortopulmonary collaterals, previous talc pleurodesis, a previous pulmonary-to-systemic arterial shunt ("reverse Potts shunt") via thoracotomy (posterolateral or lateral approach), and refusal of recommended vaccinations. Psychosocial risk factors are addressed in detail in a subsequent section. As with adults, multiple concurrent risk factors may increase the risk for adverse post-transplant outcomes. Referring the patient early and not at a time of urgency or duress allows for adequate time to address modifiable barriers to transplant if candidacy is declined. Factors leading to declination may include the above

Table 2 Information Required for Complete Referral for Pediatric Lung Transplant

Referring center	Referring medical professional and contact information, location/hospital, allied health professionals' contact if available (i.e., case manager, social worker)
Patient demographics	Name, date of birth, caregiver information, contact information for patient/caregiver
Medical and surgical history	Primary and secondary diagnoses, comorbidities
	Prior surgeries (including operative reports for thoracic surgeries; pleurodesis procedure and type), previous interventions for vascular access including any devices required
	Summary of patient's course, and trajectory of lung disease
	Functional status (exercise tolerance, ability to perform age-appropriate daily activities)
	Current respiratory support and last increases in needs, including history of oxygen therapy or ventilator support (CPAP/BiPAP/invasive ventilation), prior intubations
	Hospitalizations (reason for admission, length of stay, treatment course and outcomes)
	Growth history: growth charts including BMI or weight-for-length percentiles
	Developmental history: milestones achieved, potential developmental delays and assessments
	Vaccination status (complete list including all series, COVID-19 vaccines, and influenza vaccines)
	Medications: current and historic, including dose, frequency, and duration; documented response or lack of response to therapies used for advanced lung disease
	Allergies: medications, food, environmental
Investigations	Pulmonary function testing (current and historic) including spirometry, lung volumes, DLCO; MBW or infant PFT if relevant/available[17,18]; graph of trends if available
	6-minute walk test, if available, including trends
	Radiologic reports and images: CXR, chest CT, echocardiography, other imaging such as MRI or ultrasound if available
	If relevant: cardiac catheterization reports, cardiac MRI if available
	History of infections: including current and previous positive respiratory cultures (sputum and BAL if available) including nontuberculous mycobacteria and atypical organisms
	Genetic testing reports, including panel results
	Blood testing results, if available: blood type (ABO), HLA, and PRA results (if available), blood gas, CBC and differential, renal function tests, liver enzymes, liver function tests, and clotting factors
Social and family history	Caregiver supports, familial situation, family dynamics
	Detailed psychosocial history
	History of compliance or non-adherence, including effective strategies and supports used by the referring center to address any adherence concerns
	Smoking history, including patient or household exposures to tobacco, vaping, or cannabis use
	Family history of pulmonary disease, genetic disorders, or familiarity with transplantation
Allied health involvement	Dietician notes if available, including nutritional support (i.e., gastrostomy-tube), prior history of failure to thrive, special diets
	Neuropsychological evaluations if available; behavioral or psychologic conditions
	Social worker evaluation if available
	Palliative care discussions, advanced directives, or goals of care
	Additional consults which may include cardiology, gastroenterology, hematology, nephrology, neurology, oncology, etc.

Abbreviations: BAL, bronchoalveolar lavage; BMI, body mass index; BiPAP, bilevel positive airway pressure; CPAP, continuous positive airway pressure; CT, computed tomography; CXR, chest X-ray; DOB, date of birth; DLCO, diffusion capacity for carbon monoxide; HLA, human leukocyte antigen; MBW, multiple breath washout; MRI, magnetic resonance imaging; PFT, pulmonary function testing; PRA, panel reactive antibody.

risk factors, in addition to malnutrition, medical comorbidities, or incomplete age-appropriate vaccinations at some centers. Importantly, early referral allows time for potential candidates to be vaccinated as some vaccine series require multiple injections over time.

RISK FACTORS: Risk factors that place pediatric candidates at increased risk for poor outcomes are essentially the same as adults. Risk factors unique to pediatrics include caregiver unwillingness to pursue transplant, unreliable support system or caregiving plan, persistent poor adherence despite interventions, uncontrolled aspiration into lower airway, significant aortopulmonary collaterals, a previous pulmonary-to-systemic arterial shunt ("reverse Potts shunt") via thoracotomy (posterolateral or lateral approach), and refusal of essential vaccinations.
Average response = +4.38
Range = 0 to +5
SD = 1.17

3.3. Evaluation

Evaluation for pediatric LTx includes the assessment of lung disease and its severity, thoracic anatomy, surgical candidacy, degree of frailty, growth and nutritional status, presence and severity of comorbidities, psychosocial circumstances, and health-related behaviors that may impact short-term recovery and long-term survival. The term frailty, which may include malnutrition and/or sarcopenia, is used in this document with the caveat that it requires better definition in pediatric care as it relates to both physical and nutritional rehabilitation potential.[19] Panelists unanimously and strongly agreed that evaluation in children is essentially the same when compared to adults, with the exception of certain factors unique to pediatrics. These factors include evaluating family dynamics and support, developmental status, weight-for-length growth, and childhood vaccination. Developmental status as part of the pediatric evaluation is less relevant for candidacy at most pediatric institutions and is most relevant to allow for developmentally appropriate interactions, rapport building, and effective teaching around pretransplant and post-transplant processes. Age-appropriate considerations around testing are important considerations in this population. For instance, younger children, especially those under the age of 6 years, are unlikely to reliably complete reproducible pulmonary function testing, including spirometry and lung volume and diffusion capacity assessments. A 6-minute walk test may also be difficult to perform in younger children, and age-dependent normative values should be considered (including forced expiratory volume in 1 second (FEV_1) z-scores based on age and height). Interpretation of testing during a pediatric evaluation requires intimate knowledge of pediatric-specific criteria for acceptability and normative values; for example, for children aged 6 years or younger, an acceptable forced expiratory volume in 0.75 seconds ($FEV_{0.75}$) may be obtained from spirometry even with early termination.[20] Alternatively, lung function testing methods involving tidal breathing as multiple breath washout, can be considered for pediatric patients.[17]

Sedation may be required in some circumstances for accurate chest computed tomography imaging based on age, and testing for isohemagglutinins is unique to ABO incompatible LTx in infant candidates. Assessment of quality of life and neuropsychological testing should account for the age of the patient and level of development. Regarding timing of evaluation, whether completed early or urgently in the setting of a precipitous decline, every effort should be made to evaluate every potential candidate in a similar manner to other candidates. Decision around candidacy for LTx is an intricate process. Candidacy in a child should be informed by a thorough and unbiased evaluation process that includes confirmation of an accepted indication for LTx, absence of medical or surgical contraindications, and resolution of any modifiable risk factors associated with poor transplant outcomes.

EVALUATION: Evaluation for lung transplant in pediatrics is essentially the same when compared to adults. Factors that are unique to pediatrics include evaluating: family dynamics and supports, developmental status, weight-for-length, and vaccines. Age-appropriate considerations around testing are also important.
Average response = +4.61
Range = +3 to +5
SD = 0.66

3.4. Listing

Published criteria guiding timing of listing for LTx in children are still relevant.[8,21] However, there was strong agreement among the panelists that the previous recommendation, "Patients with CF < 18 years of age should be listed when FEV_1 is < 30% predicted" requires reevaluation in the era of highly effective CFTR modulators (HEMT),

Table 3 General Criteria for Listing of Children for Pediatric Lung Transplant
Source: Adapted from.[8]

Listing for lung transplant should be considered for children who meet ALL of the following general criteria:	1. Chronic, end-stage lung disease without surgical contraindication.[a]
	2. Progressive lung disease without available therapy or not responding to optimal treatment.
	3. Increased risk of death (> 50%) from primary lung disease within 2 y if lung transplantation is not performed.
	4. High likelihood (> 80%) of 5-year post-transplant survival from a general medical perspective, provided there is adequate graft function.

[a]Refer to disease-specific criteria in the appropriate subsequent section (i.e., obstructive lung diseases section for CF criteria, PVD section for pulmonary hypertension [PH], etc.).

including elexacaftor/tezacaftor/ivacaftor.[22,23] Timing of listing children with CF in the modern era is undoubtedly multi-faceted. This is further addressed in the Obstructive Lung Diseases section as part of disease-specific criteria for candidate listing. Additionally, with the changing demographics and shifting landscape of indications for pediatric LTx,[3,4] specific criteria for children with pulmonary hypertension (PH) and ILD need clarification. All panelists either agreed or strongly agreed that, in general, listing for LTx in pediatrics should be considered for chronic, end-stage lung disease if there is progressive lung disease without available therapy or not responding to optimal treatment and there is an increased chance of death (> 50% mortality) from the primary lung disease within 2 years (Table 3).[7,8] Moreover, the potential for improvement in quality of life for the patient was another criteria for listing consideration that was unanimously agreed upon by the panel. Compared to adults, timing of listing for transplant for children is different and must take into consideration the results of the full evaluation, including disease severity and trajectory, estimated wait time for suitable donor organs (i.e., candidate size and degree of allosensitization), waitlist survival time, frailty, and candidate/family's readiness for transplant.

LISTING: Published criteria guiding timing of listing for lung transplant in pediatrics is still relevant. However, the previous recommendation, "Patients with CF < 18 years of age should be listed when FEV1 < 30% predicted" needs to be reevaluated in the era of highly effective CFTR modulators. Some children with CF and FEV1 < 30% predicted may have life expectancy > 2 years, particularly when receiving these new agents. Timing of listing children with CF is multi-faceted. Also, with changing demographics in pediatric LTx, specific criteria for children with PH and ILD need to be clarified.
Average response = +4.24
Range = +1 to +5
SD = 1.18

LISTING: Listing for lung transplant in pediatrics should be considered for chronic, end-stage lung disease if there is progressive lung disease without available therapy or not responding to optimal treatment, and if there is an increased chance of death (> 50% mortality) from primary lung disease within 2 years.
Average response = +4.72
Range = +4 to +5
SD = 0.46

3.5. Further timing considerations

First, the panelists want to stress that the goal is not getting to LTx but being alive after with optimal quality of life. Moreover, there are modifiable factors that may exist before LTx that could impact outcomes after the transplant, so that is an additional consideration of care delivery during the pretransplant period. Optimization during the pretransplant period in pediatrics is often different than in adults. This involves ensuring patient and family goals are aligned and considering social and familial factors beyond the child's control. For children who are too early for full transplant evaluation or with contraindications for transplant that are potentially modifiable, specific parameters for the timing of rereferral and recommendations for ongoing optimization of candidacy should be provided.

OPTIMIZATION: Pretransplant optimization in pediatrics is often different than in adults. This involves ensuring patient and family goals are aligned and considering social factors beyond the child's control. Clear instructions are essential on goals for candidacy and timing for rereferral.
Average response = +4.75
Range = +2 to +5
SD = 0.68

Allocation policies should be carefully considered for a given region to help guide timing of referral, evaluation, and listing. Significant variation has been reported in lung allocation systems worldwide both in terms of prioritization and distribution for children.[24] Although most allocation systems worldwide aim to provide judicious LTx distribution for children, the definition of pediatrics varies from < 12 years of age to < 18 years of age, depending on the country and allocation organization. Therefore, in some jurisdictions compared to others, earlier listing may be appropriate due to the influence of donor allocation protocols on predicted waitlist time. The majority of panelists strongly agreed that LTx centers and referring providers should be alerted to pediatric policies through professional organizations, regional organ donation websites, and peer-to-peer education.

ALLOCATION: Lung allocation policies should be carefully considered for a given region to guide timing of referral, evaluation, and listing. Providers should be alerted to pediatric policies through professional organizations, regional organ donation websites, and peer-to-peer education.
Average response = +4.83
Range = +3 to +5
SD = 0.49

Evaluation and listing of children on life-sustaining respiratory support, including mechanical ventilation and/or extracorporeal life support (ECLS), may be considered at specialized centers depending on local expertise. The more common approach to ECLS in children requiring LTx is extracorporeal membrane oxygen (ECMO) with either a veno-venous (VV) or veno-arterial (VA) configuration via peripheral or central cannulation. In the setting of right ventricular (RV) failure, an ECLS approach may include paracorporeal lung assist device (PLAD) to further support the right ventricle before the transplant and optimize its recovery afterwards. For this document, we will use the terminology of ECLS as it may apply to any one of these configuration and cannulation strategies. Notably, there are challenges in bridging children with ECLS to LTx, which may impact the timing of referral and evaluation. Nearly all panelists strongly agreed that bridging to LTx with ECLS should only be considered at select centers, ideally with experience in pediatric LTx, pediatric acute care, and ECLS capabilities for children. An important caution regarding the use of ECLS as a bridge to LTx in children is that limited donor organ availability presents a challenge for the pediatric population; this is often the reason for optimizing both physical condition and nutritional status due to extended wait times. This contrasts with adults in whom adult donor organs are often more readily available.

