Heart-Lung Transplant

Policy
Blue Cross and Blue Shield of Kansas City (Blue KC) will provide coverage for a heart-lung transplant when it is determined to be medically necessary because the criteria shown below are met.

When Policy Topic is covered
Heart/lung transplantation may be considered medically necessary for carefully selected patients with end-stage cardiac and pulmonary disease including, but not limited to, one of the following diagnoses:
- irreversible primary pulmonary hypertension with heart failure;
- non-specific severe pulmonary fibrosis, with severe heart failure;
- Eisenmenger complex with irreversible pulmonary hypertension and heart failure;
- cystic fibrosis with severe heart failure;
- chronic obstructive pulmonary disease with heart failure;
- emphysema with severe heart failure;
- pulmonary fibrosis with uncontrollable pulmonary hypertension or heart failure.

Heart/lung retransplantation after a failed primary heart/lung transplant may be considered medically necessary in patients who meet criteria for heart/lung transplantation.

When Policy Topic is not covered
Heart/lung transplantation is considered investigational in all other situations.

Considerations
Heart-lung transplants should be considered for coverage under the Transplant Benefit.

Potential contraindications subject to the judgment of the transplant center:
1. Known current malignancy, including metastatic cancer
2. Recent malignancy with high risk of recurrence
3. Untreated systemic infection making immunosuppression unsafe, including chronic infection
4. Other irreversible end-stage disease not attributed to heart or lung disease
5. History of cancer with a moderate risk of recurrence
6. Systemic disease that could be exacerbated by immunosuppression
7. Psychosocial conditions or chemical dependency affecting ability to adhere to therapy

General
When the candidate is eligible to receive a heart in accordance with United Network for Organ Sharing (UNOS) guidelines for cardiac transplantation, the lung(s) shall be allocated to the heart-lung candidate from the same donor. When the candidate is eligible to receive a lung in accordance with the UNOS Lung Allocation System (LAS), the heart shall be allocated to the heart-lung candidate from the same donor if no suitable Status 1A isolated heart candidates are eligible to receive the heart. Status 1A is described below. (3)

Cardiac Specific
The United Network for Organ Sharing (UNOS) prioritizes donor thoracic organs according to the severity of illness as follows:

**Status 1A**
A patient is admitted to the listing transplant center hospital and has at least 1 of the following devices or therapies in place:

- Mechanical circulatory support for acute hemodynamic decompensation that includes at least 1 of the following:
  - Left and/or right ventricular assist device implanted
  - Total artificial heart
  - Intra-aortic balloon pump: or
  - Extracorporeal membrane oxygenator (ECMO)
- Mechanical circulatory support
- Mechanical ventilation
- Continuous infusion of inotropes and continuous monitoring of left ventricular filling pressures
- If criteria a, b, c, and d are not met, such status can be obtained by application to the applicable Regional Review Board

**Status 1B**
A patient has at least 1 of the following devices or therapies in place:

1. left and/or right ventricular device implanted, or
2. continuous infusion of intravenous inotropes

A patient that does not meet Status 1A or 1B is listed as Status 2.

Status 7 patients are considered temporarily unsuitable to receive a thoracic organ transplant.

**Transplant Benefit**
The date on which the Transplant Benefit starts accumulating is determined by the transplant coordinator. The Transplant Benefit ends when the Transplant Lifetime Maximum benefit (if applicable) has been exhausted.

Benefits include:
- hospitalization of the recipient for medically recognized transplants from a donor to a transplant recipient;
- evaluation tests requiring hospitalization to determine the suitability of both potential (member's benefits must be verified with regard to the potential donor who does not turn out to be the actual donor) and actual donors, when such tests cannot be safely and effectively performed on an outpatient basis (Note: The member's benefits must be verified with regard to the potential donor who does not turn out to be the actual donor);
- hospital room, board and general nursing in semi-private rooms;
- special care units, such as coronary and intensive care;
- hospital ancillary services;
- physicians' services for surgery, technical assistance, administration of anesthetics, and medical care;
- acquisition, preparation, transportation, and storage of organ / tissue / cells;
- diagnostic services;
- drugs which require a prescription by federal law;
- medical and surgical care of the donor (related to the procurement of the organ / tissue / cells) if coverage is not available to the donor from any other source. (Covered services provided to a donor will be applied against the recipient's transplant maximum benefit, if applicable)

If the donor and recipient are both listed on the same (family) policy, BCBSKC charges only one deductible and one coinsurance, if applicable.

