Clinical Policy: Heart-Lung Transplant

Reference Number: NC.CP.MP.132
Date of Last Revision: ~~04/24~~

Coding Implications
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 Carolina Complete Health that heart-lung transplant is **medically necessary** for members who meet all 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 severe heart failure;
   b. Nonspecific idiopathic 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. Congenital heart disease (CHD) meeting one of the following: ~~Non-complex congenital heart disease associated with pulmonary hypertension that is not amenable to lung transplantation and repair by standard surgery;~~
      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);
         ~~a)~~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);

~~i.~~iv. Reactive pulmonary hypertension and a potential risk of developing fixed, irreversible elevation of PVR that could preclude heart failure 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;

~~a. Congenital heart disease lesion been previously repaired or palliated;~~

~~b. Member/enrollee is an infant with a single functional ventricle and one of the following:~~

~~i. Severe stenosis (stenoses) or atresia in proximal coronary arteries;~~

~~ii. Moderate to severe stenosis and/or insufficiency of the atrioventricular and/or systemic semilunar valve(s);~~

~~iii. Severe ventricular dysfunction;~~

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 failure in the future (Class 1);
v. Neonatal cyanotic CHD with high-risk features (Class 2a);

i. Cystic fibrosis;
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: 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 except where optimal management can be demonstrated by a physician with generally recognized expertise in HIV care;
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 re-transplantation is requested;
5. Less than 6 months have passed since the primary transplantation and re-transplantation is requested;
6. Active, potentially life-threatening, malignancy (except when transplant is done for a cure);
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 < 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 h~~History of nicotine ~~or~~ tobacco, alcohol, or illicit drug use, without documentation noting ~~notes~~ abstinence from all ~~tobacco and nicotine products~~ (including nicotine replacement therapy) for ≥ 6 months prior to transplant.
17. 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~~ American Heart Association (AHA) Heart Failure Stages[1]	
Classification	**Characteristics**
A	~~At high risk for developing heart failure~~ Patients at high risk for heart failure but does not yet have symptoms or structural or functional heart disease.
B	~~Abnormal cardiac structure and/or function; no symptoms of heart failure~~ Patients with no current or previous symptoms of heart failure but have structural heart disease, increased filling pressures in the heart, or other risk factors.
C	~~Abnormal cardiac structure and/or function; Past or present symptoms of heart failure~~ Patients with structural heart disease with current or previous 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~~ Patients who have heart failure with symptoms that interfere with daily life functions or result in recurrent hospitalizations despite continued guideline-directed medical therapy.

*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

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 2021, 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, 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

Reviews, Revisions, and Approvals	Revision Date	Approval Date
Policy adapted for use by North Carolina Health Plan (Carolina Complete Health), per state feedback and requirements: removed the following contraindications: active peptic ulcer disease and chest wall/spinal deformity causing restriction; replaced contraindication of malignancy within past 2 years with "active, potentially life-threatening malignancy."	06/19	06/19
Annual review completed. No changes to policy content.	03/21	03/21
In B.2., removed "adequate functional status with the ability for rehabilitation." In section C, 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."	08/21	11/21
Annual review. References reviewed, updated, and reformatted. Updated 1.C. with some contraindications from ISHLT 2021 guidelines. Background updated with no clinical significance.	05/22	05/22
Added specific congenital heart disease criteria to 2.i. Removed contraindication regarding specific congenital heart disease lesion.	08/22	08/22
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	04/23	04/23

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.		
Annual review. Added indication to criteria I.A.1.j. Revised criterion C.1.References reviewed and updated.	04/24	04/24
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.8., added "unless being…transplant". Reworded I.C.16.a. to include "alcohol and illicit drug use without documentation noting abstinence...". 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.		

References

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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. The Health Plan 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. "Health Plan" means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan's affiliates, as applicable.

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 Health Plan-level administrative policies and procedures.

This clinical policy is effective as of the date determined by the Health Plan. The date of posting may not be the effective date of this clinical policy. This clinical policy may be subject to applicable legal and regulatory requirements relating to provider notification. If there is a discrepancy between the effective date of this clinical policy and any applicable legal or regulatory requirement, the requirements of law and regulation shall govern. The Health Plan retains the right to change, amend or withdraw this clinical policy, and additional clinical policies may be developed and adopted as needed, at any time.

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 members. This clinical policy is not intended to recommend treatment for members. Members should consult with their treating physician in connection with diagnosis and treatment decisions.

Providers referred to in this clinical policy are independent contractors who exercise independent judgment and over whom the Health Plan has no control or right of control. Providers are not agents or employees of the Health Plan.

Note: For Medicaid members, when state Medicaid coverage provisions conflict with the coverage provisions in this clinical policy, state Medicaid coverage provisions take precedence.

Please refer to the state Medicaid manual for any coverage provisions pertaining to this clinical policy.

Note: For Medicare members, to ensure consistency with the Medicare National Coverage Determinations (NCD) and Local Coverage Determinations (LCD), all applicable NCDs, LCDs, and Medicare Coverage Articles should be reviewed prior to applying the criteria set forth in this clinical policy. Refer to the CMS website at http://www.cms.gov for additional information.