Clinical Policy: Lung Transplantation

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Coding Implications
Revision Log

Description
This policy describes the medical necessity criteria for the review of lung transplantation requests.

The below criteria are sourced from the International Society for Heart and Lung Transplantation (ISHLT) 2021 Consensus Document for the Selection of Lung Transplant Candidates.[19]

The ISHLT consensus document that the below criteria are derived from provides guidelines based on expert synthesis of the current literature with a goal of improving survival and quality of life in transplant candidates. ISHLT recognizes that donor lungs are a limited societal resource, requiring that guidance on candidate selection be based on survival benefit. Given the rigor of the guidelines on which this policy is based, the benefits of receiving a lung transplant in individuals meeting the criteria below outweighs the potential risk of adverse outcomes related to receiving a transplant that is not indicated or not receiving a transplant that is indicated.

Policy/Criteria

I. It is the policy of Pennsylvania Health and Wellness® that lung transplantation for members/enrollees with chronic, end-stage lung disease who have failed maximal medical (including pulmonary rehabilitation, as applicable) or surgical therapy is **medically necessary** when all the following criteria are met:

A. High (> 50%) risk of death from lung disease within two years if lung transplantation is not performed[19];

B. High (> 80%) likelihood of five-year post-transplant survival from a general medical perspective provided there is adequate graft function[19];

C. Does not have ANY of the following absolute contraindications[19]:

1. Malignancy with high risk of recurrence or death related to cancer;
2. Glomerular filtration rate < 40 mL/min/1.73m^2 unless being considered for multi-organ transplant;
3. Acute renal failure with rising creatinine or on dialysis and low likelihood of recovery;
4. Acute liver failure, or cirrhosis with portal hypertension or synthetic dysfunction unless being considered for multi-organ transplant;
5. Stroke, acute coronary syndrome, or myocardial infarction (excluding demand ischemia) within 30 days;
6. Septic shock;
7. Active extrapulmonary or disseminated infection;
8. Active *tuberculosis* infection;
9. HIV infection with detectable viral load;
10. Progressive cognitive impairment;
11. Inability to adhere to the regimen necessary to preserve the transplant, even with caregiver support;
12. Other severe, uncontrolled medical condition expected to limit survival after

transplant;

13. Active substance use or dependence including current tobacco use, vaping, marijuana smoking, or IV drug use;

D. Has one of the following disease states (not an all- inclusive list):

1. Adult members/enrollees, age ≥ 18:

a. Interstitial lung disease and any of the following[19]:*
   i. Absolute decline in forced vital capacity (FVC) > 10% in the past six months despite appropriate treatment;
   ii. Absolute decline in diffusing capacity of the lung for carbon monoxide (DLCO) > 10% in the past six months despite appropriate treatment;
   iii. Absolute decline in forced vital capacity (FVC) > 5% with radiographic progression in the past six months despite appropriate treatment;
   iv. Desaturation to < 88% on six-minute-walk test (6MWT) or > 50 m decline in 6MWT distance in the past six months;
   v. Pulmonary hypertension on right heart catheterization or two dimensional echocardiography (in the absence of diastolic dysfunction);
   vi. Hospitalization because of respiratory decline, pneumothorax, or acute exacerbation;

b. Cystic fibrosis (CF) or other causes of bronchiectasis and any of the following[19]:
   i. FEV1 < 25% predicted despite optimal medical management including a trial of elexacaftor/tezacaftor/ivacaftor if eligible;
   ii. Both of the following:
      a) Any of the following despite optimal medical management including a trial of elexacaftor/tezacaftor/ivacaftor if eligible:
         1) FEV1 < 30% predicted;
         2) FEV1 < 40% predicted and any of the following:
            i. Six-minute walk distance < 400 meters;
            ii. PaCO2 > 50 mmHg;
            iii. Hypoxemia at rest or with exertion;
            iv. Pulmonary hypertension (PA systolic pressure > 50 mmHg on echocardiogram or evidence of right ventricular dysfunction);
            v. Worsening nutritional status despite supplementation;
            vi. Two exacerbations per year requiring intravenous antibiotics;
            vii. Massive hemoptysis (>240 mL) requiring bronchial artery embolization;
            viii. Pneumothorax;
         3) FEV1 <50% predicted and rapidly declining based on pulmonary function testing or progressive symptoms;
         4) Any exacerbation requiring positive pressure ventilation;
      b) Any of the following[19]:
         1) Rapid decline in lung function or progressive symptoms (>30% relative decline in FEV_1 over 12 months);
         2) Frequent hospitalization, particularly if > 28 days hospitalized in the preceding year;
         3) Any exacerbation requiring mechanical ventilation;

