Anthem

# Medical Policy

**Subject:** Heart/Lung Transplantation
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## Description/Scope

This document addresses heart and lung (heart/lung) transplantation criteria for individuals who have both cardiac (heart) and lung disease. A heart/lung transplant refers to the harvesting of one or both lungs and the heart from a single cadaver donor, which is then implanted into a single recipient in a coordinated surgical procedure.

**Note:** Please see the following related documents for additional information:

- TRANS.00009 Lung and Lobar Transplantation
- TRANS.00033 Heart Transplantation

## Position Statement

**Medically Necessary:**

Heart/lung transplantation is considered **medically necessary** when the following clinical indications **and** the general individual selection criteria listed below are met.

**Clinical Indications -** Individuals must meet **one** of the following criteria:

1. Irreversible primary pulmonary hypertension with heart failure;**or**
2. Secondary pulmonary hypertension and resulting heart failure due to pulmonary fibrosis, cystic fibrosis, chronic obstructive pulmonary disease or emphysema; **or**
3. Eisenmenger's complex or other types of congenital heart disease with irreversible pulmonary hypertension and heart failure.

**Investigational and Not Medically Necessary:**

A heart/lung transplantation is considered **investigational and not medically necessary** when the above criteria are not met.

**Note:** For multi-organ transplant requests, criteria must be met for each organ requested. In those situations, an individual may present with concurrent medical conditions which would be considered an exclusion or a comorbidity that would preclude a successful outcome, but would be treated with the other organ transplant. Such cases will be reviewed on an individual basis for coverage determination to assess the member's candidacy for transplantation.

**General Individual Selection Criteria**

In addition to having one of the clinical indications above, the member must not have a contraindication, as defined by the American Society of Transplantation in *Guidelines for the Referral and Management of Patients Eligible for Solid Organ Transplantation*(2001) listed below.

**Absolute Contraindications – for Transplant *Recipients**** include, but are not limited to, the following:

A. Metastatic cancer;
B. Ongoing or recurring infections that are not effectively treated;
C. Serious cardiac or other ongoing insufficiencies that create an inability to tolerate transplant surgery;
D. Serious conditions that are unlikely to be improved by transplantation as life expectancy can be finitely measured;
E. Active, systemic lupus erythematosus or sarcoid with multisystem involvement;
F. Any systemic condition with a high probability of recurrence in the transplanted heart;
G. Demonstrated patient noncompliance, which places the organ at risk by not adhering to medical recommendations;
H. Potential complications from immunosuppressive medications are unacceptable to the patient;
I. Acquired immune deficiency syndrome (AIDS) (diagnosis based on Centers for Disease Control and Prevention [CDC] definition of CD4 count, 200cells/$mm^3$) unless the following are noted:
    1. CD4 count greater than 200 cells/$mm^3$ for greater than 6 months;
    2. HIV-1 RNA undetectable;
    3. On stable anti-retroviral therapy greater than 3 months;
    4. No other complications from AIDS (for example, opportunistic infection, including aspergillus, tuberculosis, coccidioidomycosis, resistant fungal infections, Kaposi's sarcoma or other neoplasm);
    5. Meeting all other criteria for heart-lung transplantation.

*Steinman, Theodore, et al. Guidelines for the Referral and Management of Patients Eligible for Solid Organ Transplantation. Transplantation. 2022; 71(9):1189-1204.

## Rationale

Heart/lung transplantation is a standard treatment for individuals who meet the Medical Necessity criteria listed in this document. Combined heart/lung transplants are reserved for candidates in whom either a heart transplant or a lung transplant alone would not improve the individual's condition and chances of survival.

The Pulmonary Transplantation Council of the International Society for Heart and Lung Transplantation issued a 2021 updated consensus document for the selection of heart-lung transplant candidates. The authors (Leard and colleagues, 2021) include the following information:

> Candidates should meet the criteria for lung transplant listing and have significant dysfunction of one or more additional organs, or meet the listing criteria for a non-pulmonary organ transplant and have significant pulmonary

dysfunction.

The primary indication for heart-lung transplantation is pulmonary hypertension, either secondary to idiopathic pulmonary arterial hypertension or congenital heart disease (CHD). Criteria for heart-lung transplant listing described in a previous version of this document include the presence of New York Heart Association (NYHA) functional class IV symptoms despite maximal medical management, a cardiac index below 2 l/min/$m^2$, and a mean right atrial pressure above 15 mmHg; however, the decision about whether to list a patient for heart-lung transplant remains difficult.

