Clinical Policy: Heart-Lung Transplant

Reference Number: LA.CP.MP.132 Coding Implications
Date of Last Revision: ~~2/22~~5/22 Revision Log

See Important Reminder at the end of this policy for important regulatory and legal information.

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 Louisiana Healthcare Connections that heart-lung transplant is **medically necessary** for member/enrollee 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. Life expectancy in the absence of cardiopulmonary disease ≥ 2 years;

C. Does not have any of the following contraindications:
   1. HIV infection with detectable viral load;
   ~~1.~~2.~~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/µl for >3 months;~~
   ~~2.~~3. Inability to adhere to the regimen necessary to preserve the transplant, even with caregiver support;
   ~~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 with high risk of recurrence or death related to cancer;
   ~~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 renal failure with rising creatinine or on dialysis and low likelihood of recovery;
   10. Acute liver failure or cirrhosis with portal hypertension or synthetic dysfunction;
   11. Stroke, acute coronary syndrome, or myocardial infarction (excluding demand ischemia) within 30 days;
   12. Glomerular filtration rate < 40 mL/min/1.73m^2;
   13. Septic shock;
   14. Active extrapulmonary or disseminated infection;
   15. Active *tuberculosis* infection;
   16. Progressive cognitive impairment;
   17. Other severe, uncontrolled medical condition expected to limit survival after transplant;
   18. Active substance use or dependence (including current tobacco use, vaping, marijuana smoking, or intravenous drug use) 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.
      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 ≥ 6 months prior to transplant.
   19. Active peptic ulcer disease.

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

~~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.[1]~~

~~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.[2]~~

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

Coding Implications

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CPT® Codes	Description
33930	Donor cardiectomy-pneumonectomy, (including cold preservation)
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
J84.10	Pulmonary fibrosis, unspecified
M32.9	Systemic lupus erythematosus (SLE), unspecified
Q33.0-Q33.9	Congenital malformations of lung
Q34.9	Congenital malformations of respiratory system, unspecified

Reviews, Revisions, and Approvals	Revision Date	Approval Date
Converted corporate to local policy.	10/2020	
References reviewed and updated. Replaced all instances of "member" with "member/enrollee." In B.2., removed "adequate functional status with the ability for rehabilitation." Replaced contraindications of "history 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." Added "may not support medical necessity" in Coding Implications.	2/22	2/22
Annual review. References reviewed, updated, and reformatted. Updated 1.C. with some contraindications from ISHLT 2021 guidelines. Background updated with no clinical significance. Added and may not support medical necessity to Coding Implications section	5/22	

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Research Interdisciplinary Working Group [published correction appears in Circulation. 2007 Apr 3;115(13):e385. Friedman, Allen H [corrected to Friedman, Alan H]]. *Circulation*. 2007;115(5):658-676. doi:10.1161/CIRCULATIONAHA.106.180449

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Important Reminder

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. LHCC makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved.

The purpose of this clinical policy is to provide a guide to medical necessity, which is a component of the guidelines used to assist in making coverage decisions and administering benefits. It does not constitute a contract or guarantee regarding payment or results. Coverage decisions and the administration of benefits are subject to all terms, conditions, exclusions and limitations of the coverage documents (e.g., evidence of coverage, certificate of coverage, policy, contract of insurance, etc.), as well as to state and federal requirements and applicable LHCC administrative policies and procedures.

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This clinical policy does not constitute medical advice, medical treatment or medical care. It is not intended to dictate to providers how to practice medicine. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of member/enrollee. This clinical policy is not intended to recommend treatment for member/enrollee. Member/enrollee should consult with their treating physician in connection with diagnosis and treatment decisions.

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herein through the terms of their contracts. Where no such contract exists, providers, member/enrollee and their representatives agree to be bound by such terms and conditions by providing services to member/enrollee and/or submitting claims for payment for such services.