

# Clinical Policy: Heart-Lung Transplant

Reference Number: PA.CP.MP.132

Effective Date: 01/18

Last Review Date: 06/01/2020

Coding Implications

Revision Log

## Description

Heart-lung transplantation is treatment of choice for patients with both end-stage heart and end-stage lung disease. This policy establishes the medical necessity requirements heart-lung transplants.

## Policy/Criteria

**I.** It is the policy of Pennsylvania Health and Wellness® that heart-lung transplant is **medically necessary** for members who meet all of the following guidelines:

**A.** End-stage heart and end-stage lung disease due to one of the following:

1. Age > 18 years and any of the following:
  - a. Irreversible primary pulmonary hypertension with heart failure;
  - b. Nonspecific severe pulmonary fibrosis;
  - c. Eisenmenger complex with irreversible pulmonary hypertension and heart failure;
  - d. Cystic fibrosis with severe heart failure;
  - e. Chronic obstructive pulmonary disease with heart failure;
  - f. Emphysema with severe heart failure;
  - g. Pulmonary fibrosis with uncontrollable pulmonary hypertension or heart failure;
  - h. Non-complex congenital heart disease associated with pulmonary hypertension that is not amenable to lung transplantation and repair by standard surgery;
  - i. Severe coronary artery disease or cardiomyopathy with irreversible pulmonary hypertension;
2. Age ≤ 18 years and any of the following:
  - a. Eisenmenger syndrome;
  - b. Heart re-transplant;
  - c. Alpha 1 antitrypsin deficiency;
  - d. Lung re-transplant;
  - e. Alveolar proteinosis;
  - f. Primary pulmonary hypertension;
  - g. Pulmonary vascular disease;
  - h. Restrictive cardiomyopathy;
  - i. Congenital heart disease;
  - j. Cystic fibrosis with progressive, irreversible cardiac dysfunction;
  - k. Dilated cardiomyopathy;

**B.** Meets the following disease severity criteria:

1. Meets one of the following staging criteria:
  - a. Age > 18 years: New York Heart Association classification of heart failure III or IV (Table 1); or
  - b. Age ≤ 18 years: American Heart Association Stage C or Stage D heart disease, (Table 2);
2. Adequate functional status with the ability for rehabilitation;
3. Life expectancy in the absence of cardiopulmonary disease ≥ 2 years;

- C. Does not have any of the following contraindications:
1. HIV and any of the following:
    - a. Active or prior opportunistic infections (progressive multifocal leukoencephalopathy or chronic intestinal cryptosporidiosis > 1 month);
    - b. Has not been clinically stable and compliant on combination antiretroviral therapy for > 3 months;
    - c. Detectable HIV RNA;
    - d. Has not had CD4 counts > 200 cells/ $\mu$ l for >3 months;
  2. History of psychological, behavioral, or cognitive disorders, poor family support structures, or documented noncompliance with previous therapies that could interfere with successful performance of care regimens after transplantation;
  3. Severe, irreversible disease in other organ systems or when it is part of a severe, irreversible, multisystemic disease process;
  4. Severe hypoplasia of the central branch pulmonary arteries or pulmonary veins;
  5. Any specific congenital heart lesion;
  6. Current episode of ongoing acute allograft rejection, even in the presence of graft vasculopathy, and retransplantation is requested;
  7. Less than 6 months have passed since the primary transplantation and retransplantation is requested;
  8. 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;
  9. Acute medical instability, including, but not limited to, acute sepsis, myocardial infarction, and liver failure;
  10. Uncorrectable bleeding diathesis;
  11. Chronic infection with highly virulent and/or resistant microbes that are poorly controlled pre-transplant;
  12. Evidence of active Mycobacterium tuberculosis infection;
  13. Significant chest wall/spinal deformity expected to cause severe restriction after transplantation;
  14. Class II or III obesity (body mass index  $\geq 35.0$  kg/m<sup>2</sup>);
  15. 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;
  16. Substance abuse or dependence (including tobacco and alcohol) 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.
  17. Active peptic ulcer disease.

<b>Table 1: NYHA Classifications of Heart Failure</b>	
<b>Classification</b>	<b>Characteristics</b>
<b>Class I</b>	Patients with cardiac disease but without the resulting limitations in physical activity. Ordinary activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain
<b>Class II</b>	Patients with heart disease resulting in slight limitations of physical activity. They are comfortable at rest. Ordinary physical activity results in fatigue, palpitation, dyspnea or anginal pain
<b>Class III</b>	Patients with cardiac disease resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary physical activity causes fatigue, palpitation, dyspnea, or anginal pain.
<b>Class IV</b>	Patients with cardiac disease resulting in inability to carry on any physical activity without discomfort. They symptoms of cardiac insufficiency or of the anginal syndrome may be present even at rest. If any physical activity is undertaken, discomfort increases.

<b>Table 2: Heart Failure Stages in Pediatric Heart Disease</b>	
<b>Classification</b>	<b>Characteristics</b>
<b>A</b>	At high risk for developing heart failure
<b>B</b>	Abnormal cardiac structure and/or function; no symptoms of heart failure
<b>C</b>	Abnormal cardiac structure and/or function; Past or present symptoms of heart failure
<b>D</b>	Abnormal structure and/or function; continuous infusion of intravenous inotropes or prostaglandin E <sub>1</sub> to maintain of a ductus arteriosus; mechanical ventilatory and/or mechanical circulatory support

\*Note: Heart lung transplantations may be considered medically necessary for other congenital cardiopulmonary anomalies as determined upon individual case review.