ECLS BRIDGING: There are challenges in bridging children with ECLS to lung transplant, impacting timing of referral and evaluation. Bridging with ECLS should only be considered at select centers, ideally with experience in pediatric LTx, pediatric acute care, and ECLS capabilities for children.
Average response = +4.91
Range = +4 to +5
SD = 0.29

4. GENERAL PEDIATRIC CONSIDERATIONS

Because pediatric LTx is a complex therapy with significant morbidity and mortality, it is imperative to discuss some unique considerations that apply to children.

4.1. Growth and nutrition

Children being considered for LTx are more often underweight than overweight, with body mass index (BMI) frequently used as a surrogate in the assessment of their growth, nutritional status, and weight. Using BMI, underweight body habitus did not negatively impact survival after LTx in children while being overweight was associated with poorer survival.[25] However, more recently, a study using BMI percentage (BMI%) identified that children with a severely low BMI% (< third percentile) at listing had poorer allograft and overall survival, whereas the other BMI%, low-normal (3rd-85th percentile) and overweight-obese (≥85th percentile), did not influence mortality risk from the time of placement on the waitlist.[26] It remains unclear whether BMI, BMI%, or some other growth/nutrition/weight surrogate is optimal for assessing children with severe advanced lung disease being considered for LTx. Based on these limited data, the panelists determined that nutrition still has a significant impact on LTx outcomes, so aggressive nutritional optimization should occur pre and post listing.

GROWTH and NUTRITION: Nutrition has a significant impact on lung transplant outcomes, so aggressive nutritional optimization should occur pre and post listing.
Average response = +4.83
Range = +3 to +5
SD = 0.48

4.2. Unclear diagnosis

At times, pediatric patients may be too unstable to wait for the results of investigations done to determine the underlying diagnosis leading to referral for LTx; for example, a newborn with presumed fatal surfactant deficiency, who has genetic testing that is pending and is too unstable for a lung biopsy. In such cases, if no other contraindications are present, there was consensus amongst the panelists that such a patient could be evaluated and listed for LTx and could go on to be transplanted. Importantly though, panelists agreed that relevant genetic testing (where available) should be utilized in all cases before deeming the diagnosis unclear, and that all suspected therapeutic options should be exhausted before listing for LTx.

UNCLEAR DIAGNOSIS: Pediatric patients can be listed for lung transplantation with an unknown cause for their lung disease (e.g., infant with presumed fatal surfactant dysfunction but genetic testing pending and too unstable for lung biopsy) if no other contraindications.
Average response = +4.35
Range = +3 to +5
SD = 0.75

4.3. Vaccinations

Infection prevention is imperative for pediatric LTx recipients as infections in these patients lead to significant morbidity and mortality, with antimicrobial therapies often proving less effective than in immunocompetent individuals. The period before transplantation offers a critical opportunity to optimize immunity against vaccine-preventable diseases.[27] While vaccine responses may be reduced in some patients with advanced lung disease, they are typically even weaker post transplant when immunosuppression is in effect.[28] It is imperative that live virus vaccines (i.e., measles, mumps, rubella, and varicella vaccines) be given in the pretransplant period as they are generally avoided post LTx. The Infectious Diseases Society of America advises a minimum waiting period of 4 weeks between administering live virus vaccines and undergoing transplantation.[29] While newer data exist in nonlung pediatric solid organ transplantation (SOT) that live vaccines may safely and effectively be administered up to 2 weeks before transplant in patients with chronic liver disease for instance,[30] this has not yet translated to practice in pediatric LTx, especially given higher

immunosuppression requirements in LTx. Vaccinations of household contacts are also highly recommended. The panelists overall agreed that refusal of recommended pediatric vaccines is a contraindication for LTx, but 1 panelist disagreed somewhat with this statement.

VACCINATIONS: Refusal of recommended pediatric vaccinations is a contraindication for pediatric lung transplantation.
Average response = +3.60
Range = −2 to +5
SD = 1.83

4.4. Extracorporeal lung support

ECLS as a bridge to LTx in children should be used with the goal of stabilizing, enhancing their chances of receiving an LTx, and improving their opportunity of a successful post-transplant outcome.[31] Individual pediatric centers with extensive experience using ECLS as a bridge to LTx can even improve candidacy for children who are otherwise debilitated from their respiratory failure. Historically, ECLS as a bridge to transplantation was considered a relative contraindication in some centers for pediatric patients. However, recent evidence indicates, that when carefully selected—particularly in awake and extubated patients—ECLS may not negatively impact post-transplant outcomes, especially when performed in centers with significant expertise.[32-34]

A large analysis of pediatric Organ Procurement and Transplantation Network data on the use of ECLS at time of transplant assessed outcomes among 3 groups: bridged with ECMO support (with or without mechanical ventilation), mechanical ventilation, or neither. Although at hospital discharge patients bridged to LTx with ECMO had an associated increased risk of mortality, long-term survival of children receiving ECMO as a bridge to LTx was comparable to those who required neither ECMO nor ventilatory support.[35] At 5 years post transplant, both groups had an estimated survival of approximately 50%.

Based on the evidence, the consensus of the panel is that ECLS is not contraindicated for listed patients if they deteriorate (with no other contraindications, i.e., coagulopathy, progressive renal failure, etc.) and require bridging. Although ideally children bridging to transplant with ECLS should be fully evaluated and already listed candidates on the waiting list, the consensus of the panel was that children that are already on ECLS, whether as a rescue or for prolonged duration, should still be considered for LTx assessment and listing. ECLS can be used to optimize the patient before LTx because they are unstable or because they are in a poor state for transplant (e.g., intubated, heavily sedated or paralyzed, non mobile). ECLS should be used to rehabilitate the patient physically and mentally. The goal should be to extubate the patient, ambulate or maximize their physical therapy, significantly reduce or stop all sedation; nutritionally replenish them, and allow for social interactions and be purposeful in addressing their psychological state/wellness. The use of ECLS to simply limp a frail patient to transplant should be minimized since the outcomes for these patients are poor.

Infants and smaller children often face lengthy waiting times on the transplant list before a suitable donor organ becomes available. Infant LTx is exceedingly rare worldwide, with only 1 to 3 cases reported annually according to the ISHLT registry.[2] Due to the limited number of infant transplants each year, comprehensive studies on outcomes for various bridging techniques in this population are lacking.[36] However, studies on pediatric ECLS as a bridge to LTx provide valuable insights for transplant teams caring for neonates. In infants under 1 year, the primary respiratory indications for LTx are surfactant protein B deficiency and PH,[2] and risks of ECLS are most commonly hematologic in nature.[36,37] The consensus of the panel was that ECLS can be considered in some circumstances for infant LTx but should only be done at centers with experience in infant ECLS and infant LTx. Optimal physical and nutritional rehabilitation are a requirement for children on ECLS support awaiting LTx, as their wait time can be significantly longer than adults. Therefore, optimization of their candidacy is a necessity.

ECLS: ECLS is NOT contraindicated for listed patients if they deteriorate (with no other contraindications, e.g., coagulopathy, renal failure) and require bridging. Average response = +4.90 Range = +4 to +5 SD = 0.30
ECLS: Pediatric patients that are already on ECLS (rescued onto ECLS, or prolonged duration) should still be considered for lung transplant assessment and listing. Average response = +4.33 Range = +2 to +5 SD = 0.91
ECLS: ECLS can be considered in some circumstances for infant candidates but should only be done at centers with experience in infant ECLS and infant lung transplantation. Average response = +3.37 Range = –3 to +5 SD = 2.22

4.5. Additional factors to be considered

4.5.1. Age

For transplantation, the chronological age of 18 years old is the threshold of a child versus an adult. Although not universal, adult centers offering LTx for pediatric candidates may establish specific age limits.[8] For pediatric centers performing LTx, upper age limits exist due to the inability to provide comprehensive care to recipients, which is the reason to refer to an adult-focused center, or for administrative and/or legal reasons in certain health jurisdictions. Even if the primary team is comfortable with offering transplantation for individuals in an older age group over 18 years of age, other medical and surgical teams affiliated with the center must also be comfortable when asked to assist in the care of older patients. If comprehensive care is unavailable within a pediatric center due to age restrictions, referral to an adult center should strongly be considered. In contrast, evidence demonstrated that pre- and post-transplant outcomes for patients under 18 years of age are superior if LTx is performed at a pediatric-specific center,[38,39] so patients in this age category should ideally receive care at a pediatric center.

4.5.2. Functional status and frailty

Functional status is the ability of an individual to perform basic and instrumental activities of daily living with some degree of mobility, while frailty is a physiological state of heightened vulnerability to stressors that can forecast functional impairment and patient outcomes. Functional status is an established predictor of pre- and post-transplant outcomes in children.[40,41] Using Lansky Play-Performance Scores, a recent study showed that children with severe limited functional status at the time of LTx had significantly worse 1-year post-transplant outcomes.[42] In contrast, frailty in children with advanced lung disease has not been clearly defined to date according to the authors' knowledge despite it increasingly being studied in adults with advanced lung disease before and after LTx.[43-49] Whilst assessment tools for frailty are being developed, they are not currently accepted, so caution is warranted in using frailty for candidacy decisions, especially in the pediatric population. Importantly, pulmonary rehabilitation should be recommended for pediatric LTx candidates and recipients.

4.5.3. Gastroesophageal reflux/Gastrointestinal dysmotility

Gastroesophageal reflux (GER) and gastrointestinal dysmotility have variably been associated with adult lung allograft dysfunction, including acute cellular rejection, infection, and chronic lung allograft dysfunction (CLAD) with anti-reflux surgery linked to a reduction in the dysfunction.[50-56] The impact of GER and gastrointestinal dysmotility on lung allograft function in children is less robust due to fewer studies. It is known that high rates of GER occur in pediatric LTx recipients.[57] A single-center study found that GER burden nor fundoplication status impacted survival or rejection, while delayed gastric emptying appears significantly linked to CLAD development independent of GER.[58] Until further studies are performed to better delineate how GER and gastrointestinal dysmotility impacted allograft function in pediatric LTx recipients, centers caring for these children should follow

local management strategies in close collaboration with their gastroenterology, otolaryngology, and speech therapy colleagues depending on the cause of post-LTx dysphagia and aspiration, such as vocal cord paralysis, structural laryngeal abnormalities, and/or other etiologies.

4.5.4. Cardiac function

Cardiac diseases in children being considered for LTx are different than what commonly occurs in adults. Although some children with cardiac dysfunction undergo LTx, few data exist reporting outcomes in this patient population. In adult LTx recipients, elevated pretransplant left ventricular end-diastolic pressure and mean pulmonary capillary wedge pressure were associated with severe primary graft dysfunction (PGD).[59] Notably, a patient with left ventricular end-diastolic pressure and mean pulmonary capillary wedge pressure > 15 mm Hg pre LTx has a significantly increased risk of developing severe PGD.[59] These findings suggest that restrictive cardiac physiology and/or diastolic dysfunction impact the postoperative course after LTx. Therefore, a thorough assessment of cardiac function should be an integral part of the evaluation of a child being considered for LTx to assist in perioperative management, such as implementing ECLS as a preventive strategy for those at high risk for PGD or consideration of combined heart-lung transplant (HLT). Some of the participating panelists reported performing right and left cardiac catheterization at their center as a part of their comprehensive evaluation for LTx.