In addition to the specific organ criteria, transplant candidates must also meet the following general criteria, including, but not limited to:
· Since compliance is a major factor in transplant graft survival, the patient (or legal guardian) must have the ability to accept and understand the transplant procedure and to maintain compliance with long-term medical management and immunosuppression.
· If applicable, patients with a history of malignancy must have passed the recommended length of time to be considered cured for that specific cancer. A complete metastatic evaluation must be performed before a patient will be considered an acceptable transplant candidate.
· Patients with a history of alcohol or substance abuse must have a six month history of abstinence as evidenced by negative urine or serum drug screens taken randomly.
· The patient must have adequate cardiopulmonary status.
· The patient must be free from active infection.

A covered person is eligible for retransplantation as deemed medically necessary and appropriate by BCBSKC. Review of a retransplantation request will include review of the covered person’s compliance with relevant transplant selection criteria including, but not limited to, adherence to medication regimens, follow-up examinations and abstinence from the use of alcohol and drugs.

The specific member contract should be reviewed for coverage related to donors and recipients, out of network treatment, drugs and other possible limitations or exclusions.

Coverage will not be provided for:
· transplant services when the cost is covered by government, foundation or charitable grants,
· the purchase price or organs which are sold rather than donated to the recipient,
· an artificial organ.

Clinical trials for conditions other than those allowed in this policy may be available in the research setting. However, these trials are considered investigational and/or experimental and therefore contract exclusions.

Note: There are some state mandates in place that require insurance carriers to cover certain clinical trials under very specific guidelines. Please contact your BCBSKC representative for more information.

Due to the special nature of this procedure, this transplant may require an out of network exception.

**Description of Procedure or Service**
The heart/lung transplantation involves a coordinated triple operative procedure consisting of procurement of a donor heart-lung block, excision of the heart and lungs of the recipient, and implantation of the heart and lungs into the recipient. A heart/lung transplantation refers to the transplantation of one or both lungs and heart from a single cadaver donor.

**Background**
Combined heart/lung transplantation is intended to prolong survival and improve function in patients with end-stage cardiac and pulmonary diseases. The majority of recipients have Eisenmenger syndrome (37%), followed by idiopathic pulmonary artery hypertension (28%) and cystic fibrosis (14%). Eisenmenger syndrome is a form of congenital heart disease in which systemic-to-pulmonary shunting leads to pulmonary vascular resistance. Eventually, pulmonary hypertension may lead to a reversal of the intracardiac shunting and inadequate peripheral oxygenation, or cyanosis. (1)

However, the total number of patients with Eisenmenger syndrome has been declining in recent years, as a result of corrective surgical techniques and improved medical management of pulmonary hypertension. Heart/lung transplants have not increased appreciably for other indications either, as it has become more common to transplant a single or double lung and maximize medical therapy for heart failure, rather than perform a combined transplant. In these, patient survival rates are similar to lung transplant rates. Bronchiolitis obliterans syndrome is a major complication; 1-, 5-, and 10-year patient survival rates are 68%, 50%, and 40%, respectively. (1)
In 2012, 29 individuals received heart/lung transplants in the United States. As of the end of September 2013, there were 48 patients on the waiting list for heart/lung transplants.(2)

**Rationale**

**Literature Review**

This policy was originally created in 1996 and updated regularly with searches of the MEDLINE database. The most recent literature search was performed for the period September 2012 through September 26, 2013. Due to the nature of the population, there were no randomized controlled trials (RCTs) comparing heart/lung transplant to alternatives. Systematic reviews are based on case series and registry data. The extant RCTs compare surgical technique, infection prophylaxis, and immunosuppressive therapy and are not germane to this policy. The following is a summary of evidence based on registry data, case series, and expert opinion.

**Patient Selection**

Patients who are eligible for heart/lung transplantation can be listed under both the heart and lung allocation systems in the United States. In 2005, United Network for Organ Sharing (UNOS) changed the method by which lungs were allocated, from one based on length of time on the waiting list, to a system that incorporates the severity of the patient’s underlying disease, as well as likelihood of survival. (3) However, it has been noted that the individual systems underestimate the severity of illness in patients with both end-stage heart and lung failure, and modification of the lung allocation score can be appealed for patients who meet the following criteria:

- Deterioration on optimal therapy
- Right arterial pressure greater than 15 mm Hg
- Cardiac index less than 1.8 L/min/m².