Heart-lung and other multi-organ transplantation should be limited to centers with experience in such procedures and where specialists are available to manage each of the transplanted organs.

In 2018, the American Heart Association/American College of Cardiology published guidelines for the management of adults with congenital heart disease (CHD). Typically, CHD is treated with simultaneous heart-lung transplantation for conditions that result in irreversible pulmonary hypertension such as Eisenmenger syndrome (Stout, 2019).

Valapour and colleagues, (2021) reported in the Organ Procurement Transplant Network (OPTN)/ Scientific Registry of Transplant Recipients that lung transplants declined in 2020, this coincided with the COVID-19 pandemic. The number of candidates added to the waiting list increased after a decline in 2020, the waitlist mortality increased with a decreased number of transplants, time to transplant improved with 38.0% of candidates waiting fewer than 90 days for a transplant, post-transplant 85.3% of recipients are surviving to 1 year; 67%, to 3 years; and 54.3%, to 5 years.

In September 2022, United Network of Organ Sharing (UNOS) reported 31 candidates currently on the active waiting list for heart-lung transplant. A total of 45 combined heart-lung transplants occurred in 2021, 2 of the heart-lung transplants were performed in children. According to the 2020 Annual Report of the U.S. Organ Procurement and Transplantation Network (OPRN) and the Scientific Registry of Transplant Recipients, the survival rates reported from transplants performed from 2008-2015 at 1, 3, and 5 years post-transplant were 81%, 58.3%, and 50.2%.

Potential lung transplant recipients were ranked according to the lung allocation score (LAS). In 2021, the OPTN Lung Transplant Committee updated how lung transplants were allocated in the United States. UNOS made changes to the updated cohort for calculation of the LAS approved by the OPTN Board of Directors, the update improved the predication of candidates' expected survival on the waitlist and post-transplant to improve equity in lung allocation. In 2023, the OPTN published a new policy for matching lung transplant candidates with organs from deceased donors. The new "continuous distribution" methodology states that all of the factors in the organ match are included in a single weighted score, calculated for each lung transplant candidate, and each potential lung from a donor. This new document states:

> Statistical modeling suggests this will reduce the number of lung candidates who die awaiting a lung transplant, but it will increase transplant access for a number of candidates. This includes candidates who are:
>
> - The most medically urgent
> - Younger than age 18
> - A prior living organ donor
> - More likely to have immune system rejection of many organs
> - Short in stature
> - Expected to live longer after a transplant

All lung transplant candidates aged 12 and older will receive a lung Composite Allocation Score (lung CAS) that replaces the LAS. For candidates younger than age 12, the two existing priority rankings will still be utilized. With the CAS, candidates receive varying numbers of points based on a set of different attributes. The attributes are weighted. The categories of attributes include the following:

- Candidate medical urgency (maximum 25 points)
- Likelihood of recipient survival over five years post-transplant (maximum 25 points)
- Potential biological challenges in matching, such as the candidate's blood type, height or immune sensitivity (maximum 15 points)
- Whether the candidate was younger than age 18 when listed for a transplant (20 points)
- Whether the candidate was a prior living organ donor (five points)
- A final category of attributes, worth as many as 10 points, is determined by each lung offer from a donor who is a potential match for the candidate. More points will be assigned to matches where the donor hospital and the candidates' transplant hospital are closer to one another, and where the logistics of preserving and transporting the lungs between the two hospitals are more likely to result in a successful transplant. Because this portion of the lung CAS may be different for every organ offer, the total score will not remain the same for all patients and all matches.

In March 2023, the Organ Procurement and Transplantation Network (OPTN) also changed the policy to expand the eligibility to pediatric heart status 1A and 1B candidates, who are registered on the waiting list prior to turning age 2, to accept incompatible blood type (ABOi) donors.

