# Clinical Policy: Heart-Lung Transplant

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Effective Date: 01/2018
Date of Last Revision: 05/2025

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 PA Health and Wellness® that heart-lung transplant is **medically necessary** for members/enrollees who meet all of the following criteria:

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 severe heart failure;
   b. Nonspecific Idiopathic severe pulmonary fibrosis;
   c. Eisenmenger syndrome with irreversible pulmonary hypertension and heart failure;
   d. Cystic fibrosis with severe heart failure;
   e. Chronic obstructive pulmonary disease with severe heart failure;
   f. Emphysema with severe heart failure;
   g. Pulmonary fibrosis with uncontrollable pulmonary hypertension or heart failure;
   h. Congenital heart disease (CHD) meeting one of the following:
      i. Member/enrollee with single ventricle CHD and a Fontan circulation (total cardiopulmonary anastomosis) and one of the following:
         a) Symptomatic heart failure (HF) and reduced systolic function (Class 1);
         b) Symptomatic HF, preserved systolic function, and abnormal systemic ventricular filling pressures (Class 1);
         c) Lymphatic abnormalities including plastic bronchitis and protein-losing enteropathy refractory to lymphatic interventions and medical management (Class 2a);
         d) Cirrhosis or CKD attributed to chronically elevated central venous pressures (Class 2a);
      ii. Member/enrollee with single ventricle CHD and one of the following:
         a) Palliation to a shunted circulation or a superior cavo-pulmonary anastomosis (first procedure of a staged Fontan) and prohibitive risk for further single ventricle palliation;
         b) Cyanotic heart disease with severe atrio-ventricular valve regurgitation and prohibitive risk for operative repair;
         c) Pulmonary atresia with an intact ventricular septum, right ventricular dependent coronary circulation, and atresia of at least one aorto-coronary ostium;
      iii. HF symptoms or ventricular arrhythmias refractory to medical, interventional, and device therapies (Class 1);
      iv. Reactive pulmonary hypertension and a potential risk of developing fixed, irreversible elevation of PVR that could preclude heart transplant in the future (Class 1);
   i. Severe coronary artery disease or cardiomyopathy with irreversible pulmonary hypertension;
   j. Right ventricular failure with objective evidence of right ventricular fibrosis or infarction or refractory left ventricular failure;
2. Age < 18 years and any of the following:
   a. Eisenmenger syndrome;
   b. Heart re-transplant;
   c. Lung re-transplant;
   d. Pulmonary alveolar proteinosis;
   e. Primary pulmonary hypertension;

f. Pulmonary vascular disease;
g. Restrictive cardiomyopathy;
h. Congenital heart disease meeting one of the following:
    i. Member/enrollee with single ventricle CHD and a Fontan circulation (total cardiopulmonary anastomosis) and one of the following:
        a) Symptomatic HF and reduced systolic function (Class 1);
        b) Symptomatic HF, preserved systolic function, and abnormal systemic ventricular filling pressures (Class 1);
        c) Lymphatic abnormalities including plastic bronchitis and protein-losing enteropathy refractory to lymphatic interventions and medical management (Class 2a);
        d) Cirrhosis or CKD attributed to chronically elevated central venous pressures (Class 2a);
    ii. Member/enrollee with single ventricle CHD and one of the following:
        a) Palliation to a shunted circulation or a superior cavo-pulmonary anastomosis (first procedure of a staged Fontan) and prohibitive risk for further single ventricle palliation;
        b) Cyanotic heart disease with severe atrio-ventricular valve regurgitation and prohibitive risk for operative repair;
        c) Pulmonary atresia with an intact ventricular septum, right ventricular dependent coronary circulation, and atresia of at least one aorto-coronary ostium;
        d) Neonatal hypoplastic left heart syndrome with high-risk features including HF symptoms, ventricular dysfunction, left ventricular-coronary artery fistulae;
    iii. HF symptoms or ventricular arrhythmias refractory to medical, interventional, and device therapies (Class 1);
    iv. Reactive pulmonary hypertension and a potential risk of developing fixed, irreversible elevation of PVR that could preclude heart transplant in the future (Class 1);
    v. Neonatal cyanotic CHD with high-risk features (Class 2a);
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 unless all of the following are noted:
        a. CD4 cell count > 200 cells/mm$^3$ for at least 3 months before transplantation;
        b. Absence of active AIDS-defining opportunistic infection (unless treated efficaciously or prevented, can be included on the heart transplant waiting list) or malignancy;
        c. Member/enrollee is currently on effective antiretroviral therapy (ART);
        d. Member/enrollee does not have chronic wasting or severe malnutrition;
    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 six 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, unless being considered for multi-organ transplant;
    9. Stroke, acute coronary syndrome, or myocardial infarction (excluding demand ischemia) within 30

days;
10. Glomerular filtration rate < 30 mL/min/1.73m2, 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;
17. History of nicotine, tobacco, alcohol, or illicit drug use, without documentation noting abstinence from all (including nicotine replacement therapy) for ≥ six months prior to transplant;
18. Lung transplantation alone will restore right ventricular function.

**Table 1.** New York Heart Association (NYHA) Classifications of Heart Failure[1]

| 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.** American Heart Association (AHA) Heart Failure Stages[1]

| 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.[2]

Contraindications for combined heart-lung transplantation are similar to those for isolated heart and lung transplantation.[2] The International Society for Heart Lung Transplantation (ISHLT) provides listing criteria and best practice recommendations for heart transplants and for lung transplants.[3,4,5]

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.[2]

**Coding Implications**

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| 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 | 06/2020 | |

| Reviews, Revisions, and Approvals | Revision Date | Approval Date |
|---|---|---|
| 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. | | |
| 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 | |
| Annual review. Added indication to criteria I.A.1.j. Expanded criteria I.C.1. to I.C.1.a. through c. Removed contraindication I.C.17., active peptic ulcer disease. References reviewed and updated. | 03/2024 | 04/2025 |
| Annual review. Changed I.A.1. to ≥ 18. I.A.1.a now reflects "severe" heart failure and I.A.1.b. now reflects "nonspecific idiopathic". In I.A.1.h., "non-complex congenital…standard surgery" was removed, and now reflects "Congenital heart disease…", adding I.A.1.h.i – iv., followed by i. and j. Age changed to < 18 in I.A.2 and in I.A.2.d, added "pulmonary" Changes made to I.A.2.h in addition to adding h.i.-v. Ages changed in I.B.1.a. "> 18" and I.B.1.b. "≤ 18". In I.C.1.a, added "for at least | 05/2025 | |

| Reviews, Revisions, and Approvals | Revision Date | Approval Date |
|---|---|---|
| …transplantation". Added I.C.1.d "Member/enrollee…severe malnutrition". In I.C.8., added "unless being…transplant". Reworded I.C.16.a. now reflecting "alcohol or illicit drug use". Table 2 has been changed to the American Heart Association Heart Failure Stages, removing Heart Failure Stages in Pediatric Heart Disease. Background reviewed and updated. Coding verified. Updated criteria I.A.1.h.iv. and I.A.2.h.iv. from, "…could preclude heart failure in the future…" to "…could preclude heart transplant in the future..." Internal and external specialist reviewed. References reviewed and updated | | |

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