4) Chronic respiratory failure with hypoxemia or hypercapnia, particularly for those with increasing oxygen requirements or needing long-term non-invasive ventilation therapy;
5) Pulmonary hypertension (Pulmonary arterial systolic pressure >50 mmHg on echocardiogram or evidence of right ventricular dysfunction);
6) Worsening nutritional status particularly with body mass index (BMI) <18 kg/m^2 despite nutritional interventions;
7) Recurrent massive hemoptysis despite bronchial artery embolization;
8) World Health Organization (WHO) Functional Class IV;

c. Chronic obstructive pulmonary disease (COPD), and any of the following[19]:
   i. BODE score (includes BMI, degree of airflow obstruction, degree of dyspnea, and exercise capacity) of 7 to 10;
   ii. FEV_1 (forced expiratory volume in one second) < 20% predicted;
   iii. History of severe exacerbations;
   iv. Chronic hypercapnia;
   v. Moderate to severe pulmonary hypertension;
d. Pulmonary vascular diseases and any of the following[19]:
   i. European Society of Cardiology/European Respiratory Society (ESC/ERS) high risk or Registry to Evaluate Early and Long-term Pulmonary Arterial Hypertension Disease Management (REVEAL) risk score >10 on appropriate pulmonary arterial hypertension (PAH) therapy, including intravenous (IV) or subcutaneous (SC) prostacyclin analogues;
   ii. Progressive hypoxemia;
   iii. Progressive, but not end stage, liver, or kidney dysfunction due to PAH
   iv. Life-threatening hemoptysis;
   v. PAH in the European Pediatric Pulmonary Vascular Disease Network (EPPVDN) high risk category and on optimal therapy without improvement[19];
e. Lymphangioleiomyomatosis (LAM) with evidence of disease progression despite mTOR inhibitor therapy and any of the following[19]:
   i. Severely abnormal lung function (e.g. FEV_1 <30% predicted);
   ii. Exertional dyspnea (NYHA class III or IV);
   iii. Hypoxemia at rest;
   iv. Pulmonary hypertension;
   v. Refractory pneumothorax;
f. Primary lung graft failure;
g. Acute respiratory distress syndrome (ARDS) with a persistent requirement for mechanical ventilatory support and /or extracorporeal life support (ECLS) without expectation of clinical recovery and with evidence of irreversible lung destruction[19];

2. *Pediatric members/enrollees, age <18*[19]*:*
   a. Cystic fibrosis, and any of the following[19]:
      i. FEV_1 < 25% predicted despite optimal medical management including a trial of elexacaftor/tezacaftor/ivacaftor if eligible[19];