In recent years the discussion of heart transplantation as a treatment for cardiomyopathy caused by amyloidosis has gathered interest. Cardiac amyloidosis causes restrictive cardiomyopathy, progressive heart failure, and death. Heart transplantation outcomes have been poorer in individuals with cardiac amyloidosis due to the systemic nature of the condition. Light chain suppressive therapies for light chain amyloidosis (AL) may improve outcomes. Barrett and colleagues (2020) reported a retrospective (2004-2017), single center study of heart transplantation outcomes in 31 individuals with cardiac amyloidosis (13 with AL, and 18 with transthyretin [ATTR] amyloidosis). Screening included evaluation for extracardiac amyloidosis with organ specific biopsies, and extracardiac amyloid deposits constituted an absolute contraindication. In individuals with vascular or mucosal amyloid positive gastrointestinal biopsy, transplantation candidacy was evaluated by clinical symptoms. Individuals were considered for transplantation if there was no evidence of dysmotility, bleeding, or malabsorption. Pre-transplantation response to light chain reducing therapies in AL amyloidosis was not an absolute requirement for transplantation. Post-transplantation immunosuppression regimens were followed. Autologous stem cell transplants were performed in select participants with AL amyloidosis once they achieved clinical stability following the transplant, but no sooner than 6 months post-heart transplantation. The results demonstrated that post-transplantation outcomes were similar between the AL and ATTR groups, including post-transplantation infection and organ rejection. Two participants had difficulty with post-transplantation hemostasis, and 5 developed post-transplantation renal dysfunction that required renal replacement therapy. One participant with gastrointestinal amyloid deposits developed a post-operative ileus, which resolved. Of the 2 participants with renal amyloidosis, 1 developed progressive renal dysfunction that required dialysis 7 years after transplantation. There were 4 deaths after transplantation, but none determined to be due to amyloidosis or amyloidosis therapy complications. There were no differences in mortality between the 31 participants and the 599 individuals without amyloidosis. The authors concluded that in carefully selected individuals with AL and ATTR cardiac amyloidosis, the results were similar in both post-transplantation outcomes

and survival compared to individuals without amyloidosis who received transplantation for all other conditions. These results are promising; however, the study was limited by its small size and single center transplant practices. Treatment practices and transplant eligibility policies can vary between institutions and could lead to different results, therefore larger studies are needed to confirm the outcomes.

In 2023 the ACC published the "Expert Consensus Decision Pathway on Comprehensive Multidisciplinary Care for the Patient With Cardiac Amyloidosis." The consensus supports heart transplant in some individuals with cardiac amyloidosis:

> In select patients with ATTR-CM and AL-CM with advanced/stage D HF, heart transplantation may be an option, and the current adult donor allocation system provides priority as Status 4 to amyloid CM, given the lack of durable (MCS) support options. The traditional signs of advanced HF apply in patients with cardiac amyloidosis and should be recognized as triggers for a discussion of prognosis and advanced HF therapies that incorporates the patient's goals, values, and preferences.

The ACC acknowledges that no single test can identify which individuals may be candidates for advanced HF therapies; multiple factors including the history, physical examination, laboratory testing, and imaging studies must be considered together to identify those at greatest risk of future decompensation and those who will derive the greatest benefit.

## Background/Overview

Combined heart-lung transplantation is intended to prolong survival and improve function in individuals with end-stage cardiopulmonary or pulmonary disease. The technique involves a coordinated triple operative procedure, consisting of procurement of a single donor heart-lung block, excision of the heart and lungs of the recipient, and implantation of the new donor heart and lungs into the recipient.

The limiting factor for heart-lung transplantation is the short supply of donor organs. The procurement and distribution of heart-lung organs for transplantations in the United States is under the discretion of the United Network for Organ Sharing (UNOS). A national database of transplant candidates, donors, recipients, and donor-recipient matching and histocompatibility is maintained by UNOS.

Heart-lung transplantation remains a treatment option for carefully selected individuals with end-stage heart and lung diseases.

## Definitions

End-stage heart failure: In people with heart failure, the body does not receive an adequate supply of oxygen. As a result, they can feel weak, fatigued or short of breath. Everyday activities such as walking, climbing stairs, carrying groceries and yard work can become quite difficult. In end-stage heart failure, the heart is so weakened the individual will die without a heart transplant.

Heart/Lung transplant: Removal of an individual's heart and lungs and replacing it with a heart and lungs from a single donor.

New York Heart Association (NYHA) definitions:

- Class III. Individuals with cardiac disease resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity causes fatigue, palpitation, dyspnea (difficulty breathing) or anginal (chest) pain.
- Class IV. Individuals with cardiac disease resulting in inability to carry on any physical activity without discomfort and/ or dyspnea. Symptoms of heart failure or anginal (chest) pain may be present even at rest. If any physical activity is undertaken, cardiac symptoms are increased.