4.5.5. Renal function

Renal function in the perioperative period after LTx is important and can be negatively impacted by hypotension and hypoperfusion of the kidneys along with the initiation of nephrotoxic immunosuppression. Data in this area for children after LTx are rather limited. In adults, an estimated glomerular filtration rate (eGFR) < 60 ml/min/1.73 m^2 (Chronic Kidney Disease Epidemiology Collaboration equation) at the time of waitlisting and need for early dialysis after LTx are associated with worse outcomes.[60,61] In children after LTx, many patients experience renal dysfunction, particularly early post transplant; but unlike adults, predictors of renal impairment in this small pediatric population have not been identified.[62] Estimating eGFR using the modified Schwartz formula, an ISHLT Registry report on pediatric LTx recipient characteristics reported that an eGFR of 90 ml/min/1.73 m^2 was associated with a reduction in short-term survival at 1-year post transplant, whereas there was no difference in survival at 5 years post-transplant conditional on survival to 1 year.[2] A more recent ISHLT Registry Report identified that 5.6% of pediatric LTx recipients developed renal failure requiring dialysis during the first-year post transplant with associated risk factors being ECMO support, mechanical ventilation support, pretransplant dialysis, higher pulmonary capillary wedge pressures at time of transplant, and female donors.[63] The need for early dialysis during the first post-LTx year in children was associated with significantly worse outcome when compared to the nondialysis group and had a 50% mortality in less than 3 months post LTx (dialysis vs nondialysis patient survival: 71.8% vs 95.8% 30 day, 54.7% vs 94.6% 60 day, 38.7% vs 85.3% 1-year survival).[63] Therefore, a thorough assessment of renal function should be an integral part of the evaluation of a child being considered for LTx, especially in the setting of critical illness requiring mechanical respiratory or circulatory support. If the eGFR is below 90 ml/min/1.73 m^2, centers caring for these patients should follow local management strategies in close collaboration with their nephrology colleagues with the potential consideration of MOT with lung and kidney if the expertise exists.

4.5.6. Liver function

In certain patient populations, elevated bilirubin levels or the presence of liver disease may influence post-LTx outcomes or require MOT.[64,65] Due to limited data specific to children being considered for LTx, centers caring for these patients should follow local management strategies in close collaboration with their gastroenterology colleagues with the potential consideration of MOT with lung and liver if the expertise exists.

4.5.7. Genetic disorders/mutations

As genetic testing becomes more widely used, there will be a growing number of lung diseases associated with genetic disorders and genetic mutations in children. Historically, the genetic disorders most associated with LTx in children are CF, surfactant disorders and inherited pulmonary arterial hypertension or group 1 PH.[66,67] As more children with ILD undergo LTx,[3,4] it is likely an increasing number of individuals will have an associated genetic disorder or genetic mutation identified, so it will be imperative to delineate outcomes across these different genetic diseases while establishing best approaches to the management of this patient population as they undergo LTx.

4.5.8. Hematology/Oncology factors

Pediatric cancer survivors and bone marrow transplant (BMT)/hematopoietic stem cell transplant (HSCT) recipients are a growing patient population being referred for consideration for LTx with post-treatment bronchiolitis obliterans (BO) and pleuro-parenchymal fibroelastosis (PPFE).[68-70] Patients with a prior history of malignancy must undergo evaluation and testing to confirm there is no evidence of residual or metastatic disease. The determination of LTx candidacy should not necessarily be based on a cancer-free period but on the risk of recurrence or death as determined by a thorough evaluation by oncology and BMT/HSCT experts at the local center.

4.5.9. Allosensitization/Human leukocyte antigen mismatch

Pretransplant allosensitization to human leukocyte antigens (HLAs) increases waitlist time and risk for allograft dysfunction and mortality after LTx.[71] Moreover, increased donor-recipient HLA mismatching increases risk for acute cellular rejection, CLAD, and mortality in LTx recipients.[72-74] Implications for HLA mismatching appear to be significantly different between adults and children.[75] Despite the challenges that allosensitization to HLAs and the development of de novo donor specific anti-HLA antibodies (DSAs) pose, LTx can be successfully performed in children, even in the setting of a perioperative positive cross match.[76,77] Threshold antibody levels for donor organ acceptance and optimal detection for clinically significant DSAs have not been determined. Furthermore, there is wide variability in approaches to desensitization and treatment of DSAs with insufficient evidence to determine best practice. Due to a lack of consensus, there is no singular approach to managing allosensitized candidates on the waitlist or recipient with DSAs, and many centers utilize strategies that have historically worked best in their experience. For example, a single-center published successful LTx with good outcomes using repeated IgA- and IgM-enriched intravenous immunoglobulin infusions in combination with plasmapheresis before infusions and a single anti-CD20 antibody dose in adult and pediatric candidates with preformed DSAs.[78] The panel felt that multicenter studies for determining best practice in the management of allosensitization and HLA mismatch in children are urgently needed and would advance the field.

5. SURGICAL CONSIDERATIONS

5.1. Volume reduction surgery ("Trimming")

The practice of reducing the size of donor lungs to fit the shape and volume of the recipient thorax is now in its 4th decade,[79] and is an attractive option to widen the potential donor pool for pediatric candidates[80] or to address some of the issues that arise with abnormalities of chest wall shape. Techniques described include wedge resection ("segmentectomy"), lobectomy,[81] or a combination of these procedures.[82] High-volume centers with experience using these techniques in children and adolescents report similar outcomes to "classical" LTx.[81-84] Panelists agreed that lung volume reduction surgery should be offered at experienced centers, with the goal of widening the potential donor pool for pediatric lung candidates.

VOLUME REDUCTION SURGERY: Lung volume reduction surgery should be offered in experienced centers to widen the potential donor pool for pediatric candidates for lung transplantation.
Average response = +4.60
Range = +3 to +5
SD = 0.60

5.2. Previous thoracic procedures

Prior cardiothoracic procedures including pleurodesis, chest tube placement, thoracoscopy and thoracotomy, and median sternotomy should not be considered an absolute contraindication to LTx.[85] There was reasonable agreement amongst panelists, with some wide variation of the standard deviation. Additionally, panelists generally agreed that prior pneumonectomy is not an absolute contraindication to subsequent lung transplantation, if the resulting intrathoracic airway and vascular anatomy allow a successful anastomosis. Although previous talc pleurodesis was not considered a

contraindication by our panel, 1 surgical panelist felt this group was at much higher risk for complications and suggested caution. Detailed assessment on an individual basis, ideally with the records of previous procedures, should be conducted by the multidisciplinary team when assessing candidate suitability for listing for transplantation; this assessment should include consideration of factors that include intra- and perioperative risk.

PREVIOUS THORACIC PROCEDURES: Previous pleural procedures increase the time for dissection and risk of bleeding.
Average response = +3.55
Range = −3 to +5
SD = 1.99

PREVIOUS THORACIC PROCEDURES: Prior pneumonectomy is NOT an absolute contraindication to subsequent single- or double-lung transplantation if the resulting intrathoracic airway and vascular anatomy allow a subsequent successful anastomosis.
Average response = +4.20
Range = +2 to +5
SD = 0.77

5.3. Specific diagnoses for surgical planning

While rare in children, the relatively poor outcomes described in PPFE[86,87] suggest that this disease entity carries increased risk, and careful consideration regarding listing should be made on an individual basis. Similarly, reports on transplantation in the context of fibrosing mediastinitis are limited; centers should be confident that alternative medical or surgical options have been excluded and liaise closely with surgical teams. Similar to these pathologies, a patient having undergone successful talc pleurodesis can prove to be a significant challenge with an increased risk of bleeding and extended operative times. There was reasonable agreement that these specific conditions (such as PPFE and fibrosing mediastinitis) are not absolute contraindications and should be discussed on an individual basis. In addition, bleeding risk is increased for children with major aortopulmonary collateral arteries (MAPCAs).[88] There was strong agreement amongst panelists that MAPCAs present a relative contraindication for LTx (and are an absolute contraindication at some centers); as a result, patients, families, and caregivers should be counseled about an increased perioperative risk in these situations by a center that is willing to consider LTx for a child with MAPCAs.

SPECIFIC DIAGNOSES: Specific diagnoses that impact lung explantation, such as pleuro-parenchymal fibroelastosis and fibrosing mediastinitis, are NOT absolute contraindications and should be discussed on an individual basis.
Average response = +4.15
Range = 0 to +5
SD = 1.14

SPECIFIC DIAGNOSES: The presence of major aortopulmonary collateral arteries (MAPCAs) is a relative contraindication for lung transplantation, is associated with an increased risk of significant bleeding, and patients/families/caregivers should be counseled about increased perioperative risk.
Average response = +4.50
Range = +3 to +5
SD = 0.67

5.4. Infant lung transplantation

Infant LTx is well-established in experienced centers with comparable survival to older patients,[89] and as genetic testing increases for infants presenting with signs and symptoms consistent with ILD (particularly surfactant disorders), congenital PVD, and groups 1 and 3 PH, rates of referral for consideration of transplantation may increase in this group. There was strong agreement amongst panelists that infant lung transplantation is indeed feasible and should be performed at select experienced centers. Unlike groups undergoing LTx, intubation with or without a tracheostomy in infants is not a risk factor for poor outcomes.[90] Infants with severe bronchopulmonary dysplasia (BPD) who have

progressive respiratory failure despite maximal medical therapy are a particularly challenging group; potential for rehabilitation and consideration of comorbidities in other organ systems must be carefully considered for LTx in this group of infants. Moreover, infants with diagnoses of both BPD and severe tracheobronchomalacia present challenges after LTx, and therefore require consideration on an individual basis at select centers who also have experience in caring for advanced pediatric airway disorders. Limited reports to date describe acceptable respiratory outcomes in small numbers of infants and young children after LTx, with a high incidence of poor neurodevelopmental outcomes that may require additional assistance in early childhood.[89,91,92]

INFANT TRANSPLANTATION: Infant lung transplantation is feasible and should be performed in selected experienced centers.
Average response = +4.95
Range = +4 to +5
SD = 0.23

6. ECLS BRIDGING TO LTX OR ALTERNATIVE PROCEDURES

6.1. ECLS bridging

There are several ECLS strategies to consider when deciding on a means for mechanical support to bridge to LTx. In the setting of single-organ lung failure outside of PVD, VV ECMO support is often the preferred configuration due to the ability to use a single-site cannulation strategy which enhances an awaken patient to rehabilitate with ambulation if age appropriate. With VV ECMO, this configuration only improves gas exchange but does not allow for return of blood to the left heart, so if the underlying lung process continues to progress, the right ventricle may eventually fail, thus, necessitating VA ECMO or PLAD (pulmonary artery to left atrial circuit with pump and oxygenator) for those individual patients. Therefore, VA ECMO or PLAD in a configuration that will optimize rehabilitation seems the preferred approach in PVD or other refractory lung disease with associated RV failure. Sequential interrogations of the United Network for Organ Sharing (UNOS) database demonstrate that the use of ECLS bridging to LTx is increasing, with medium to longer outcomes that are not demonstrated to be inferior to those for children who did not require ECLS.[33,35] These studies do not capture data on those who go on to ECLS with the intention to transplant and do not survive to a suitable organ offer and are based on small numbers of cases (with most patients being older adolescents). It appears that ambulatory and rehabilitation strategies in highly experienced centers confer an advantage, in countries where waitlist times and offering schemes confer a reasonable probability of an organ offer within a feasible bridging period. ECLS, either peripheral ECMO or PLAD[93] should be considered in progressive hypoxic and/or hypercapnic respiratory failure not responding to mechanical ventilation. These higher risk evaluations ideally should occur at an experienced center and in children and young people who have previously been assessed for transplantation, with the recognition that in some disease entities (acute respiratory distress syndrome (ARDS), rapidly deteriorating neonate with chronic respiratory failure), this may not be feasible. Panelists strongly agreed that (1) ECLS should be considered as a bridging strategy for lung transplantation in the above situations, and (2) that referral for pediatric LTx should occur ideally before a patient requires ECLS (if in the setting of acute/chronic respiratory failure associated with preexisting advanced lung disease). There was consensus amongst the panelists that providers should consider referral to a pediatric LTx program if a child with refractory acute respiratory failure has remained on ECMO for > 2 weeks without any signs of improvement. This is based on previous publications in children who survive ECMO support for ARDS, with a mean length of support being 10 ± 8 days.[94-97] Factors that should be considered before ECLS use include: vascular access, bleeding risk, neurological concerns, infectious concerns, use of anticoagulation, need for paralysis or sedation (ideally minimal), participation in physiotherapy (PT) / occupational therapy (OT), nutrition (ideally predominantly enteral), psychosocial supports, waiting period, and potential allosensitization. There was strong agreement amongst panelists that at experienced pediatric ECLS centers, the implementation of ambulatory and rehabilitation strategies should be considered for pediatric patients on ECLS, with a focus on individualized care and close collaboration between transplant, ECLS, and rehabilitation teams. Risks inherent in ECLS may result in a subsequent change in suitability for LTx candidacy, particularly neurological complications and systemic infection,[93] and where possible patients and families should be carefully

counseled about this before embarking on ECLS. Counseling around potential major consequences should include discussing: bleeding, vascular injury, threatened limbs, thromboembolic events, infection, neurological complications, and the need for extended rehabilitation. The uncertain waiting period, outcomes, and potential withdrawal from ECLS without transplant should also be discussed. Finally, there was unanimous strong agreement that criteria requiring removal from transplant listing should be communicated before ECLS initiation; these may include severe neurologic deficit, bleeding disorders, uncontrolled infection, high degree of allosensitization, or multiorgan failure.