ii. Both of the following[19]:
  a) Any of the following despite optimal medical management including a trial of elexacaftor/tezacaftor/ivacaftor if eligible:
    1) FEV_1 < 40% predicted[19];
    2) FEV_1 < 50% predicted and any of the following[19]:
      i) Six-minute walk distance < 400 meters;
      ii) $PaCO_2$ > 50 mmHg;
      iii) Hypoxemia at rest or with exertion;
      iv) Pulmonary hypertension (PA systolic pressure > 50 mmHg on echocardiogram or evidence of right ventricular dysfunction;
      v) Worsening nutritional status despite supplementation;
      vi) Two exacerbations per year requiring intravenous antibiotics;
      vii) Massive hemoptysis (>240 mL) requiring bronchial artery embolization;
      viii) Pneumothorax.
    3) FEV_1 <50% predicted and rapidly declining based on pulmonary function testing or progressive symptoms;
    4) Any exacerbation requiring positive pressure ventilation;
  b) Any of the following[19]:
    1) Rapid decline in lung function or progressive symptoms (>30% relative decline in FEV_1 over 12 months);
    2) Frequent hospitalization, particularly if > 28 days hospitalized in the preceding year;
    3) Any exacerbation requiring mechanical ventilation;
    4) Chronic respiratory failure with hypoxemia or hypercapnia, particularly for those with increasing oxygen requirements or needing long-term non-invasive ventilation therapy;
    5) Pulmonary hypertension (Pulmonary arterial systolic pressure >50 mmHg on echocardiogram or evidence of right ventricular dysfunction);
    6) Worsening nutritional status particularly with BMI <18 kg/m^2 despite nutritional interventions;
    7) Recurrent massive hemoptysis despite bronchial artery embolization;
    8) WHO Functional Class IV.

b. Pulmonary vascular disease and any of the following[19]:
  i. ESC/ERS high risk or Registry to Evaluate Early and Long-term Pulmonary Arterial Hypertension Disease Management (REVEAL) risk score >10 on appropriate PAH therapy, including IV or SC prostacyclin analogues;
  ii. Progressive hypoxemia;
  iii. Progressive, but not end stage, liver, or kidney dysfunction due to PAH;
  iv. Life-threatening hemoptysis;
  v. PAH in the European Pediatric Pulmonary Vascular Disease Network (EPPVDN) high risk category and on optimal therapy without improvement;[19]

c. Interstitial lung disease and any of the following[19]:

i. Absolute decline in FVC > 10% in the past six months despite appropriate treatment;
ii. Absolute decline in DLCO > 10% in the past six months despite appropriate treatment;
iii. Absolute decline in FVC > 5% with radiographic progression in the past six months despite appropriate treatment;
iv. Desaturation to < 88% on 6MWT or > 50 m decline in 6MWT distance in the past six months;
v. Pulmonary hypertension on right heart catheterization or two dimensional echocardiography (in the absence of diastolic dysfunction);
vi. Hospitalization because of respiratory decline, pneumothorax, or acute exacerbation.

d. COPD, and any of the following[19]:
i. BODE score (includes BMI, degree of airflow obstruction, degree of dyspnea, and exercise capacity) of seven to 10;
ii. FEV_1 < 20% predicted;
iii. History of severe exacerbations;
iv. Chronic hypercapnia;
v. Moderate to severe pulmonary hypertension.

e. Primary lung graft failure[19];
f. LAM with evidence of disease progression despite mTOR inhibitor therapy and any of the following[19]:
i. Several abnormal lung function (e.g. FEV1 <30% predicted);
ii. Exertional dyspnea (NYHA class III or IV);
iii. Hypoxemia at rest;
iv. Pulmonary hypertension;
v. Refractory pneumothorax.

g. ARDS with a persistent requirement for mechanical ventilatory support and /or ECLS without expectation of clinical recovery and with evidence of irreversible lung destruction[19].

**Note:* FVC may be a less reliable parameter for those with concomitant emphysema[19].

Background

Lung transplantation is an accepted therapy for the management of a range of severe lung disorders. Single, double, and lobar-lung transplants have all been successful for carefully selected patients with end-stage pulmonary disease. The most common disease processes for which lung transplants are performed include COPD, idiopathic pulmonary fibrosis, cystic fibrosis, pulmonary arterial hypertension, and sarcoidosis.

COPD is one of the most common lung diseases and is the most common indication for lung transplantation in adults. Chronic bronchitis and emphysema are the two main forms of COPD, both most commonly caused from smoking. Non-smokers with an alpha-1 antitrypsin deficiency can also develop emphysema. These conditions are the most common indications for single lung transplants. Cystic fibrosis, emphysema, and alpha-1 antitrypsin deficiency are the most common indications for double lung transplant, or sequential replacement of both lungs.

