Clinical Policy: Lung Transplantation

Reference Number: PA.CP.MP.57
Effective Date: 06/18
Last Review Date: 12/18

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 transplant for members with chronic, end-stage lung disease who have failed maximal medical therapy is medically necessary when all of 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 surviving at least 90 days after lung transplantation.

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

D. Does not have ANY of the following absolute contraindications:
   1. Malignancy in the past two years, except for non-melanoma localized skin cancer that has been treated appropriately;
   2. Untreatable significant dysfunction of another major organ system unless combined organ transplantation can be performed;
   3. Uncorrected atherosclerotic disease with suspected or confirmed end-organ ischemia or dysfunction and/or coronary artery disease not amenable to revascularization;
   4. Acute medical instability, including, but not limited to, acute sepsis, acute viral respiratory infection, myocardial infarction, and liver failure;
   5. Uncorrectable bleeding diathesis;
   6. Chronic infection with highly virulent and/or resistant microbes that are poorly controlled pre-transplant;
   7. Evidence of active *Mycobacterium tuberculosis* infection and/or smear-positive nontuberculous mycobacterial infection;
   8. Significant chest wall/spinal deformity expected to cause severe restriction after transplantation;
   9. Class II or III obesity (body mass index ≥ 35.0 kg/m²);
   10. Current non-adherence to medical therapy or a history of repeated or prolonged episodes of non-adherence to medical therapy that are perceived to increase the risk of non-adherence after transplantation;
   11. Psychiatric or psychological condition associated with the inability to cooperate or comply with medical therapy;
   12. Absence of an adequate or reliable social support system;
   13. Severely limited functional status with poor rehabilitation potential;
14. Substance abuse or dependence (including tobacco and alcohol) without appropriate risk reduction behaviors, such as meaningful and/or long-term participation in therapy for substance abuse and/or dependence; 
   a. Documentation of abstinence from smoking for 6 months before consideration to be eligible for transplant.

E. Has one of the following disease states and meets its corresponding criteria (not an all-inclusive list):

1. *Adult Members, Age ≥ 18.*
   a. Interstitial Lung Disease and any of the following:
      i. Decline in forced vital capacity (FVC) ≥ 10% during 6 months of follow-up (note: a 5% decline is associated with a poorer prognosis and may warrant listing);
      ii. Decline in diffusing capacity of the lung for carbon monoxide (DLCO) ≥15% during 6 months of follow-up;
      iii. Desaturation to < 88% or distance < 250 m on 6-minute-walk test (6MWT) or > 50 m decline in 6MWT distance over a 6-month period;
      iv. Pulmonary hypertension on right heart catheterization or 2-dimensional echocardiography;
      v. Hospitalization because of respiratory decline, pneumothorax, or acute exacerbation;
   b. Cystic fibrosis (CF) or other causes of bronchiectasis, and any of the following:
      i. Chronic respiratory failure and one of the following:
         a) With hypoxia alone (partial pressure of oxygen [PaO₂] < 8 kPa or < 60 mm Hg);
         b) With hypercapnia (partial pressure of carbon dioxide [PaCO₂] > 6.6 kPa or > 50 mmHg);
      ii. Long-term non-invasive ventilation therapy;
      iii. Pulmonary hypertension;
      iv. Frequent hospitalization with a clinical trajectory of worsening quality of life and lung function;
      v. Rapid lung function decline;
      vi. World Health Organization Functional Class IV.
   c. Chronic obstructive pulmonary disease (COPD), and any of the following:
      i. BODE index (includes BMI, degree of airflow obstruction, degree of dyspnea, and exercise capacity) ≥ 7;
      ii. FEV1 (forced expiratory volume in 1 second) < 15 to 20% of predicted;
      iii. Three or more severe exacerbations during the preceding year;
      iv. One severe exacerbation with acute hypercapnic respiratory failure;
      v. Moderate to severe pulmonary hypertension;
   d. Pulmonary vascular diseases and any of the following:
      i. New York Heart Association (NYHA) Functional Class III or IV despite a trial of at least 3 months of combination therapy including prostanoids;
      ii. Cardiac index of < 2 liters/min/m2;
      iii. Mean right atrial pressure > 15 mm Hg;
      iv. 6MWT of < 350 m;
v. Development of significant hemoptysis, pericardial effusion, or signs of progressive right heart failure (renal insufficiency, increasing bilirubin, brain natriuretic peptide, or recurrent ascites);
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 and any of the following:
   i. Severe impairment in lung function and exercise capacity (e.g., VO2 max <50% predicted);
   ii. Hypoxemia at rest;
g. Primary lung graft failure or bronchiolitis obliterans.