**Pediatric Considerations**

In 2012, the Registry of the International Society for Heart and Lung Transplantation (ISHLT) reported on pediatric heart/lung transplant data collected through June 2011.(4) In recent years, the number of heart/lung transplant procedures in children has decreased, and the number of lung transplants has increased. There have not been any heart/lung transplants in infants since 2007. Overall, survival rates after heart/lung transplants are comparable in children and adults (median half-life of 4.7 and 5.3 years, respectively). For pediatric heart/lung transplants that occurred between January 1990 and June 2010, the 5-year survival rate was 49%. The 2 leading causes of death in the first year after transplantation were noncytomegalovirus infection and graft failure. Beyond 3 years posttransplant, the major cause of death was bronchiolitis obliterans syndrome.

**Retransplantation**

Repeat heart-lung transplant procedures have been performed; only 1 published study was found that reported on outcomes after repeat heart-lung transplants. The study, published by Shuhaiber and colleagues in 2008, involved a review of data from the UNOS registry.(5) The authors identified 799 primary heart-lung and 19 repeat heart-lung transplants. According to Kaplan-Meier survival analysis, the observed median survival times were 2.08 years after primary transplant and 0.34 years after repeat transplants. In addition, the authors analyzed survival data in matched pairs of primary and repeat transplant patients, who were matched on a number of potentially confounding demographic and clinical characteristics. Matches were not available for 4 repeat transplant patients. For the 15 repeat transplant patients with primary transplant matches, survival time did not differ significantly in the 2 groups. Being on a ventilator was statistically significantly associated with decreased survival time. The main limitation of this analysis is the small number of repeat transplant procedures performed.

**Potential Contraindications**

Individual transplant centers may differ in their guidelines, and individual patient characteristics may vary within a specific condition. In general, heart transplantation is contraindicated in patients who are not expected to survive the procedure, or in whom patient-oriented outcomes, such as morbidity or mortality, are not expected to change due to comorbid conditions unaffected by transplantation, eg, imminently terminal cancer or other disease. Further, consideration is given to conditions in which the
necessary immunosuppression would lead to hastened demise, such as active untreated infection. However, stable chronic infections have not always been shown to reduce life expectancy in heart transplant patients.

**Malignancy**
Concerns regarding a potential recipient's history of cancer were based on the observation of significantly increased incidence of cancer in kidney transplant patients. In fact, carcinogenesis is 2 to 4 times more common, primarily skin cancers, in both heart transplant and lung transplant patients, likely due to the higher doses of immunosuppression necessary for the prevention of allograft rejection. The incidence of de novo cancer in heart transplant patients approaches 26% at 8 years posttransplant, the rate for lung transplant is 28% at 10 years. For renal transplant patients who had a malignancy treated prior to transplant, the incidence of recurrence ranged from zero to more than 25%, depending on the tumor type. However, it should be noted that the availability of alternate treatment strategies informs recommendations for a waiting period following high-risk malignancies: in renal transplant, a delay in transplantation is possible due to dialysis; end-stage cardiopulmonary failure patients may not have an option. A small study of survivors of lymphoproliferative cancers who subsequently received cardiac transplant had 1-, 5-, and 10-year survival rates of 77%, 64%, and 50%, respectively. By comparison, overall 1-, 5-, and 10-year survival rates are expected to be 88%, 74%, and 55%, respectively for the general transplant candidate. The evaluation of a candidate who has a history of cancer must consider the prognosis and risk of recurrence from available information including tumor type and stage, response to therapy, and time since therapy was completed. Although evidence is limited, patients in whom cancer is thought to be cured should not be excluded from consideration for transplant. United Network for Organ Sharing (UNOS) has not addressed malignancy in current policies.

**HIV**
Solid organ transplant for patients who are HIV-positive (HIV+) has been controversial, due to the long-term prognosis for human immunodeficiency virus (HIV) positivity and the impact of immunosuppression on HIV disease. Although HIV+ transplant recipients may be a research interest of some transplant centers, the minimal data regarding long-term outcome in these patients consist primarily of case reports and abstract presentations of liver and kidney recipients. Nevertheless, some transplant surgeons would argue that HIV positivity is no longer an absolute contraindication to transplant due to the advent of highly active antiretroviral therapy (HAART), which has markedly changed the natural history of the disease.