## Coding

*The following codes for treatments and procedures applicable to this document are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Please refer to the member's contract benefits in effect at the time for service to determine coverage or non-coverage of these services as it applies to an individual member.*

**When services may be Medically Necessary, when criteria are met:**

| | |
|---|---|
| **CPT** | |
| 00580 | Anesthesia for heart transplant or heart/lung transplant |
| 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 |
| **ICD-10 Procedure** | |
| 02YA0Z0 | Transplantation of heart, allogeneic, open approach |
| 02YA0Z1 | Transplantation of heart, syngeneic, open approach |
| 0BYM0Z0 | Transplantation of bilateral lungs, allogeneic, open approach |
| 0BYM0Z1 | Transplantation of bilateral lungs, syngeneic, open approach |
| **ICD-10 Diagnosis** | |
| | All diagnoses |

**When services are Investigational and Not Medically Necessary:**
For the procedure codes listed above when criteria are not met, or when the code describes a procedure indicated in the Position Statement section as investigational and not medically necessary.

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**Peer Reviewed Publications:**


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JACC Heart Fail. 2020; 8(6):461-468.
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**Government Agency, Medical Society, and Other Authoritative Publications:**

1. Hunt SA, Abraham WT, Chin MH, et al. 2009 focused update incorporated into the ACC/AHA 2005 Guidelines for the Diagnosis and Management of Chronic Heart Failure in the Adults: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. Circulation. 2009; 119(14):e391-e479.
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## Websites for Additional Information

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## Index



## Document History

| Status | Date | Action |
|---|---|---|
| Revised | 11/09/2023 | Medical Policy & Technology Assessment Committee review (MPTAC). Revised reference in Position Statement section. Updated Description, Background/Overview, References, and Website sections. |
| Reviewed | 11/10/2022 | MPTAC review. Updated Rationale, Background, References and Websites sections. |
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| Reviewed | 11/07/2019 | MPTAC review. Updated Rationale, References and Websites sections. |

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|---|---|---|
| Reviewed | 01/24/2019 | MPTAC review. Updated References and Websites sections. |
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| Reviewed | 02/02/2017 | MPTAC review. Updated formatting in Position Statement section. Updated References and Websites sections. |
| Revised | 02/04/2016 | MPTAC review. Reformatted investigational and not medically statement. Defined abbreviation in investigational and not medically necessary absolute contraindication section. Updated Rationale, Background and References sections. Removed ICD-9 codes from Coding section. |
| Reviewed | 02/05/2015 | MPTAC review. Updated Description, Rationale, References and Websites sections. |
| Reviewed | 02/13/2014 | MPTAC review. Updated Description, Definitions, References and Web Sites sections. |
| Reviewed | 02/14/2013 | MPTAC review. Rationale, Definitions, and Websites Updated. |
| Reviewed | 02/16/2012 | MPTAC review. Updated Rationale, References and Websites |
| Reviewed | 02/17/2011 | Medical Policy & Technology Assessment Committee (MPTAC) review. Updated Rationale, References and Websites. |
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| Pre-Merger Organizations | Last Review Date | Document Number | Title |
|---|---|---|---|
| Anthem, Inc. | 11/15/2001 | TRANS.00006H Archived | Heart/Lung Transplant |
| WellPoint Health Networks, Inc. | 09/23/2004 | 7.04.01 | Heart/Lung Transplantation |

Applicable to Commercial HMO members in California: When a medical policy states a procedure or treatment is investigational, PMGs should not approve or deny the request. Instead, please fax the request to Anthem Blue Cross Grievance and Appeals at fax # 818-234-2767 or 818-234-3824. For questions, call G&A at 1-800-365-0609 and ask to speak with the Investigational Review Nurse.

Federal and State law, as well as contract language, including definitions and specific contract provisions/exclusions, take precedence over Medical Policy and must be considered first in determining eligibility for coverage. The member's contract benefits in effect on the date that services are rendered must be used. Medical Policy, which addresses medical efficacy, should be considered before utilizing medical opinion in adjudication. Medical technology is constantly evolving, and we reserve the right to review and update Medical Policy periodically.