### **Background**

Heart-lung transplantation is a strong surgical option for selected patients with simultaneous end-stage heart failure and end-stage lung disease. However, due to a shortage of suitable donors, it is a rare procedure. Only about one hundred such transplants are performed each year in the USA. The 2016 International Society for Heart Lung Transplantation provides listing criteria and best practice recommendations for heart-lung transplants.<sup>1</sup>

The one and five-year survival rates are reported, respectively, at 59.1% and 88.2% for patients with hypertension, 26.8% and 70.4% for patients with hyperlipidemia, and 18% and 28.9% for patients with diabetes.<sup>2</sup>

Spahr et al discusses the pediatric indications and outcomes for heart-lung transplantations and reports that primary pulmonary hypertension, congenital heart disease, and Eisenmenger's syndrome, with a penetrance at 29%, 20% and 16%, respectively, are most common indications for heart lung transplants in children.<sup>5</sup> Since 1988, 188 pediatric heart lung transplants have been reported. Of these procedures, 16 have been performed at < 1 year of age, 52 procedures for

## CLINICAL POLICY

### Heart-Lung Transplant



patients 1-5 years of age, 28 procedures for patients 6-10 years of age, 92 procedures for patients 11-17 years of age.<sup>5</sup> Of note, outcomes for heart lung transplants are largely dependent on the success on the lung graft.<sup>5</sup>

#### 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
33930	Donor cardiectomy-pneumonectomy, with preparation and maintenance of allograft
33933	Backbench standard preparation of cadaver donor heart/lung allograft prior to transplantation, including dissection of allograft from surrounding soft tissues to prepare aorta, superior vena cava, inferior vena cava, and trachea for implantation
33935	Heart-lung transplant with recipient cardiectomy-pneumonectomy

HCPCS Codes	Description
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
D86.0-D86.89	Sarcoidosis
E84.0-E84.9	Cystic fibrosis
E88.01	Alpha-1-antitrypsin deficiency
I27.0-I27.9	Other pulmonary heart diseases
I42.0-I43	Cardiomyopathy
I50.84	End stage heart failure
J44.0-J44.9	Other chronic obstructive pulmonary disease
J47.0- J47.9	Bronchiectasis
J84.1Ø	Pulmonary fibrosis, unspecified
M32.9	Systemic lupus erythematosus (SLE)
Q33.0-Q33.9	Congenital malformations of lung

**CLINICAL POLICY**  
**Heart-Lung Transplant**



ICD-10-CM Code	Description
Q34.9	Congenital malformations of respiratory system, unspecified

Reviews, Revisions, and Approvals	Date	Approval Date
Corrected codes for bronchiectasis to be J47.0-J47.9	09/18	
Reworded contraindications regarding retransplantation with no change of meaning.	12/18	
Added contraindication of “Active peptic ulcer disease”. Clarified I.A.2.j, to state “Cystic fibrosis with progressive, irreversible cardiac dysfunction.” Removed the following contraindications: Severe, irreversible, fixed elevation of pulmonary vascular resistance; and Uncorrected atherosclerotic disease with suspected or confirmed end-organ ischemia or dysfunction and/or coronary artery disease not amenable to revascularization. Edited malignancy contraindication to not specify within 2 years, and added exceptions early stage prostate cancer, cancer that has been completely resected, or that has been treated and poses acceptable future risk. Added ICD-10-CM Codes – E88.01 and I50.84. Revised References reviewed and updated. Codes reviewed and updated. Specialist reviewed.	06/2020	08/2020

**References**

1. Mehra, Mandeep R., et al. "The 2016 International Society for Heart Lung Transplantation listing criteria for heart transplantation: a 10-year update." *J Heart Lung Transplant* 35.1 (2016): 1-23. Accessed March 12, 2020.
2. Spahr, Jonathan E., and Shawn C. West. "Heart-lung transplantation: pediatric indications and outcomes." *Journal of thoracic disease* 6.8 (2014): 1129. Accessed March 12, 2020.
3. Canter, Charles E., et al. "Indications for Heart Transplantation in Pediatric Heart Disease A Scientific Statement from the American Heart Association Council on Cardiovascular Disease in the Young; the Councils on Clinical Cardiology, Cardiovascular Nursing, and Cardiovascular Surgery and Anesthesia; and the Quality of Care and Outcomes Research Interdisciplinary Working Group." *Circulation* 115.5 (2007): 658-676.
4. Singer, LG. Heart-lung transplantation in adults. In: UpToDate, Hunt SA (Ed), UpToDate, Waltham, MA. Accessed March 12, 2020.
5. Weill D, Benden C, Corris PA, et al. A consensus document for the selection of lung transplant candidates:2014—An update from the Pulmonary Transplantation Council of the International Society for Heart and Lung Transplantation. *J Heart Lung Transplant*. 2015 Jan;34(1):1-15. doi: 10.1016/j.healun.2014.06.014. Epub 2014 Jun 26. Accessed March 12,2020.
6. Singh RK, Singh TP. Heart failure in children: Management. In: UpToDate, Triedman JK (Ed), UpToDate, Waltham, MA. Accessed on March 12, 2020.
7. Helderman JH, Goral S. *Journal of the American Society of Nephrology*. “Gastrointestinal Complications of Transplant Immunosuppression.” 2002 Jan; 13(1):277-87.
8. Vakil NM. Unusual causes of peptic ulcer disease. In: UpToDate, Feldman M (Ed), UpToDate, Waltham, MA. Accessed March 12, 2020.

**CLINICAL POLICY**  
**Heart-Lung Transplant**



9. Weill D. Lung transplantation: indications and contraindications. J Thorac Dis. 2018 Jul;10(7):4574-4587. doi: 10.21037/jtd.2018.06.141