ECLS: ECLS should be considered as a bridging strategy in select pediatric patients with acute/chronic respiratory failure associated with advanced lung disease. Average response = +4.95 Range = +4 to +5 SD = 0.22
ECLS: Referral for pediatric lung transplant should occur ideally before a patient requires ECLS, if in the setting of acute/chronic respiratory failure associated with preexisting advanced lung disease. Providers should refer to a pediatric lung transplant program if a child with refractory acute respiratory failure has remained on ECMO for >2 weeks without any signs of improvement. Average response = +4.45 Range = +2 to +5 SD = 0.89
ECLS: Before ECLS, these factors should be considered: access, bleeding risk, neurological concerns, infectious concerns, use of anticoagulation, need for paralysis / sedation (ideally minimal), participation in rehabilitation with physical and occupational therapy, nutrition (ideally predominantly enteral), psychosocial supports, waiting period, and allosensitization. Average response = +4.95 Range = +4 to +5 SD = 0.22
ECLS: In experienced pediatric ECLS centers, the implementation of ambulatory and rehabilitation strategies should be considered for pediatric patients on ECLS, with a focus on individualized care and close collaboration between transplant, ECLS, and rehab teams. Average response = +4.86 Range = +4 to +5 SD = 0.35
ECLS: Families of those undergoing ECLS should be counseled about potential major consequences: including bleeding, vascular injury, threatened limbs, thromboembolic events, infection, neurological complications, and the need for extended rehabilitation. The uncertain waiting period, outcomes, and potential withdrawal from ECLS without transplant should be discussed. Average response = +5.00 Range = +5 to +5 SD = 0.00
ECLS: Criteria requiring removal from transplant listing should be communicated before ECLS initiation. This includes severe neurologic deficit, bleeding disorders, uncontrolled infection, high degree of allosensitization, or multiorgan failure. Average response = +5.00 Range = +5 to +5 SD = 0.00

6.2. Alternative procedures to ECLS

In selected cases of severe pediatric PH where waiting on donor lungs is not conducive for children who are critically ill where ECLS is employed, a pulmonary-to-systemic arterial shunt ("reverse Potts shunt") may provide an alternate treatment strategy with outcomes comparable to LTx.[98,99] Transcatheter approaches to pulmonary-to-systemic arterial shunt are described.[100] Acute preoperative decompensation and substantial RV dysfunction are associated with higher mortality,[99] emphasizing the importance of careful case selection. A recent clinical practice guideline recommended LTx and not the creation of a pulmonary-to-systemic arterial shunt in children with progressive severe PH who were on ECLS without reversible cause due to significant mortality differences as discovered by their meta-analysis of the current published literature.[101] Indeed, there was excellent consensus amongst panelists that alternative procedures, such as a pulmonary-to-systemic arterial shunt, along with atrial septal intervention (creation or enlargement) and pulmonary artery-to-left atrium shunt with artificial membrane, should be considered as palliation or a means to transplant based on each patient's hemodynamic condition and

center-specific experience. If a surgical pulmonary-to-systemic arterial shunt is performed and LTx to be considered in the future, the panelists felt an anterior approach via a sternotomy with patch is the preferred approach.[101,102] Importantly, we provide more detailed discussion on alternative strategies for children with severe pediatric PH and the implications for consideration of LTx in the PVD section of the consensus document.

ALTERNATIVE PROCEDURES TO ECLS: Alternative procedures such as a pulmonary-to-systemic arterial shunt ("reverse Potts shunt"), atrial septal intervention (creation or enlargement), and pulmonary artery-to-left atrium shunt with artificial membrane (Novalung, Quadrox) should be considered as a palliation or a means to transplant based on each patient's hemodynamic condition and center's experience.
Average response = +4.95
Range = +4 to +5
SD = 0.22

6.3. Infectious disease factors

Data regarding infectious disease considerations in the candidacy for pediatric LTx are extremely limited, and most opinion is based on small retrospective studies or extrapolated by data from adult LTx. Specific considerations focus on history of infections with nontuberculosis mycobacteria, Aspergillus and other molds, and multidrug-resistant (MDR) bacterial pathogens such as *Burkholderia cenocepacia*, *Pseudomonas aeruginosa*, *Acinetobacter* species, and *Achromobacter* species.

6.4. Nontuberculosis mycobacteria

Information on the impact of nontuberculosis mycobacteria on pediatric LTx is limited; however, case series relate the experience of specific to *Mycobacterium abscessus* complex species have been described.[103-105] Variability in pretransplant management and listing circumstances include differences in documented clearance of sputum smears and cultures as well as differences in antimicrobial susceptibilities, in the small cases series ranging from 2 to 16 pediatric LTx recipients. Further, the range of outcomes reported extend from early mortality to development of bronchiolitis obliterans syndrome (BOS), specifically for *M abscessus* subspecies *abscessus,* while *M abscessus* subspecies *bolletii* and *massiliense* more frequently report better outcomes.[105] Based on this limited data, the panelists determined that NTM other than *M abscessus* complex species was not a contraindication to pediatric LTx but were less definitive about how *M abscessus* complex species should be considered. The panelists favored using both smear and culture negativity with the availability of an appropriate antimicrobial plan as markers for candidacy. However, a few panelists pointed out that listing for LTx could be considered even in a candidate with culture positivity but smear negativity. One panelist suggested ensuring that lower respiratory tract cultures are used.

NTM: NTM other than M. abscessus are NOT a contraindication to pediatric lung transplantation.
Average response = +4.52
Range = +3 to +5
SD = 0.62

NTM: Transplant in a patient with history of M. abscessus can be considered, but only after documented clearance (negative smear, negative culture).
Average response = +3.18
Range = −1 to +5
SD = 1.51

NTM: Transplant in a patient with history of M. abscessus can be considered, but only if there is available antimicrobial therapy.
Average response = +3.23
Range = −4 to +5
SD = 2.22

6.5. Aspergillus and other molds

Aspergillus and other invasive fungal infections have been associated with increased morbidity and early mortality in pediatric LTx recipients with significant variability in prevention strategies.[106-108] While CF is a risk factor for pretransplant colonization, age, CF, and pretransplant colonization were not associated with post-transplant detection of fungal organisms or pulmonary fungal infection.[107] Due to a paucity of information regarding the impact of resistant Aspergillus species, Scedosporium/Lomentospora, and other molds, the panelists reached consensus around these organisms being an absolute contraindication for LTx in the setting of uncontrolled fungal infection. Although the remaining issues for Aspergillus species, Scedosporium/Lomentospora, and other molds examined by the panel did not achieve consensus, there was a trend that available therapeutic options were needed to consider a child for LTx. Due to the lack of research focused on Aspergillus species, Scedosporium/Lomentospora, and other molds in LTx, we identified a key area needing future exploration to help guide the management of patients with these pathogens. Based a lack of consensus on these findings LTx could be considered on an individual basis by centers who have more experience with resistant fungal species, especially where the infection can be controlled with the availability of therapeutic options.

ASPERGILLUS / MOLDS: Uncontrolled fungal infection is a contraindication to pediatric lung transplantation. Average response = +4.75 Range = +3 to +5 SD = 0.55
ASPERGILLUS / MOLDS: History of Aspergillus, Scedosporium/Lomentospora, and other molds without available therapeutic options are an absolute contraindication to pediatric lung transplantation. Average response = +2.83 Range = −5 to +5 SD = 2.55
ASPERGILLUS / MOLDS: History of Aspergillus, Scedosporium/Lomentospora, and other molds without available therapeutic options are a relative contraindication to pediatric lung transplantation. Average response = +2.32 Range = −5 to +5 SD = 3.07
ASPERGILLUS / MOLDS: Scedosporium/Lomentospora is a relative contraindication to pediatric lung transplantation. Average response = +3.06 Range = −1 to +5 SD = 1.78

6.6. Resistant gram-negative bacteria

Presence of MDR gram-negative bacteria in patients specifically with an underlying diagnosis of CF has been reported to have limited impact on mortality after adult LTx in some studies,[109-111] while another 2-center study reported decreased long-term survival evident starting around 1-year post transplant in a period before the availability of broader extended-spectrum beta-lactamases.[112,113] However, infection-related morbidity at 90-days and 1-year post transplant were higher in LTx recipients labeled as pan-resistant in the ISHLT Thoracic Transplant (TTX) Registry. In pediatric LTx candidates with CF, ISHLT TTX Registry data indicated that presence of pan-resistant infections increased over time but was not associated with decreased graft survival.[114] The panelists agreed that MDR or pan-resistant gram-negative organisms should not be a contraindication to transplant listing while consideration for available therapy and active infection was strongly favored as part of decision-making.

For *Burkholderia cenocepacia*, the panel did not find consensus for this organism as either a relative or absolute contraindication. Overall, *Burkholderia cenocepacia* has been associated with nonreferral for LTx in individuals with CF in the United States,[115] and has been associated with decreased post-transplant survival.[116-119] A small, single-center study in the United Kingdom reported a 1-year survival of 25% (3/12) for CF patients with *Burkholderia cenocepacia*,[116-119] while a more recent, single-center study of 87 CF patients with *Burkholderia cenocepacia* reported 1-, 5-, and 10-year survival rates of 59%, 33%, and 16%.[116-119] A few panelists suggested that pediatric candidates with *Burkholderia cenocepacia* could be considered and waitlisted at experienced transplant centers on an individual basis. Studies report that noncenocepacia *Burkholderia* species, such as *Burkholderia gladioli*, *Burkholderia multivorans*, *Burkholderia*

vietnamiensis, among others, have less impact on mortality[120]; therefore pediatric candidates with these less virulent strains of *Burkholderia* should be considered on an individual basis based on the experience of the center and the antimicrobial profile of the *Burkholderia* species.

MDR PSEUDOMONAS / ACINETOBACTER: Multidrug or pan-resistant gram-negative bacteria are NOT a contraindication pediatric lung transplantation. Average response = +3.29 Range = −1 to +5 SD = 1.61
MDR PSEUDOMONAS / ACINETOBACTER: Factors affecting consideration of listing a patient with MDR gram-negative bacteria should include: active infection, and available antimicrobial therapy. Average response = +4.65 Range = +3 to +5 SD = 0.61
BURKHOLDERIA: *Burkholderia cenocepacia* is an absolute contraindication to pediatric lung transplantation. Average response = +1.47 Range = −4 to +5 SD = 3.03
BURKHOLDERIA: *Burkholderia cenocepacia* is a relative contraindication to pediatric lung transplantation, which should be managed by experienced centers. Average response = +1.84 Range = −5 to +5 SD = 2.95

7. PSYCHOSOCIAL FACTORS

7.1. Psychosocial screening

Pretransplant psychosocial factors that are typically considered to determine transplant eligibility include: past and present adherence to medical regimens, mental health, substance use, and social supports.[121,122] Whilst the general psychosocial considerations for the potential pediatric LTx candidate are similar to those of adult recipients,[8,121] the increased dependence of children on their family or caregiver means that pretransplant screening must take into account not only the individual child, their developmental and healthcare needs, but also the socioenvironmental context in which they live, including the dynamics of the family/caregiving unit as well as the caregiver's functional and emotional capabilities to manage the child's healthcare needs.