As of February 2013, the United Network for Organ Sharing (UNOS) policy on HIV-positive transplant candidates states: “A potential candidate for organ transplantation whose test for HIV is positive should not be excluded from candidacy for organ transplantation unless there is a documented contraindication to transplantation based on local policy.”

In 2006, the British HIV Association and the British Transplantation Society Standards Committee published guidelines for kidney transplantation in patients with HIV disease. These criteria may be extrapolated to other organs:
- CD4 count greater than 200 cells/ml for at least 6 months
- Undetectable HIV viremia (less than 50 HIV-1 RNA copies/ml) for at least 6 months
- Demonstrable adherence and a stable HAART regimen for at least 6 months
- Absence of AIDS-defining [acquired immunodeficiency syndrome] illness following successful immune reconstitution after HAART.

However, concerns have been raised about the extrapolation of these criteria to lung transplants.

**Other**
Note: Considerations for heart transplantation and lung transplantation alone may also pertain to combined heart-lung transplantation. For example, cystic fibrosis accounts for the majority of pediatric...
candidates for heart-lung transplantation, and infection with *Burkholderia* species is associated with higher mortality in these patients. And, experience with kidney transplantation in patients infected with HIV in the era of HAART has opened discussion of transplantation of other solid organs in these patients. These topics are addressed more fully in the separate policies on heart transplantation and lung transplantation.

**Summary**

The available literature, consisting of case series and registry data, describes outcomes after heart-lung transplantation. Given the exceedingly poor expected survival without transplantation, this evidence is sufficient to demonstrate that heart/lung transplantation provides a survival benefit in appropriately selected patients. It may be the only option for some patients with end-stage cardiopulmonary disease. Heart/lung transplant is contraindicated in patients in whom the procedure is expected to be futile due to comorbid disease or in whom posttransplantation care is expected to significantly worsen comorbid conditions. Based on this evidence and established guidelines, heart–lung transplant may be considered medically necessary for those who meet clinical criteria and do not have contraindications to the procedure. A very limited amount of data suggest that, after controlling for confounding variables, survival rates after primary and repeat heart/lung transplants is similar. Findings are not conclusive due to the small number of cases of repeat heart-lung transplants reported in the published literature. Repeat heart-lung transplantation may be considered medically necessary in patients with a failed prior transplant who meet the clinical criteria for heart-lung transplantation.

**Practice Guidelines and Position Statements**

A key publication is the 2006 guidelines from the Pulmonary Scientific Council of the International Society for Heart and Lung Transplantation.(13) The consensus-based guidelines state that,

Lung transplantation is now a generally accepted therapy for the management of a wide range of severe lung disorders. However, the number of donor organs available remains far fewer than the number of patients with end-stage lung disease who might potentially benefit from the procedure. It is of primary importance, therefore, to optimize the use of this resource, such that the selection of patients who receive a transplant represents those with realistic prospects of favorable long-term outcomes. There is a clear ethical responsibility to respect these altruistic gifts from all donor families and to balance the medical resource requirement of one potential recipient against those of others in their society. These concepts apply equally to listing a candidate with the intention to transplant and potentially de-listing (perhaps only temporarily) a candidate whose health condition changes such that a successful outcome is no longer predicted.

Thus, for all patients, including those with end-stage cardiopulmonary disease and HIV infection, evaluation of a candidate for transplant needs to consider the probability of a successful transplant and the limited supply of organs available.

**Medicare National Coverage**

Heart/lung transplantation is covered under Medicare when performed in a facility that is approved by Medicare as meeting institutional coverage criteria.(14) The Centers for Medicare and Medicaid Services (CMS) has stated that under certain limited cases, exceptions to the criteria may be warranted if there is justification and if the facility ensures safety and efficacy objectives.

**References**


Billing Coding/Physician Documentation Information

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Additional Policy Key Words

N/A

Policy Implementation/Update Information

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11/1/10 No policy statement changes.
11/1/11 Policy statement revised to include the absolute contraindications as not medically necessary.
11/1/12 Severe heart failure added to second bullet point of medically necessary statement. Contraindications moved to Considerations. Absolute and relative contraindications combined and wording changed to be consistent with other solid organ transplant policies. Policy statement regarding transplantation not meeting criteria is clarified.
2/1/14 Two policy statements added; 1 on retransplantation and 1 stated that all other indications are considered investigational.

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