Clinical Policy Title: Lung transplant

Clinical Policy Number: 07.02.07

Effective Date: January 1, 2016
Initial Review Date: October 21, 2015
Most Recent Review Date: October 19, 2016
Next Review Date: October 2017

Policy contains:
- Lung transplants.

Related policies:

CP# 10.03.04 Corneal transplants (keratoplasty)
CP# 04.02.06 Heart valve transplants
CP# 14.02.06 Bone marrow transplants
CP# 13.02.01 Kidney transplants
CP# 04.02.05 Heart transplants
CP# 08.02.06 Pancreas transplants

ABOUT THIS POLICY: Keystone First has developed clinical policies to assist with making coverage determinations. Keystone First's clinical policies are based on guidelines from established industry sources, such as the Centers for Medicare & Medicaid Services (CMS), state regulatory agencies, the American Medical Association (AMA), medical specialty professional societies, and peer-reviewed professional literature. These clinical policies along with other sources, such as plan benefits and state and federal laws and regulatory requirements, including any state- or plan-specific definition of "medically necessary," and the specific facts of the particular situation are considered by Keystone First when making coverage determinations. In the event of conflict between this clinical policy and plan benefits and/or state or federal laws and/or regulatory requirements, the plan benefits and/or state and federal laws and/or regulatory requirements shall control. Keystone First's clinical policies are for informational purposes only and not intended as medical advice or to direct treatment. Physicians and other health care providers are solely responsible for the treatment decisions for their patients. Keystone First's clinical policies are reflective of evidence-based medicine at the time of review. As medical science evolves, Keystone First will update its clinical policies as necessary. Keystone First's clinical policies are not guarantees of payment.

Coverage policy

Keystone First considers lung transplants to be clinically proven and, therefore, medically necessary when any of the following criteria are met:

- Obstructive lung disease (e.g., emphysema, including alpha 1-antitrypsin deficiency; chronic obstructive pulmonary disease [COPD]; bronchiolitis obliterans; and bronchiectasis) in patients who meet any of the following:
  - Disease is progressive, despite maximal treatment, including medication, pulmonary rehabilitation, and oxygen therapy.
  - Individual is not a candidate for endoscopic or lung volume reduction surgery (LVRS). Simultaneous referral of individual with COPD for both lung transplant and LVRS evaluation is appropriate.
- **BODE index of 5 – 6:**

  - The BODE index is a composite score of body mass index (BMI), airway obstruction (percent predicted FEV1) (O), dyspnea (D), and exercise capacity (E). The BODE index calculator is available at [http://www.qxmd.com/calculate-online/respirology/bode-index](http://www.qxmd.com/calculate-online/respirology/bode-index). (Accessed September 30, 2016.)

- PaCO2 > 50 mmHg or 6.6 kilopascals (kPa) and/or PaO2 < 60 mmHg or 8 kPa.
- FEV1 < 25 percent predicted.
- Bronchopulmonary dysplasia.

- Congenital heart disease (Eisenmenger’s defect or complex) when any of the following is met:
  - Signs of right ventricular failure (i.e., progressive hepatomegaly, ascites, marked deterioration in functional capacity (New York Heart Association [NYHA] Class III).
  - Pulmonary hypertension with mean pulmonary artery pressure greater than 20 mm Hg by right heart catheterization.

- Cystic fibrosis in patients who display any of the following:
  - FEV1 that has fallen to 30 percent or individual with advanced disease with rapidly falling FEV1 despite optimal therapy (particularly in a female patient), infected with non-tuberculous mycobacterial (NTM) disease or BMI cepacia complex and/or with diabetes.
  - A six-minute walk distance < 400 m.
  - Development of pulmonary hypertension in the absence of a hypoxic exacerbation (as defined by a systolic pulmonary arterial pressure [PAP] > 35 mm Hg on echocardiography or mean PAP > 25 mm Hg measured by right heart catheterization).

- Clinical decline characterized by increasing frequency of exacerbations associated with any of the following:
  - An episode of acute respiratory failure requiring noninvasive ventilation.
  - Increasing antibiotic resistance and poor clinical outcomes.
  - Recovery from exacerbations.
  - Worsening nutritional status despite supplementation.
  - Pneumothorax.
  - Life-threatening hemoptysis despite bronchial embolization.

- Primary pulmonary hypertension in patients who meet any of the following:
  - NYHA Functional Class III or IV symptoms during escalating therapy.
  - Rapidly progressive disease (assuming weight and rehabilitation concerns are not present).
  - Use of parenteral targeted pulmonary arterial hypertension (PAH) therapy regardless of symptoms or NYHA Functional Class.
- Known or suspected pulmonary veno-occlusive disease (PVOD) or pulmonary capillary hemangiomatosis.