Despite recognition of the importance of psychosocial function for a favorable LTx outcome, there are few absolute psychosocial contraindications. In fact, the aim of the psychosocial evaluation in pediatrics is not to exclude pediatric patients from the opportunity of transplantation, but to understand the challenges the patient and family/caring unit might encounter on their transplant journey and to guide the transplant team on how best to support the patient and their family/caring unit to optimize their candidacy and post-transplant outcomes. Careful consideration should be given to supporting children undergoing a psychosocial evaluation, including an awareness of their ill health and what physical and psychological support they might need during the pretransplant assessment.

Despite their young age, children undergoing transplant assessment should be involved in discussions about plans for their future care in a developmentally appropriate manner. The pediatric patient's knowledge, understanding and capacity to engage in decision-making regarding their health should be assessed. Factors that are specific to the patient's personal, social and environmental resources and circumstances should be understood to elicit information regarding support needs. It is important to consider the patient, in this case a child or adolescent, in their developmental, social and environmental context. This is particularly important in terms of their understanding of their healthcare needs, and the need to tailor developmentally appropriate information.

Many of the complexities of post-transplant care are similar in children compared to adults; irrespective of age, these include lifelong immunosuppression, frequent clinic visits, and numerous blood draws amongst other surveillance procedures.[123] One main difference between pediatric and adult candidates is the role of caregivers in monitoring and supervising the post-transplant regimen. As such, psychosocial evaluation should also include the application of these

domains to the family/caring unit, to assess their ability to adequately support the child or adolescent. Particular attention should be given to the family/caring unit's understanding of their child's health needs, and any challenges they might experience in meeting those needs. Where the family/caring unit has responsibility for the decision-making, for example due to a child's age or developmental understanding of health, it is important to consider the health beliefs of the family/caring unit, including any specific religious or cultural influences.

7.2. Adolescence and early adulthood development

Adolescence and young adulthood is a distinct developmental period characterized by the transition from childhood to independent adulthood.[124] Like other young people after SOT,[125-128] adolescent LTx recipients demonstrate poorer outcomes with increased mortality compared to younger children and adults.[129] As such, factors that can influence outcomes in this age group should be specifically and carefully assessed.

The assessment of developmental stage and the capacity of the young person to become independent is crucial to allow for planning of pre- and post-transplant education as well as planning for transition to adult care. Evidence of a social support system, including supportive caregivers and the maintenance of peer relationships is protective for non-adherence.[130] Substance use and misuse is common in young people and the assessment of extent, degree and modifiable nature of substance use is essential. Non-adherence rates are thought to be highest in adolescents compared to all other age groups,[131,132] with evidence of this seen in many chronic diseases.[133] Assessment of non-adherence and implementation of strategies to improve adherence to improve candidacy are an essential part of the assessment process.

Adolescents should be assessed both in the presence of their caregivers and alone. Assessment should be developmentally appropriate and consider the psychosocial context of the young person's illness. The cornerstone of assessment is the psychosocial interview which allows for a comprehensive understanding of the young person's situation and specific needs.[134] While various psychosocial frameworks exist that can be useful to provide a scaffold around which to direct questions,[135] the important domains to review include: home life, participation in education and employment, activities enjoyed and participation, substance use and other safety issues.

Preexisting psychological issues can impact post-transplant outcomes with reported increase in non-adherence and hospitalizations.[136,137] Adolescents should therefore be screened for past and current mood disorders (including suicidal ideation or other self-injurious behavior), anxiety disorders and other mental health problems (i.e., eating disorders, psychosis, and personality disorders).

Part of the assessment should also focus on a child and young person's capacity to consent. The legal rights of a young person to consent to or refuse treatment are dependent on age and jurisdiction and vary widely internationally. Despite this, the capacity to consent should be assessed in all adolescents. Even where no legal requirement for consent exists, adolescents and young people should be supported to feel part of the decision-making around transplant and assent to the life-long commitment to medical care that it requires.

7.3. Psychosocial contraindications to transplantation

Strong consensus was achieved among the panelists that unmodifiable non-adherence, substance abuse (including smoking and vaping), and insufficient caregiver support are contraindications to pediatric LTx. These contraindications are similar to those suggested for adults,[8,121] and are also deemed critical by pulmonologists,[138,139] with evidence demonstrating a relationship between these issues and adverse outcomes in both pediatric and adult SOT recipients.[121,122]

7.4. Non-adherence

While few studies have focused on adherence to medication in pediatric LTx populations specifically,[130,131,140] data from systematic reviews in other pediatric SOT groups suggest the prevalence of non-adherence in pediatric LTx cohorts is expected to be as high as 30%.[141-143] Non-adherence to immunosuppressive regimens is a known risk factor for late acute rejection, graft loss, and mortality in other transplant populations (both adult and pediatric)[143-145]; therefore, it is reasonable to infer that persistent medication non-adherence most likely increases the risk for antibody mediated rejection, CLAD, and possibly even late mortality in pediatric lung transplantation.

Past behavior is a strong predictor of future behavior,[146,147] and non-adherence should be assessed and addressed as early as possible in the patient's disease trajectory,[12,123] preferably before individuals are referred for transplant evaluation.[12,123] Given that adherence can fluctuate over time and can be influenced by the child's developmental stage,[146] regular assessment during the entire transplant pathway is warranted.[8] Rather than to prevent referral for consideration of or acceptance for transplant, the identification of non-adherence provides the opportunity to implement interventions to improve adherence. Pediatric patients should not be waitlisted if they are not engaging in the support being offered or if they continue to have serious problems with medication adherence without signs of improvement despite intervention.

7.5. Lack of sufficient care providers to support transplantation

Unlike adult candidates, pediatric recipients cannot manage their care independently. Therefore, the availability of caregivers who can provide the necessary support pre- and post transplant is essential. In the appropriate setting and jurisdiction, elective medical foster care for pediatric candidates who do not have the support system for transplantation is a consideration. Although there are no published standards on the number of required caregivers for a child who undergoes, the accepted minimum at most individual centers is 2 care providers; however, this may vary based on candidate age and social situation. Unstable or unsupportive family environments, poor family communication and conflict, history of child maltreatment, and high parental distress are risk factors for post-transplant non-adherence and adverse clinical outcomes, including late acute rejection and unplanned hospitalization.[130-132,148,149] The identification of these issues provides an opportunity for intervention. If no improvements in family functioning can be realized by supportive interventions, and if no alternative caregivers can be identified, a lack of capable caregivers should be considered a contraindication to transplantation, either permanently or temporarily. Panelists achieved consensus and generally agreed that unmodifiable non-adherence and the lack of sufficient care providers to support the patient after LTx are contraindications in children.

PSYCHOSOCIAL: Unmodifiable non-adherence and lack of sufficient care providers to support transplantation are contraindications to lung transplant.
Average response = +4.75
Range = +2 to +5
SD = 0.74

7.6. Substance misuse

As transplant candidates may be reluctant to disclose substance use and misuse, objective measurement methods to detect substance use (nicotine, vaping, cannabis, opioid, etc.) should be used in addition to self-report. In some jurisdictions, diagnostic testing for substance use in children may require consent before it can be performed.

7.6.1. Nicotine

A scoping review on substance use in adolescents and young SOT recipients up to age 40 revealed that evidence on this topic is scarce (especially on illicit drug use), but that its prevalence is relatively similar to numbers observed in healthy peers, and that substance use might result in non-adherence and poor clinical outcomes.[150] More specifically, smoking is a preventable cause of death both in the general and transplant population, and it increases the risk for early graft dysfunction, infection, malignancy and ischemic heart diseases.[151] Various smoking cessation programs exist that show success in helping people, including young adults, to quit tobacco use successfully.[152] Regular screening pre- and post transplant, for instance by means of cotinine testing or exhaled carbon monoxide (CO) measures, is indicated to evaluate for sustained smoking cessation.

7.6.2. Vaping

Particular attention should also be given to vaping or e-cigarette use in adolescent transplant candidates. The use of vaping products is particularly prevalent amongst adolescents and young adults with rates exceeding that of

the overall adult population.[153] A growing body of evidence suggests that these devices have medium and long-term health effects and should not be regarded as safe alternatives to tobacco smoking.[154,155] In adolescents and young adults, vaping often precedes cigarette smoking and is associated with the misuse of cannabis, unprescribed ADHD medication, and/or alcohol use.[156] Interventions should focus on cessation of e-cigarette use in adolescent LTx candidates and should be implemented throughout the transplant journey.

7.6.3. Cannabis

The use of cannabis (tetrahydrocannabinol, THC) in transplant recipients remains controversial in light of its legalization for both recreational and medical use in some jurisdictions.[157] While there is no consistent association between cannabis use and poor post-transplant outcomes, it has been associated with higher risk of cigarette smoking, other illicit drug and alcohol abuse in both heart and kidney transplant recipients.[158,159] Additionally, the smoking of cannabis has been associated with increased risk of pulmonary aspergillosis due to fungal contamination of cannabis.[160,161] Similar to adult guidelines, inhaled cannabis use should be ceased before lung transplantation, and orally consumed cannabis should only be used before transplantation if approved by the LTx team. It is important to note that orally consumed cannabinoids, including those prescribed, will cause positive urinary drug tests and complicate routine drug screening efforts,[162] and that following transplantation cannabis has the potential to interact with immunosuppressants.[163-165]

7.6.4. Opioids

The safety of pre-LTx opioid use on transplant outcomes has not been widely studied in the adult population,[166,167] with no data available in pediatric candidates. The risk and benefits of opioid use as part of palliation and symptom management should be considered on an individual basis. However, opioids are addictive and excessive use with signs of dependence should be considered with caution.

7.6.5. Abstinence

Pediatric patients should be assessed for active substance misuse and, where indicated, engage in cessation treatment before LTx. The panelists generally agreed that current nicotine, cannabis, and opioid use is an absolute contraindication to pediatric LTx as previously listed in ISHLT guidelines,[8] but 1 panelist disagreed somewhat with this statement. A 6-month period of abstinence is suggested before listing, which the panelists reached consensus on, recognizing that a short duration of abstinence increases the likelihood of relapsing into these behaviors. Some panelists suggested that the language should be stronger, “recommending” or “strongly advising” this abstinence period, while a few panelists stated that it should be a mandatory period of abstinence. Although data on the effects of substance use after LTx are limited,[168] there are single cases where children required LTx due to E-cigarette/Vaping-Associated Lung Injury (EVALI).[169]

7.6.6. EVALI considerations

The period of abstinence for a child who is critically ill with irreversible lung injury from EVALI should be left up to the local transplant center if the likelihood of relapsing behaviors is low. Importantly, the use of any of these substances may resume in adolescents and young adults at any point, so routine screening should be performed in adolescents to initiate intervention and allow for access to addiction services in the post-transplant period. Notably, active vaping, tobacco use, and cannabis use is a considered a contraindication by the panel with the need for a 6-month period abstinence being required before listing a child for LTx. In panelist discussion meetings, this statement was clarified to include that active (ongoing) use at time of assessment is an important factor in considering this as a contraindication. Some panelists suggested caution regarding this length of abstinence in children who are critically ill from EVALI, possibly requiring ECLS, and that a 6-month abstinence period would be challenging to achieve in this scenario, especially if there is a low risk of relapsing to vaping or tobacco/cannabis use after LTx. Therefore, candidacy for transplantation should be considered on an individual basis by the treating lung transplant center.

SUBSTANCE MISUSE: Regular vaping, tobacco use, and cannabis use is an absolute contraindication to lung transplantation.
Average response = +3.90
Range = –2 to +5
SD = 1.61

SUBSTANCE MISUSE: 6 months of abstinence from vaping, tobacco use, and cannabis use is suggested before listing.
Average response = +4.05
Range = +1 to +5
SD = 1.02

7.7. Psychosocial assessment

Panelists unanimously agreed that mental health professionals (i.e., psychologist, psychiatrist, and/or psychiatric nurse/nurse practitioner) and social workers with insight into pediatric SOT, should take part in the assessment of LTx candidates during the evaluation process.[170] When possible, a psychosocial assessment by a local provider knowledgeable in the case should be included in the referral provided to the transplant center.

