

Clinical Policy: Lung Transplantation

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[Coding Implications](#)
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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/Enrollees* 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, except for non-melanoma localized skin cancer that has been treated appropriately, low grade prostate cancer, a malignancy that has been completely resected, or a treated malignancy determined to have a small likelihood of recurrence and acceptable future risks;
 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 non-tuberculous mycobacterial infection;
 8. Significant chest wall/spinal deformity expected to cause severe restriction after transplantation;
 9. Inability to adhere to the regimen necessary to preserve the transplant, even with caregiver support;
 10. Class II or III obesity (body mass index $\geq 35.0 \text{ kg/m}^2$);
 11. Absence of an adequate or reliable social support system;
 12. 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

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eligible for transplant.

- E. Has one of the following disease states and meets its corresponding criteria (not an all-inclusive list):
1. *Adult Members/Enrollees, Age ≥ 18.* :
 - a. Interstitial Lung Disease and any of the following:
 - i. Decline in forced vital capacity (FVC) $\geq 10\%$ 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) $\geq 15\%$ 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 prostacyclins;
 - 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);

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- 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/Enrollees , 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

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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

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 2020, 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
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

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

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ICD-10-CM Code	Description
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
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 emphysema
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	12/2020	1/28/2021

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Reviews, Revisions, and Approvals	Revision Date	Approval Date
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.		
<p>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.</p> <p>Annual review. References reviewed and updated. Reviewed by specialist.</p>	10/2021	11/19/2021

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