The most common indications for pediatric lung transplants include pulmonary vascular disease, bronchiolitis obliterans, bronchopulmonary dysplasia, graft failure due to viral pneumonitis, and cystic fibrosis.

Coding Implications

This clinical policy references Current Procedural Terminology (CPT®). CPT® is a registered trademark of the American Medical Association. All CPT codes and descriptions are copyrighted 2022, American Medical Association. All rights reserved. CPT codes and CPT descriptions are from the current manuals and those included herein are not intended to be all-inclusive and are included for informational purposes only. Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

CPT® Codes	Description
32850	Donor pneumonectomy(s) (including cold preservation), from cadaver donor
32851	Lung transplant, single; without cardiopulmonary bypass
32852	Lung transplant, single; with cardiopulmonary bypass
32853	Lung transplant, double (bilateral sequential or en bloc); without cardiopulmonary bypass
32854	Lung transplant, double (bilateral sequential or en bloc); with cardiopulmonary bypass
32855	Backbench standard preparation of cadaver donor lung allograft prior to transplantation, including dissection of allograft from surrounding soft tissues to prepare pulmonary venous/atrial cuff, pulmonary artery, and bronchus; unilateral
32856	Backbench standard preparation of cadaver donor lung allograft prior to transplantation, including dissection of allograft from surrounding soft tissues to prepare pulmonary venous/atrial cuff, pulmonary artery, and bronchus; bilateral

HCPCS Codes	Description
S2060	Lobar lung transplantation
S2152	Solid organ(s), complete or segmental, single organ or combination of organs; deceased or living donor (s), procurement, transplantation, and related complications; including: drugs; supplies; hospitalization with outpatient follow-up; medical/surgical, diagnostic, emergency, and rehabilitative services, and the number of days of pre- and post-transplant care in the global definition

Reviews, Revisions, and Approvals	Revision Date	Approval Date
Added Eisenmenger syndrome as a qualifying condition for adult transplant. Added that the list of qualifying conditions for transplant is not all-inclusive. Added primary lung graft failure and bronchiolitis obliterans as an indication for adult and pediatric transplant since ISHLT guidelines recommend retransplant in	09/18	10/18

Reviews, Revisions, and Approvals	Revision Date	Approval Date
certain cases. Updated coding. Added time frame for which smoking cessation should be documented. In criteria pertaining to substance use, removed the statement that serial blood and urine testing" may be required, as it is informational only. In the adult COPD criteria, changed "one severe exacerbation" to "at least one severe exacerbation."		
References reviewed and updated.	12/18	
References reviewed and updated. Specialist review Edited malignancy contraindication to not specify within 2 years, and added exceptions of early stage prostate cancer, cancer that has been completely resected, or that has been treated and poses acceptable future risk.	12/2020	1/28/2021
Replaced contraindications of "severely limited functional status with poor rehabilitation potential" and those regarding past or current nonadherence to medical therapy, and psychological condition associated with the inability to comply with medical therapy with "Inability to adhere to the regimen necessary to preserve the transplant, even with caregiver support." Changed "review date" in header to "Date of Last Revision" and "Date" in the revision log header to "Revision Date." Replaced "members" with "members/enrollees" in all instances. Annual review. References reviewed and updated. Reviewed by specialist.	10/2021	
Annual review. Added "or surgical therapy" to I and noted that maximal medical therapy includes pulmonary rehab when applicable. Updated the following based on ISHLT 2021 guidelines; removed criteria "High (> 80%) likelihood of surviving at least 90 days after lung transplantation.", updated I.C., I.D.1.a, I.D.1.b., I.D.1.c., I.D.1.d., I.D.1.f., I.D.2.a, I.D.2.b. Clarified nicotine and tobacco abstinence contraindication. Added CPT codes 32850, 32855, and 32856. References reviewed, updated, and reformatted. Reviewed by specialist.	2/22/2023	
Revised adult and pediatric criteria to align with ISHLT 2021 consensus document. References reviewed and updated.	08/2023	

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