Clinical Policy: Heart-Lung Transplant

Reference Number: PA.CP.MP.132
Effective Date: 01/2018
Date of Last Revision: 11/2023

Coding Implications
Revision Log

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

Policy/Criteria

I. It is the policy of Pennsylvania Health and Wellness® that heart-lung transplant is **medically necessary** for members/enrollees 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. Lung re-transplant;
      d. Alveolar proteinosis;
      e. Primary pulmonary hypertension;
      f. Pulmonary vascular disease;
      g. Restrictive cardiomyopathy;
      h. Congenital heart disease meeting one of the following:
         i. Congenital heart disease lesion that has been previously repaired or palliated;
         ii. Member/enrollee is an infant with a single functional ventricle and one of the following:
            a) Severe stenosis (stenoses) or atresia in proximal coronary arteries;
            b) Moderate to severe stenosis and/or insufficiency of the atrioventricular and/or systemic semilunar valve(s);
            c) Severe ventricular dysfunction;
      i. Cystic fibrosis with progressive, irreversible cardiac dysfunction;
      j. 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);
b. Age ≤ 18 years: American Heart Association Stage C or Stage D heart disease, (Table 2);

2. Life expectancy in the absence of cardiopulmonary disease ≥ two years;

C. Does not have any of the following contraindications:
1. HIV infection with detectable viral load;
2. Inability to adhere to the regimen necessary to preserve the transplant, even with caregiver support;
3. Severe hypoplasia of the central branch pulmonary arteries or pulmonary veins;
4. Current episode of ongoing acute allograft rejection, even in the presence of graft vasculopathy, and retransplantation is requested;
5. Less than 6 months have passed since the primary transplantation and retransplantation is requested;
6. Malignancy with high risk of recurrence or death related to cancer;
7. Acute renal failure with rising creatinine or on dialysis and low likelihood of recovery;
8. Acute liver failure or cirrhosis with portal hypertension or synthetic dysfunction;
9. Stroke, acute coronary syndrome, or myocardial infarction (excluding demand ischemia) within 30 days;
10. Glomerular filtration rate < 40 mL/min/1.73m^2, unless being considered for multi-organ transplant;
11. Septic shock;
12. Active extrapulmonary or disseminated infection;
13. Active *tuberculosis* infection;
14. Progressive cognitive impairment;
15. Other severe, uncontrolled medical condition expected to limit survival after transplant;
16. Active substance use or dependence (including current tobacco use, vaping, marijuana use (unless prescribed by a licensed practitioner), or intravenous drug use) without convincing evidence of risk reduction behaviors (unless urgent transplant timelines are present, in which case a commitment to reducing behaviors is acceptable). 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 ≥ six months prior to transplant;
17. Active peptic ulcer disease;
18. Lung transplantation alone will restore right ventricular function.

Table 1: NYHA Classifications of Heart Failure	
Classification	**Characteristics**
Class I	Patients with cardiac disease but without the resulting limitations in physical activity. Ordinary activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain
Class II	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
Class III	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.
Class IV	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.

Table 2: Heart Failure Stages in Pediatric Heart Disease	
Classification	**Characteristics**
A	At high risk for developing heart failure
B	Abnormal cardiac structure and/or function; no symptoms of heart failure
C	Abnormal cardiac structure and/or function; Past or present symptoms of heart failure
D	Abnormal structure and/or function; continuous infusion of intravenous inotropes or prostaglandin E_1 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. Complex congenital heart disease with Eisenmenger syndrome is the most common indication for heart-lung transplantation, with other common indications to include primary pulmonary hypertension and cystic fibrosis. The frequency of heart-lung transplantation is limited due to the number of suitable donors, while the need for heart-lung transplantation has declined due to the availability of new medical therapies.[4]

Contraindications for combined heart-lung transplantation are similar to those for isolated heart and lung transplantation.[4] The International Society for Heart Lung Transplantation (ISHLT) provides listing criteria and best practice recommendations for heart-lung transplants.[1,10]

According to the 2019 ISHLT registry report, survival rates in adult patients who underwent heart-lung transplantation has steadily improved with an overall median survival rate of 3.7 years

from 1992 through 2001 to 6.5 years from 2010 through 2017. This is comparable to primary lung transplantation but is inferior to the median survival rate of heart transplantation alone.[4]

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 2023, 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

Reviews, Revisions, and Approvals	Revision 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 –	06/2020	

Reviews, Revisions, and Approvals	Revision Date	Approval Date
E88.01 and I50.84. Revised References reviewed and updated. Codes reviewed and updated. Specialist reviewed.		
References reviewed and updated. Replaced all instances of "member" with "member/enrollee."	6/3/2021	
Annual review. In B.2., removed "adequate functional status with the ability for rehabilitation." Replaced contraindications of "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" and "current non-adherence to medical therapy…" with "Inability to adhere to the regimen necessary to preserve the transplant, even with caregiver support." Changed "Review Date" in policy header to "Date of Last Revision," and "Date" in the revision log header to "Revision Date." Updated 1.C. with some contraindications from ISHLT 2021 guidelines. Background updated with no clinical significance. Added specific congenital heart disease criteria to 2.i. Removed contraindication regarding specific congenital heart disease lesion. References reviewed, updated, and reformatted.	9/27/2022	
Annual review completed. Removed pediatric indication of Alpha- 1 antitrypsin deficiency. Added "Lung transplantation alone will restore right ventricular function" to I.C. Updated I.C.10. to include "unless being considered for multi-organ transplant". Criteria I.C.16. updated to exclude marijuana use when prescribed by a licensed practitioner and include required commitment to reducing substance use behaviors if urgent transplant timelines are present. ICD-10 diagnosis code table removed. Minor rewording with no clinical significance. References reviewed and updated. External specialists reviewed.	11/2023	

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