PSYCHOSOCIAL: Psychosocial evaluation should be undertaken by a pediatric-trained psychologist or other health professional with training in mental health.
Average response = +4.75
Range = +3 to +5
SD = 0.61

The psychosocial assessment should be part of the multidisciplinary approach at the time of the evaluation and, if possible, at multiple points throughout the transplant process. It is important to reassess patients that have received mental health treatment to identify changes during the wait period. Special circumstances, for example acute decompensation or newly diagnosed end-stage lung disease, may create a situation of extreme duress for the child and their caregivers. In these situations, the assessment may be less comprehensive, and there may not be time to intervene therapeutically before transplantation.[171]

Despite their use in adult candidates, the application of standardized psychosocial assessment tools to the pediatric population is challenging as they have not been adequately validated in children.[171] More importantly than the tool used, the importance of the assessment is to take into consideration not only the specific patient factors that will influence transplant outcomes but also the socio-cultural context of the candidate. It is an opportunity to not only gain insights into the candidate and their caregivers, but also to provide education around the transplant process that is developmentally appropriate and tailored to the patient's learning needs, and to identify issues which require intervention to improve candidacy.[172]

The majority of panelists agreed that neuropsychological assessment can be of aid in the assessment of the pediatric LTx recipient but is not regarded as an essential part of suitability assessment for all potential candidates. Rather than being used to identify reasons that would exclude individuals from accessing transplantation, the goal of neuropsychological testing is to identify the individual patient's needs and to develop early interventions to improve candidacy. Pretransplantation, such testing allows the multidisciplinary team to learn about the child's cognitive level and their capacity to participate in medical conversations, as well as the ability to manage their care. Clinical neuropsychologists can recommend patient-specific accommodations for transplant education and can also participate in the creation of action plans to optimize patient and family's readiness for successful LTx.[173] Regarding the psychological assessment, panelists achieved consensus and agreed on several important issues, including the need for psychosocial functioning of both the child and the caring / family unit by a psychologist or other health professional with pediatric experience and training in mental health. There was general agreement that this assessment should be similar to other pediatric SOT recipients. There was strong agreement achieved among panelists that, when assessing adolescents and young people, there should also be a focus on adherence, substance use, and the development of independence. Finally, panelists generally agreed, with some variation in responses, that neuropsychological testing should not necessarily be universally performed in children undergoing evaluation for LTx but should be considered on an individual basis.

PSYCHOSOCIAL: Psychosocial assessment for pediatric lung transplant recipients differs to that of adult recipients and should include assessment of both the psychosocial functioning of the child and the caring / family unit.
Average response = +4.96
Range = +4 to +5
SD = 0.20

PSYCHOSOCIAL: Psychosocial assessment of pediatric lung transplant recipients is similar to that of other pediatric SOT recipients.
Average response = +3.68
Range = 0 to +5
SD = 1.39

PSYCHOSOCIAL: Psychosocial assessment of adolescents and young people require an additional focus on adherence, substance use, and the development of independence.
Average response = +4.92
Range = +4 to +5
SD = 0.28

PSYCHOSOCIAL: The role of neuropsychological testing should be determined on an individual and is NOT an essential part of all psychosocial assessments.
Average response = +3.91
Range = 0 to +5
SD = 1.24

8. LUNG RETRANSPLANTATION

Given the limited longevity of lung allografts, lung retransplantation is more frequently being considered and ultimately performed in pediatric LTx recipients under 18 years of age. In earlier eras (1992-2000), according to the ISHLT International Thoracic Organ Transplant (TTX) Registry, retransplant accounted for 6.4% of all pediatric LTx internationally, whereas it accounted for 5.2% in the more recent era (2010-2018).[174] This varies slightly based on location: 6.4% in Europe, 5.0% in North America, and 2.1% in other countries in the more recent era.[175] Retransplant has the lowest survival in the first 12 months post transplant compared to CF or any other diagnosis, with 6-month survival of 78.5% and 1-year survival of only 70.3%, based on international data from 2000 to 2017.[175] Early acute retransplantation within 12 months after primary LTx has been reported to have very poor outcomes in children,[176] similarly to in adults.[177] However, further analysis is required given the lack of available data on this rare occurrence. Importantly, CLAD due to AMR being an indication for retransplantation is now considered a relative contraindication for repeat LTx as the outcomes for this population are often poor.[8] Overall, all panelists strongly agreed that children should be considered for lung retransplantation in select cases, recognizing the risks and poorer outcomes that are also present in the adult population when comparing to primary LTx.

Evaluation of pediatric LTx recipients for retransplantation should pay special attention to the reason for graft failure, adherence and support, potential allosensitization, and patient and family goals of care (strong agreement by all panelists). Due to the importance that non-compliance may play in graft failure, particularly in the adolescent or recipient transitioning to adult care, this should be carefully assessed for and mitigated. All panelists unanimously agreed that the most important considerations around timing of listing for retransplantation are: (1) time since initial transplant, (2) rate of deterioration after initial LTx, (3) reason for graft failure including non-adherence, (4) need for supportive care and therapies, and (5) donor lung availability. This is ultimately a multifactorial decision that involves judicious assessment of all potential risk factors, including high degree of allosensitization, for potential poor outcomes. All panelists agreed that factors that substantially increase the risk for a child undergoing retransplantation include: unknown etiology of graft failure, early graft failure within 1 year (particularly AMR or restrictive CLAD), refractory non-adherence after first transplant, poor follow-up, active PTLD, and significant comorbidities due to first transplant. Additionally, ethical considerations should be considered when considering pediatric lung retransplantation. These include the overall scarcity of pediatric organs (considerations around justice), poorer outcomes compared to initial lung transplant, and patient versus parental wishes which would include conversations around goals of care.

RETRANSPLANTATION: Children should be considered for lung retransplantation in select cases.
Average response = +5.00
Range = +5 to +5
SD = 0

RETRANSPLANTATION: Evaluation for pediatric lung retransplantation should pay special attention to the reason for graft failure, adherence and supports, potential allosensitization, and patient and family goals of care.
Average response = +5.00
Range = +5 to +5
SD = 0

RETRANSPLANTATION: The most important considerations around timing of listing for retransplantation are: time since initial transplant, rate of deterioration after initial lung transplant, reason for graft failure including non-adherence, need for supportive care and therapies, and donor lung availability.
Average response = +4.87
Range = +4 to +5
SD = 0.34

RETRANSPLANTATION: Factors that substantially increase the risk for a child undergoing retransplantation include: unknown etiology of graft failure, early graft failure within 1 year (particularly AMR or restrictive CLAD), refractory non-adherence after first transplant, poor follow-up, active PTLD, and significant comorbidities due to first transplant.
Average response = +4.91
Range = +4 to +5
SD = 0.29

RETRANSPLANTATION: Ethical considerations should be taken into account when considering pediatric lung retransplantation. These include scarcity of pediatric organs, poorer outcomes compared to initial lung transplant, and patient versus parental wishes.
Average response = +4.43
Range = +3 to +5
SD = 0.66

9. MULTIORGAN TRANSPLANTATION

Pediatric MOT including LTx is a rare and complex endeavor that should be performed by select centers with expertise in thoracic +/- abdominal SOT for children. Analyses from North America reveal that adult MOT resulted in lower rejection rates for combined transplants that included heart, liver, and kidney, which appears to be due to various immunologic mechanisms.[178] Historically, HLT has been the most common MOT in children, followed by heart-kidney, followed by lung-liver, with other combinations being even rarer. As of 2017 in the United States, there were > 700 HLTs in children reported to the UNOS since 1990, with 20 lung-liver transplants, 6 heart-lung-other transplants, and 1 lung-kidney transplant.[66] In addition, an early phase trial of combined pancreatic islet-lung transplantation for CF-related diabetes demonstrated feasibility and promising 1-year outcomes.[179] Importantly, the frequency of HLT in children has dramatically decreased over time as more children with CF and PH are undergoing LTx rather than HLT.[2,180,181] Additional indications include cardiopulmonary failure as a complication of primary LTx, ILD, and other lung diseases.[2,182] The most recent estimate of median survival for children undergoing HLT was 5.2 years.[2] In studies of UNOS and the United Kingdom national registries, the 5-year survival has been estimated at 48% to 61.5%.[183,184] Waitlist mortality for HLT in children is estimated to be approximately 40%, and median waitlist time is estimated at 103 days (IQR: 34-342).[181,183] Analyses in children have revealed that pediatric MOT that include lungs resulted in similar survival rates compared to LTx alone; however, HLT is known to result in poorer survival compared to heart alone (albeit similar to lung alone).[2,185]

Published outcomes on pediatric lung-liver transplant (LLTx) outcomes are even more scarce, with the literature demonstrating that survival after combined lung-liver transplant in children is also poorer compared to transplanting either organ alone.[65] However, in a small series, children who underwent LLTx had lower rates of BO compared to matched LTx controls, which was posited to be due to potential differences in rejection.[186] The primary indication for LLTx is end-stage liver disease in the setting of advanced lung disease in CF.[186-189] Previous work in adults shows that the indications for lung-kidney transplantation are diverse, and often related to calcineurin inhibitor-related toxicity, although these procedures remain rare.[190] Overall, survival for non-HLT related MOT involving lungs is generally encouraging, albeit this being a scarce occurrence in the pediatric population.

Ultimately, based on worldwide experience, panelists unanimously agreed that children should be considered for MOT with lung transplantation. Timing of referral for pediatric candidates being considered for MOT including LTx should be done as early as possible, given the additional evaluation time needed and the scarcity of high quality pediatric multiorgan donors. All panelists agreed that the timing of referral for MOT should be earlier than that for single-organ transplantation. The approval process for these candidates requires agreement by all pediatric SOT teams, with multidisciplinary discussion being strongly recommended by all panelists from each organ group involved. Finally, MOT with LTx in pediatrics should be performed at select centers with this expertise; these centers should have well-established synergies between all pediatric SOT programs.

MULTIORGAN TRANSPLANTATION: Children should be considered for multiorgan transplantation with lung transplant. Average response = +5.00 Range = +5 to +5 SD = 0
MULTIORGAN TRANSPLANTATION: Timing for referral for candidates being considered for multiorgan transplantation with lung transplant should be as early as possible, given additional evaluation time needed and scarcity of pediatric multiorgan donors. This should be earlier than for single organ transplant. Average response = +4.92 Range = +4 to +5 SD = 0.28
MULTIORGAN TRANSPLANTATION: The approval process for pediatric candidates undergoing multiorgan transplantation with lung transplant requires agreement by all pediatric solid organ transplant teams, with multidisciplinary discussion from each organ group involved. Average response = +5.00 Range = +5 to +5 SD = 0
MULTIORGAN TRANSPLANTATION: Multiorgan transplantation with lung transplant in pediatrics should be performed at centers with this expertise and with well-established synergies between pediatric solid organ transplant programs. Average response = +4.88 Range = +4 to +5 SD = 0.34

10. DISEASE-SPECIFIC CONSIDERATIONS

10.1. Obstructive lung diseases

10.1.1. Cystic fibrosis

CF has historically been the most common indication for lung transplantation in pediatric patients, representing approximately half of all transplants in children over the last 30 years.[2,191] There are established criteria providing guidance on when patients with advanced CF lung disease should be referred for transplant evaluation.[12,192] It is recommended that children be referred when 1) FEV1 is < 50% predicted and the patient is rapidly declining, 2) FEV1 is < 40% predicted with markers of shortened survival (such as concomitant PH, malnutrition, resting hypoxemia, and frequent hospitalizations, among others), and 3) FEV1 is < 30% predicted based on published guidelines.[8] A general concept in these guidelines is that early referral provides adequate time for multidisciplinary evaluation and education, establishment of goals of care, and provides an opportunity to optimize the patient before ultimate listing.

Advances in care for CF, most notably the development of HEMT, have resulted in a dramatic decrease in the need for LTx for patients of all ages.[193,194] Recent data from the United States CF Registry show that the total number of annual LTx has decreased by approximately 80% from the premodulator era.[195] Published criteria guiding timing of listing for LTx in pediatrics is still relevant. Due to the ever-changing landscape of CF care with HEMT, the panelists reached consensus that the previous recommendation of children under 18 years of age should be listed when FEV1 < 30% predicted[8] needs reconsideration. Some children with CF and FEV1 < 30% predicted may have life expectancy > 2 years, particularly when receiving these new agents. The ideal timing to list children with CF is multi-faceted and requires a holistic evaluation of the individual child. It is important to understand a child's current and future projected respiratory disease state, including the risk for acute life-threatening complications.