2. Pediatric Members, Age < 18
   a. Cystic fibrosis, and any of the following:
      i. Progressive lung disease and disability despite optimal medical therapy;
      ii. FEV1 < 30%;
      iii. Increasingly frequent hospitalizations;
      iv. Hypoxemia, (PaO₂] < 8 kPa or < 60 mm Hg);
      v. Hypercapnia, (partial pressure of carbon dioxide [PaCO₂ > 6.6 kPa or > 50 mmHg);
   b. Idiopathic pulmonary arterial hypertension, and any of the following:
      i. NYHA or WHO functional class III or IV despite vasodilator therapy;
      ii. Low exercise tolerance with 6MWT < 350 meters;
      iii. Uncontrolled syncope;
      iv. Hemoptysis;
      v. Right 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 impairing 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.
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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<table>
<thead>
<tr>
<th>CPT® Codes</th>
<th>Description</th>
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<tbody>
<tr>
<td>32851</td>
<td>Lung transplant, single; without cardiopulmonary bypass</td>
</tr>
<tr>
<td>32852</td>
<td>Lung transplant, single; with cardiopulmonary bypass</td>
</tr>
<tr>
<td>32853</td>
<td>Lung transplant, double (bilateral sequential or en bloc); without cardiopulmonary bypass</td>
</tr>
<tr>
<td>32854</td>
<td>Lung transplant, double (bilateral sequential or en bloc); with cardiopulmonary bypass</td>
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<table>
<thead>
<tr>
<th>HCPCS Codes</th>
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<tbody>
<tr>
<td>S2060</td>
<td>Lobar lung transplantation</td>
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<tr>
<td>S2152</td>
<td>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</td>
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ICD-10-CM Diagnosis Codes that Support Coverage Criteria

<table>
<thead>
<tr>
<th>ICD-10-CM Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>C96.6</td>
<td>Unifocal Langerhans-cell histiocytosis</td>
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<tr>
<td>D86.0</td>
<td>Sarcoidosis of lung</td>
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<tr>
<td>E84.0-E84.9</td>
<td>Cystic fibrosis</td>
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<tr>
<td>E88.01</td>
<td>Alpha-1-antitrypsin deficiency</td>
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<tr>
<td>I27.0</td>
<td>Primary pulmonary hypertension</td>
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<tr>
<td>I27.83</td>
<td>Eisenmenger’s syndrome</td>
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<tr>
<td>I27.89</td>
<td>Other specified pulmonary heart disease</td>
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<tr>
<td>I27.9</td>
<td>Pulmonary heart disease, unspecified</td>
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<tr>
<td>J41.8</td>
<td>Mixed simple and mucopurulent chronic bronchitis</td>
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<tr>
<td>J42</td>
<td>Unspecified chronic bronchitis</td>
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<tr>
<td>J43.0-J43.9</td>
<td>Emphysema</td>
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<tr>
<td>J44.0-J44.9</td>
<td>Other chronic obstructive pulmonary disease</td>
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<tr>
<td>J47.0-J47.9</td>
<td>Bronchiectasis</td>
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<tr>
<td>J60</td>
<td>Coal worker’s Pneumoconiosis</td>
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<td>J61</td>
<td>Pneumoconiosis due to asbestos and other mineral fibers</td>
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<tr>
<td>J62.0-J62.8</td>
<td>Pneumoconiosis due to dust containing silica</td>
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<tr>
<td>J63.0-J63.6</td>
<td>Pneumoconiosis due to other inorganic dusts</td>
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<tr>
<td>J84.10</td>
<td>Pulmonary fibrosis, unspecified</td>
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<td>J84.111-J84.17</td>
<td>Idiopathic interstitial pneumonia</td>
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<tr>
<td>J84.81</td>
<td>Lymphangioleiomyomatosis</td>
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<td>J84.82</td>
<td>Adult pulmonary Langerhans cell histiocytosis</td>
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<td>Surfactant mutations of the lung</td>
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<td>Other specified interstitial pulmonary disease</td>
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<tr>
<td>J98.2</td>
<td>Interstitial emphysema</td>
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<td>J99</td>
<td>Respiratory disorders in diseases classified elsewhere</td>
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<td>P27.0-P27.9</td>
<td>Chronic respiratory disease originating in the perinatal period</td>
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<tr>
<td>Q21.8</td>
<td>Other congenital malformations of cardiac septa</td>
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<tr>
<td>Q33.0-Q33.9</td>
<td>Congenital malformations of the lung</td>
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<tr>
<td>Z99.89</td>
<td>Dependence on other enabling machines and devices</td>
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Reviews, Revisions, and Approvals

<table>
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<tr>
<th>Date</th>
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<td>09/18</td>
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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.”

References reviewed and updated.

References


http://optn.transplant.hrsa.gov/ContentDocuments/OPTN_Policies.pdf#nameddest=Policy_10

http://dx.doi.org/10.1016/j.healun.2014.06.014