Clinical Policy: Lung Transplantation

Reference Number: PA.CP.MP.57
Effective Date: 06/18
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Coding Implications
Revision Log

Description
Medical necessity guidelines for the review of lung transplantation requests.

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 2 years if lung transplantation is not performed.

B. High (> 80%) likelihood of 5-year post-transplant survival from a general medical perspective provided that there is adequate graft function.

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 intravenous drug use) without convincing evidence of risk reduction behaviors, such as meaningful and/or long-term participation in therapy for substance abuse and/or dependence. Serial blood and urine testing may be used to verify abstinence from substances that are of concern.
    a. If there is a history of nicotine or tobacco use, documentation notes abstinence from all tobacco and nicotine products (including nicotine replacement therapy) for ≥ 6 months prior to transplant.

A. 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:*
   i. Absolute decline in forced vital capacity (FVC) ≥ 10% in the past 6 months despite appropriate treatment;
   ii. Absolute decline in diffusing capacity of the lung for carbon monoxide (DLCO) ≥10% in the past 6 months despite appropriate treatment;
   iii. Absolute decline in forced vital capacity (FVC) ≥ 5% with radiographic progression in the past 6 months despite appropriate treatment;
   iv. Desaturation to < 88% on 6-minute-walk test (6MWT) or > 50 m decline in 6MWT distance in the past 6-months;
   v. Pulmonary hypertension on right heart catheterization or 2-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:
   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:
            (a) Six-minute walk distance < 400 meters;
            (b) PaCO2 >50mmHg;
            (c) Hypoxemia at rest or with exertion;
            (d) Pulmonary hypertension (PA systolic pressure >50mmHg on echocardiogram or evidence of right ventricular dysfunction);
            (e) Worsening nutritional status despite supplementation;
            (f) 2 exacerbations per year requiring intravenous antibiotics;
            (g) Massive hemoptysis (>240 mL) requiring bronchial artery embolization;
            (h) 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:
         - Rapid decline in lung function or progressive symptoms (>30% relative decline in FEV_1 over 12 months);
         - Frequent hospitalization, particularly if >28 days hospitalized in the preceding year;
         - Any exacerbation requiring mechanical ventilation;
         - Chronic respiratory failure with hypoxemia or hypercapnia, particularly for those with increasing oxygen requirements or needing long-term non-invasive ventilation therapy;
         - Pulmonary hypertension (Pulmonary arterial systolic pressure >50 mmHg on echocardiogram or evidence of right ventricular

dysfunction);
- Worsening nutritional status particularly with BMI <18 kg/m^2 despite nutritional interventions;
- Recurrent massive hemoptysis despite bronchial artery embolization;
- World Health Organization (WHO) Functional Class IV;

c. Chronic obstructive pulmonary disease (COPD), and any of the following:
   i. BODE score (includes BMI, degree of airflow obstruction, degree of dyspnea, and exercise capacity) of 7-10;
   ii. FEV_1 (forced expiratory volume in 1 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:
   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 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;

e. Eisenmenger syndrome with pulmonary hypertension despite therapy aimed at avoiding polycythemia, iron deficiency and dehydration, and the associated profound hypoxemia and impaired quality of life;

f. Lymphangioleiomyomatosis (LAM) with evidence of disease progression despite mTOR inhibitor therapy and any of the following:
   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;

g. Primary lung graft failure or bronchiolitis obliterans;

2. *Pediatric members/enrollees, age < 18:*
   a. Cystic fibrosis, and any of the following:
      i. Progressive lung disease and disability despite optimal medical therapy;
      ii. FEV_1< 30% predicted;
      iii. Increasingly frequent hospitalizations;
      iv. Hypoxemia, (PaO_2 < 8 kPa or < 60 mm Hg);
      v. Hypercapnia, ($PaCO_2$ > 6.6 kPa or > 50 mmHg);
   b. Idiopathic pulmonary arterial hypertension, and any of the following:
      i. European Pediatric Pulmonary Vascular Disease Network (EPPVDN) high risk category and on optimal therapy without improvement;
      ii. Low exercise tolerance with 6MWT < 350 meters;
      iii. Uncontrolled syncope;
      iv. Hemoptysis;
      v. Right-sided heart failure;

vi. Failure to respond to vasodilator therapy;

c. Pulmonary vascular disease and failure to respond to medical management;
d. Eisenmenger syndrome with pulmonary hypertension despite therapy aimed at avoiding polycythemia, iron deficiency and dehydration, and the associated profound hypoxemia and impaired quality of life;
e. Surfactant dysfunction disorders with unrelenting respiratory failure, or progressive interstitial lung disease with respiratory insufficiency, unresponsive to medical interventions;
f. Bronchopulmonary dysplasia, and any of the following:
   i. Extended time requiring ventilator support without clinical improvement;
   ii. Pulmonary hypertension unresponsive to oxygen therapy;
   iii. Repeated episodes of respiratory failure without improvement in clinical trajectory over time, despite good medical support;
   iv. Progressive pulmonary hypertension;
g. Diffuse parenchymal lung disease, and any of the following:
   i. Disease progression despite optimal management;
   ii. Poor quality of life;
h. Primary lung graft failure or bronchiolitis obliterans.

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

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. 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 CF.

Coding Implications

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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

ICD-10-CM Diagnosis Codes that Support Coverage Criteria

ICD-10-CM Code	Description
C96.6	Unifocal Langerhans-cell histiocytosis
D86.0	Sarcoidosis of lung
E84.0-E84.9	Cystic fibrosis
E88.01	Alpha-1-antitrypsin deficiency
I27.0	Primary pulmonary hypertension
I27.83	Eisenmenger's syndrome
I27.89	Other specified pulmonary heart disease
I27.9	Pulmonary heart disease, unspecified
J41.8	Mixed simple and mucopurulent chronic bronchitis
J42	Unspecified chronic bronchitis
J43.0-J43.9	Emphysema
J44.0-J44.9	Other chronic obstructive pulmonary disease
J47.0-J47.9	Bronchiectasis
J60	Coal worker's Pneumoconiosis

ICD-10-CM Code	Description
J61	Pneumoconiosis due to asbestos and other mineral fibers
J62.0-J62.8	Pneumoconiosis due to dust containing silica
J63.0-J63.6	Pneumoconiosis due to other inorganic dusts
J84.10	Pulmonary fibrosis, unspecified
J84.111-J84.17	Idiopathic interstitial pneumonia
J84.81	Lymphangioleiomyomatosis
J84.82	Adult pulmonary Langerhans cell histiocytosis
J84.83	Surfactant mutations of the lung
J84.89	Other specified interstitial pulmonary disease
J98.2	Interstitial emphesema
J99	Respiratory disorders in diseases classified elsewhere
P27.0-P27.9	Chronic respiratory disease originating in the perinatal period
Q21.8	Other congenital malformations of cardiac septa
Q33.0-Q33.9	Congenital malformations of the lung
Z99.89	Dependence on other enabling machines and devices

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 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."	09/18	10/18
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	10/2021	

Reviews, Revisions, and Approvals	Revision Date	Approval Date
the revision log header to "Revision Date." Replaced "members" with "members/enrollees" in all instances. Annual review. References reviewed and updated. Reviewed by specialist.		
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	

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