CYSTIC FIBROSIS TIMING OF LISTING: Published criteria guiding timing of listing for lung transplant in pediatrics is still relevant. However, the previous recommendation, "Patients with CF < 18 years of age should be listed when FEV1 < 30% predicted" needs to be reevaluated in the era of highly effective CFTR modulators. Some children with CF and FEV1 < 30% predicted may have life expectancy >2 years, particularly when receiving these new agents. Timing of listing children with CF is multi-faceted. Also, with changing demographics in pediatric LTx, specific criteria for children with PH and ILD need to be clarified.
Average response = +4.24
Range = +1 to +5
SD = 1.18

While uncontrolled CF-related diabetes and malnutrition can be significant barriers, it is possible to achieve post-transplant success when these comorbidities can be appropriately managed. The time from evaluation until transplant provides an opportunity for optimization. There are guidelines on how to best treat CF comorbidities as part of long-term post-transplant care.[196] However, severe coexistent hepatic disease (such as cirrhosis with portal hypertension) should be considered a contraindication to LTx alone, and patients should be referred for consideration of combined LLTx.[197,198] For the CF population, panelists reached consensus that severe malnutrition, poorly controlled diabetes mellitus, and advanced liver disease as relative contraindications with the need to improve these specific comorbidities where possible or consider MOT in the case of advanced lung and liver disease.

CYSTIC FIBROSIS COMORBIDITIES: Severe malnutrition in CF should be considered a relative contraindication to lung transplantation.
Average response = +3.05
Range = −1 to +5
SD = 1.70

CYSTIC FIBROSIS COMORBIDITIES: Poorly controlled diabetes mellitus in CF should be considered a relative contraindication to lung transplantation.
Average response = +3.15
Range = −1 to +5
SD = 1.57

CYSTIC FIBROSIS COMORBIDITIES: Advanced liver disease in CF should be considered a relative contraindication to lung transplant in children, and patients should be evaluated for combined lung-liver transplantation.
Average response = +4.21
Range = +1 to +5
SD = 0.98

Even with the onset of CFTR modulators, approximately 10% of patients with CF do not yet have an effective therapy available based on their individual genotype, and even more may not have access to this therapy due to financial or other social limitations.[195,199] As the international CF community continues to strive towards developing highly effective therapies and ensuring access for all patients, it is important to recognize that those patients not currently receiving modulators are at higher risk for clinical deterioration and poor outcomes. As such, panelists strongly agreed that children who are not treated with HEMT should be referred and undergo early evaluation and should be given special consideration for transplant listing.

CFTR MODULATOR THERAPY: Early evaluation and special consideration for transplant should be given to children with advanced CF lung disease who are NOT eligible for effective CFTR modulator therapy based on genotype.
Average response = +4.70
Range = +4 to +5
SD = 0.47

10.2. Non-CF bronchiectasis

Children with advanced lung disease from non-CF bronchiectasis (such as primary ciliary dyskinesia) may benefit from transplant.[200] There is data, based primarily in adult patients, that suggests those with non-CF bronchiectasis

may have better survival awaiting transplant, but worse post-transplant survival, than those with CF.[201,202] Therefore, it is important for children with non-CF bronchiectasis to be considered on an individual basis when considering optimal timing of transplant, with caution in extrapolating guidelines taken from the CF experience.

10.3. Bronchiolitis obliterans

BO is a pathological term describing obstruction of the small airways. It can be further classified based on etiology including post-infectious BO, BO due to other inflammatory or immune-mediated insults, post-BMT/HSCT graft-versus host disease BOS, and post-LTx chronic lung allograft dysfunction BOS. These subtypes of BO are important to recognize as they have different treatments and prognosis. In pediatrics, BO can be progressive and result in advanced lung disease and the need for LTx.[203] Children that develop BO as a sequela to post-BMT/HSCT related pulmonary graft versus host disease are challenging candidates. Panelists agreed that patients with BO / pulmonary graft versus host disease related to BMT/HSCT should undergo comprehensive evaluation to confirm that they are cancer-free and at low risk for recurrence before being listed for LTx. It is widely recognized that LTx recipients may develop BO as a manifestation of CLAD. Providing retransplant to children with a BO phenotype of CLAD is an important life-sustaining treatment option in many cases.[176]

BRONCHIOLITIS OBLITERANS: Children with bronchiolitis obliterans / pulmonary graft versus host disease related to bone marrow transplantation / hematopoietic stem cell transplantation should undergo comprehensive evaluation to confirm a cancer-free status as well as low-risk for future recurrence before being listed for lung transplant.
Average response = +4.45
Range = +3 to +5
SD = 0.76

10.4. Bronchopulmonary dysplasia

BPD is chronic lung disease related to premature birth and has an increasing incidence as BPD treatments have improved in recent decades. While most transplants for patients with BPD occur in young adults, successful life-sustaining transplantation has also been reported in young children.[91,92] Therefore, LTx should be considered as a treatment option for infants and young children with severe BPD. Additional studies are needed to understand specific comorbidities that may predict children who would benefit most from early transplant versus chronic management. Panelists reached consensus, with some variation noted in responses, that LTx is a treatment option for infants and young children with severe BPD, and those being transplanted should be patients for whom sustained growth has not resulted in improvement in their respiratory health and/or quality of life.

BRONCHOPULMONARY DYSPLASIA: Lung transplantation should be considered as a treatment option for infants and young children with severe bronchopulmonary dysplasia.
Average response = +3.84
Range = –3 to +5
SD = 1.83

11. RESTRICTIVE LUNG DISEASES

Restrictive lung diseases requiring LTx in children are predominantly lung parenchymal disorders. Due to new therapies for CF, advancements in managing PVD, and improvements in ECMO, indications for pediatric LTx have transformed over recent years, accommodating more severe cases as expertise grows.[98,204-206] Consequently, children with restrictive lung diseases and ILD, including inherited surfactant disorders, refractory ARDS (from infectious and other causes), fibrotic diseases (post-malignancy related to chemotherapy/radiotherapy, poststem

cell transplant, i.e., PPFE), rheumatologic diseases and other ILDs, now make up a larger proportion of the total pediatric LTx population.[3,207,208] Many ILDs in children carry both a higher waitlist mortality and a higher frequency of comorbidities related to the severity of illness and other end-organ toxicity resulting from treatment. Therefore, early referral for LTx consideration is essential to ensure an opportunity for candidacy.

Childhood ILD is an umbrella term for a variety of rare diffuse lung diseases of different etiologies and prognoses.[209-211] These have become more important over time, accounting for a greater proportion of the indications for LTx in recent years: LTx for childhood ILDs accounted for 32.4% of all pediatric LTx in the United States in 2020-2023, compared to 20.3% in 2016-2019.[3,4] Some forms are associated with significant morbidity and mortality, particularly if they manifest in the first year of life.[206] Surfactant dysfunction disorders make up the largest group, often following a severe course in infants and young children.[212-215] Since the efficacy of treatments (i.e., systemic corticosteroids) has yet to be established in randomized trials, and due to the potential for rapid progression, children with severe progressive disease should be considered for LTx at an early stage. The panelists achieved high-level consensus regarding the consideration of LTx in infants and children with restrictive lung diseases, with the recommendation of referring these patients early.

RESTRICTIVE DISEASES: Early referral to a transplant center is essential for any child with a restrictive lung disease that may progress to organ failure.
Average response = +4.95
Range = +4 to +5
SD = 0.22

Achieving an accurate etiologic diagnosis is important for this process but is not absolutely required.[207,209,216] If a pathogenic genetic variant is identified (where genetic testing is available), an additional lung biopsy may not provide benefit in assessing prognosis or guiding treatment decisions and carries a significant risk of complications in critically ill children.[207,209] However, in the absence of a genetic diagnosis, a biopsy may be of utility, except in cases of advanced fibrosis or when the risk of complications in critically ill children outweighs the potential benefits of the procedure. Panelists universally agreed that genetic testing (where available) is useful for assessing children with childhood ILD / surfactant dysfunction disorders that are being considered for LTx and agreed that a lung biopsy is not always needed for determining candidacy, particularly in genetically confirmed ILD or surfactant dysfunction disorder cases.

RESTRICTIVE DISEASES: Patients with childhood ILD / surfactant dysfunction disorders referred for lung transplantation should undergo genetic testing for a panel of known candidate genes for these disorders and pulmonary hypertension.
Average response = +4.85
Range = +4 to +5
SD = 0.37

RESTRICTIVE DISEASES: In genetically confirmed childhood ILD / surfactant dysfunction disorders, performing a lung biopsy is NOT required for the decision of listing.
Average response = +4.40
Range = +3 to +5
SD = 0.75

After BMT/HSCT, pediatric patients can develop restrictive lung diseases such as chronic graft-versus-host disease (GVHD), pulmonary fibrosis, ILD, and PPFE, among others. These conditions often result in significant lung dysfunction, potentially leading to the need for LTx. Although LTx can be a life-saving option for patients with these restrictive pathologies, it can be associated with significant postoperative morbidity and mortality in some scenarios, particularly in cases of advanced disease or malnutrition.[217,218] Additionally, children with autoimmune diseases and rheumatologic disorders such as stimulator of interferon genes (STING)-associated vasculopathy of infancy (SAVI) and STAT3 GOF mutations can develop ILD either as cause of the disease or due to their treatment.[219,220] Given that these entities are unique to the pediatric population, childhood ILD specialist involvement is important in these cases. Panelists strongly agreed that infants and young children with ILDs and restrictive lung diseases should undergo evaluation and transplantation exclusively at centers with expertise in the comprehensive care of these patients.

RESTRICTIVE DISEASES: Infants and young children with interstitial lung diseases and restrictive lung diseases should undergo evaluation and transplantation exclusively at centers with expertise in the comprehensive care of these patients, both before and after transplantation.
Average response = +4.55
Range = +3 to +5
SD = 0.60

The risk of concomitant PH is increased in children with restrictive lung disease, which may affect timing of listing, so all patients referred for consideration of LTx should undergo echocardiographic screening for PH, ideally performed by an experienced operator either at the referring site or at the transplant center as part of the initial assessment.[221] If detected, PH should be phenotyped and risk-stratified as per local protocols, as outlined in the subsequent section on PVD. Panelists were unanimous about the importance of screening for coexisting PH (i.e., group 3 PH) in children with childhood ILD.

RESTRICTIVE DISEASES: Children with restrictive lung diseases being assessed for potential lung transplantation should undergo a thorough evaluation for pulmonary hypertension.
Average response = +5.00
Range = +5 to +5
SD = 0

Abnormal lung parenchyma is associated with abnormal lung growth and may have secondary impacts on the developing chest wall and/or spine; these may result in additional restrictions to ventilation. Specific diagnoses are associated with typical abnormalities including pectus excavatum with childhood ILD or PPFE.[218] Abnormalities of the chest wall and/or spine may impact perioperative planning in restrictive lung diseases, as the intrathoracic space may not be an appropriate shape or size to receive donor lungs or may preclude effective secretion clearance post-LTx. Panelists unanimously agreed that in children with restrictive pulmonary physiology should undergo assessment of the chest wall structure and intrathoracic capacity for evaluating surgical feasibility, optimizing future graft function, assessing risks, tailoring individualized treatment plans, and predicting post-transplant outcomes.

RESTRICTIVE DISEASES: Assessing the chest wall structure and intrathoracic capacity is essential for evaluating surgical feasibility, optimizing graft function, assessing risks, tailoring individualized treatment plans, predicting post-transplant outcomes, and effectively allocating resources in patients with restrictive lung diseases undergoing lung transplantation.
Average response = +4.95
Range = +4 to +5
SD = 0.23

Supportive care for children with restrictive lung diseases can reduce symptom burden and influence quality of life; however, due to the rarity of the conditions and paucity of specific pediatric research, most treatment algorithms are empirical, influenced by evidence in adult ILDs and based on case series with few randomized control trials conducted in children thus far.[222,223] While trialing pharmacotherapeutic options (such as a tyrosine kinase inhibitor, Janus kinase-2 inhibitor, etc.) may be appropriate in select cases, referral and listing for LTx should not be delayed awaiting treatment response.

Finally, due to several multisystemic autoimmune or autoinflammatory disorders being contributory to childhood ILDs, the panelists agreed these conditions should be considered as relative contraindications, and LTx candidacy should be considered on a case-by-case basis, considering potential multiorgan dysfunction, nutritional and mobility status, and the risk of primary disease recurrence after transplant. The unmet needs of childhood ILD remain high with further research into this heterogenous group of diseases required, which will likely lead to changes to these recommendations based on future advancements in the field. Children referred for LTx should also be offered entry into childhood ILD registries and clinical trials where available.

Table 4 Specific Indications for Lung Transplant in Pediatric Pulmonary Vascular Disease*

The following are indications for transplant referral (evaluation / planning) for lung or heart-lung transplantation in the setting of pulmonary vascular disease in children:	
Meeting referral criteria based on available guidelines for pediatric pulmonary vascular disease (including EPPVDN consensus, endorsed by ISHLT[8,13,15,224]). This includes remaining in an intermediate or high-risk disease category despite maximal therapy.	
Worsened functional class and deterioration of RV dysfunction despite optimal therapy for 3 to 6 months.	Average response = +5.00 Range = +5 to +5 SD = 0
Life-threatening hemoptysis.	Average response = +4.90 Range = +4 to +5 SD = 0.30
Progressive pulmonary vein stenosis despite interventions.	Average response = +4.76 Range = +3 to +5 SD = 0.54
Certain genetic variants; these impact risk stratification and timing of referral and/or listing for transplant: that is, *FOXF1*, *E1F2AK4*, *BMPR2*, and other mutations.	Average response = +4.62 Range = +3 to +5 SD = 0.59

*Refer to panelist voting results in the text and in the appendix for supporting statements that refer to each of these points.

RESTRICTIVE DISEASES: Interstitial lung diseases primarily resulting from multisystemic autoimmune / autoinflammatory disorders are relative contraindications to lung transplantation. Patients should be considered on an individual basis, taking into account the degree of multiorgan dysfunction, nutritional / mobility status, and the risk of recurrence after the transplant.
Average response = +4.53
Range = +3 to +5
SD = 0.61

12. PULMONARY VASCULAR DISEASES

Pediatric PH is a complex and progressive disease with multiple distinct etiologies.[224,225] While many treatment options exist, LTx has an important role in the treatment of advanced PVD.[3,101] Guidelines for patient management have included recommendations for evaluation and referral for LTx in pediatric patients with PH.[8,13,15] Indications for referral, evaluation, and listing for LTx are based on functional status and markers of disease severity in pediatric PVD,[42,224] and are defined in Table 4, based on panelist voting.

Panelists unanimously agreed that critical factors for referral/evaluation include deterioration in functional class *and* worsening RV dysfunction *despite* optimal treatment for 3 to 6 months. In general, the panelists agreed that surrogate measures for cardiovascular function such as N-terminal pro-B-type natriuretic peptide (NT-proBNP) or B-type natriuretic peptide (BNP), RV function, and exercise testing are used to refine clinical assessment of disease severity, possible response to treatment, and management; however, in isolation, these are not individual criteria that alone can be used for or against referral for transplantation. Specifically, no consensus was reached by the panelists for the statement that the "downward trend in BNP [alone] is adequate evidence for treatment response for PAH in children"; this statement resulted in a wide range of responses, from strong disagreement to agreement. Life-threatening hemoptysis or progressive pulmonary vein stenosis that are refractory to interventions are also appropriate indications for referral/evaluation. Specific syndromes associated with less favorable outcomes, such as alveolar capillary dysplasia/misalignment of pulmonary veins (ACD/MPV) or pulmonary veno-occlusive disease, frequently associated with genetic variants in FOXF1 or E1F2AK4 respectively, should be referred for urgent transplant evaluation. There was unanimous agreement on this point. This includes other forms of rapidly progressive PH such as pulmonary capillary hemangiomatosis. Other genetic variants such as BMPR2 may help inform timing of referral and risk stratification in children with PH.

PULMONARY VASCULAR DISEASES: Worsened functional class and deterioration of RV dysfunction despite optimal therapy for 3 to 6 months are general indications for evaluation / planning for lung or heart-lung transplantation in the setting of pulmonary vascular disease in children. Average response = +5.00 Range = +5 to +5 SD = 0
PULMONARY VASCULAR DISEASES: Improvement in RV function AND functional class would constitute adequate evidence for treatment response. Average response = +4.05 Range = +2 to +5 SD = 0.86
PULMONARY VASCULAR DISEASES: Downward trend in BNP is adequate evidence for treatment response for PAH in children. Average response = +0.65 Range = −4 to +3 SD = 2.20
PULMONARY VASCULAR DISEASES: 3 to 6 months is adequate time to gauge treatment response in PAH before consideration of pediatric lung transplant. Average response = +3.67 Range = 0 to +5 SD = 1.20
PULMONARY VASCULAR DISEASES: Life-threatening hemoptysis is an indication for evaluation / planning for lung or heart-lung transplantation in the setting of pulmonary vascular disease in children. Average response = +4.90 Range = +4 to +5 SD = 0.30
PULMONARY VASCULAR DISEASES: Progressive pulmonary vein stenosis despite interventions is an indication for evaluation / planning for lung or heart-lung transplantation in the setting of pulmonary vascular disease in children. Average response = +4.76 Range = +3 to +5 SD = 0.54
PULMONARY VASCULAR DISEASES: Certain genetic variants impact risk stratification and timing of lung transplant referral / listing for pulmonary vascular disease in children: for example, FOXF1 with ACD/MPV, E1F2AK4, BMPR2, and other mutations. Average response = +4.62 Range = +3 to +5 SD = 0.59

Clinical alternatives to LTx for advanced PH include atrial septostomy or reverse Potts shunt.[98,102,226] Patients in whom a reverse Potts shunt has been performed through a thoracotomy (posterolateral or lateral approach) are at increased risk for LTx-related surgical complications including bleeding; in many centers, this is a relative or absolute contraindication to LTx. However, there was no agreement by panelists on whether a reverse Potts shunt performed through via either thoracotomy approach is a contraindication for LTx in children, with responses varying from strong disagreement to strong agreement. Upon further discussion, it was deemed that this type of procedure, if being performed, should be done at centers with experience in doing so with acceptable outcomes. Of the surgeons on the panel that perform surgical pulmonary-to-systemic arterial shunts, their opinion is in agreement with a recent clinical practice guideline[101,102] that reverse Potts shunt should be performed from an anterior approach via a sternotomy with a graft if LTx is to be considered in the future. ECMO for potential short-term circulatory support, or PLAD (pulmonary artery to left atrial circuit with pump and oxygenator) which can provide RV rehabilitation, are options for bridging pediatric patients with severe PVD awaiting LTx.[33,67] It should be noted that the occurrence of left ventricular (LV) diastolic dysfunction in the preoperative period has been associated with the development of primary graft dysfunction[227-229]; therefore, awareness of this situation is recommended to allow for early post-LTx prognostication and early graft management. Postoperative ECMO may be used in certain recipients to reduce the risk of primary graft dysfunction and has also been utilized post-operatively to assist in LV remodeling through controlled LV loading.[230,231]

Diastolic and systolic function of both the LV and RV are highly important in a child after LTx, so there was significant discussion given to this subject matter. The majority of panelists agreed, reaching consensus, that LTx alone is contraindicated in PH patients with pretransplant severe LV systolic or diastolic dysfunction as well as in

those with unrepaired complex congenital heart disease; these children may be potential candidates for HLT.[181] RV dysfunction, on the other hand, in children with PVD is a potential risk factor for post-LTx primary graft dysfunction.[227] However, given that RV function has been shown to improve after LTx, baseline RV dysfunction is not a contraindication to LTx in children.[232-234] Panelists agreed on this point, although with some variation in responses: consensus was reached, with the majority agreeing that RV dysfunction in children with PVD is a risk factor, but not a contraindication, for pediatric LTx without heart transplant. Given the complexity of treatment strategies and decision-making, all panelists agreed that it is essential that pediatric LTx teams work in close collaboration with pediatric PH specialists and other interdisciplinary providers to optimize care and timing of listing for transplantation in children with PVD.

PULMONARY VASCULAR DISEASES: Reverse Potts shunt thoracotomy (posterolateral or lateral approach) in children with PVD is a contraindication to pediatric lung transplant. Average response = –0.05 Range = –5 to +5 SD = 2.98
PULMONARY VASCULAR DISEASES: Refractory, moderate LV systolic or diastolic dysfunction in children with PVD is a contraindication for lung transplant alone, and an indication for heart-lung transplantation. Average response = +3.67 Range = +1 to +5 SD = 1.28
PULMONARY VASCULAR DISEASES: RV dysfunction in children with PVD is risk factor for poor outcome, but not a contraindication, for pediatric lung transplant (without heart transplant). Average response = +3.81 Range = –1 to +5 SD = 1.50
PULMONARY VASCULAR DISEASES: Use of ECMO in the immediate postoperative period can be a useful approach to reduce the risk of primary graft dysfunction in children with PH who undergo lung transplant. Average response = +4.24 Range = +1 to +5 SD = 1.09
PULMONARY VASCULAR DISEASES: Pediatric lung transplant providers should engage in close collaboration with the pediatric cardiologists / PH specialists to optimize care and timing of listing for patients with PVD. Average response = +5.00 Range = +5 to +5 SD = 0

13. FUTURE DIRECTIONS

As the landscape of pediatric LTx evolves and outcomes continue to slowly improve, clinicians caring for these children must consider how to continue improving access to this treatment option. We discuss a few evolving issues that would benefit more immediate attention. A prevalent obstacle in some children with childhood ILD being considered for LTx is the increasing corticosteroid dosages being prescribed pretransplant, which can be prohibitive for future LTx candidacy. A recent opinion piece published by some of the panelists provides some guidance on this important issue,[235] but this needs to be addressed by conducting longitudinal, multicenter studies in the future to help determine best practice. In addition, through the Delphi consensus process, we have identified select topics that require future exploration to help provide standardized and coordinated care to pediatric LTx candidates. These include but are not limited to: candidacy and management of pediatric patients with infections that are without therapeutic options from Aspergillus species, Scedosporium/Lomentospora, and other molds; candidacy of pediatric patients with *Burkholderia cenocepacia*; timing around listing of specific ILDs based on individual disease profiles and progression in childhood; and timing of listing for individual pediatric PVDs based on evolving pediatric PH guidelines and approaches for advanced disease.

Once approved for LTx, a major challenge facing waitlist children globally is the scarcity of donor organs as the pediatric donor consent rate has been declining[90]; an opportunity to improve access and potentially shorten

wait times is by using *ex vivo* lung perfusion.[236-238] This perfusion strategy may allow the extension of the potential organ procurement area by over 400% as compared to a standard cold-storage preservation strategy, and it is a well-established tool in the field of adult LTx by now. Another surgical approach that may assist in improving access to donor lungs for children is the use of lobes from living or deceased donors for pediatric candidates as this is being performed in Japan with great success.[239] As the complexity of children being referred for LTx increases, the pediatric LTx community needs validated tools to objectively evaluate and monitor the frailty of this pediatric population as we work towards improving access and outcomes for LTx. The evolution of these specific challenges and new ones that will undoubtedly develop will be a key part of future updates of this consensus document.

14. CONCLUSIONS

Pediatric LTx outcomes can vary significantly depending on the clinical characteristics of candidates and the expertise of the local center caring for them. This consensus statement is the first to create categories for risk factors and acknowledge those risk factors that need consideration in the context of considering children as a candidate. Moreover, certain centers may choose to develop specialized expertise in addressing certain higher risk conditions. Whenever possible, all potentially modifiable risk factors should be optimized before LTx to yield the most successful long-term outcomes. Further, no prognostic tools have solid evidence in predicting outcomes in children, so this consensus document is for guidance when discussing LTx as a treatment option with patients and caregivers based on individual desires for the remaining part of their life.

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APPENDIX A. SUPPLEMENTARY DATA

Supplementary data associated with this article can be found in the online version at doi:10.1016/j.healun.2025.08.005.

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