- Restrictive lung disease (e.g., idiopathic pulmonary fibrosis, desquamative interstitial fibrosis, post-chemotherapy allergic alveolitis, systemic sclerosis [scleroderma], collagen vascular disease, asbestosis, or eosinophilic granuloma).

- Pulmonary fibrosis in patients who meet any of the following:
  - Presence of cor pulmonale (indicative of severe pulmonary fibrosis) or pulmonary hypertension.
  - Diffusing capacity for carbon monoxide (DLCO) is less than 60 percent expected.
  - Total lung capacity (TLC) less than 70 percent expected.

- Sarcoidosis in patients who meet any of the following:
  - Presence of cor pulmonale (indicative of severe pulmonary fibrosis) or pulmonary hypertension.
  - Total lung capacity less than 70 percent predicted.
  - DLCO less than 60 percent expected.
  - Lymphangioleiomyomatosis (LAM) with end-stage pulmonary disease.
  - Graft versus host disease or failed primary lung graft.

- Including the pre-transplant and post-discharge services, and the treatment of complications as approved by the plan.

Limitations:

Keystone First considers lung transplantation for all other conditions not listed in the coverage policy above to be not medically necessary.

Relative contraindications for adults and children include, but may not be limited to:

- Age appropriateness:
  - 65 years old for single lung
  - 65 years old for double lung transplant.
  - 55 years old for heart and lung transplant.

- Active smoker (less than six months since quitting).

- Active substance use disorder.

- Chronic mechanical ventilation (unless tolerating three hours of physical therapy/day and free of bacterial colonization).

- Previous lung transplant (rare exceptions for John Hopkins Hospital primary transplant patients).

- Severe diffuse coronary artery disease (especially with poor ejection fraction (EF)).

- End-stage renal disease (creatinine clearance < 40 mg/min).
• End-stage liver disease.
• Bone marrow dysfunction.
• Human immunodeficiency virus (HIV). (HIV inclusion criteria: CD4 count greater than or equal to 200 cells/ml for at least six months, undetectable HIV viremia for six months, adherence to HAART regimen for greater than six months)
• Severe local or systemic infection.
• Severe neurologic deficits.
• Untreatable psychiatric conditions.
• A recent history of malignancy.
• Morbid obesity (BMI > 35).
• Severe malnutrition/cachexia.
• Chronic prednisone use > 20 mg/day.
• Symptomatic osteoporosis.
• Psychiatric or social problems (including noncompliance).
• Financial problems (no prescription coverage).
• Previous thoracic surgery.
• Lack of family or social support.
• Cancer in the last five years, except localized skin (never melanoma).
• Colonization with resistant organisms.

Alternative covered services:

• Maximum medical management of COPD.
• Maximum medical management of pulmonary arterial hypertension.

Background

Lung transplantation or pulmonary transplantation is a surgical procedure in which a patient's diseased lungs are partially or totally replaced by lungs that come from a donor. Donor lungs can be retrieved from a living donor or a deceased donor. A living donor can only donate one lung lobe. With some lung diseases, a recipient may only need to receive a single lung. With other lung diseases, such as cystic fibrosis, it is imperative that a recipient receive two lungs. While lung transplants carry certain associated risks, they can also extend life expectancy and enhance the quality of life for end-stage pulmonary patients.

More than 6,400 lung transplants have been performed since the first successful operations in the early 1980s. In 2010, 1,770 lung transplant procedures were performed in the U.S., yet 2,469 new candidates were added to the waiting list the same year. Lung transplant programs now exist in many countries. Internationally, the number of donor organs available is far fewer than the number of patients with end-stage lung disease. Because of this, many candidates die on the waiting list, and the average wait to receive a donor organ may approach two years.
Overall survival rates are between 60 percent and 65 percent at two years and approximately 40 percent at five years. Considering the resource limitations and the importance of assuring optimum outcomes, it is believed that international guidelines for selection of appropriate candidates for lung transplant will ensure a fair distribution of donor organs. Transplant physicians and surgeons representing the International Society of Heart and Lung Transplantation, the American Society of Transplant Physicians, the American Thoracic Society, the European Respiratory Society, and the Thoracic Society of Australia and New Zealand have agreed on the information in the following document as acceptable guidelines for candidates for lung transplantation.

Lung transplantation should be considered for patients with advanced lung disease whose clinical status has progressively declined despite maximal medical or surgical therapy. Candidates are usually symptomatic during activities of daily living and have a limited expected survival over the next two years. In addition, the ideal candidate should be free of significant other organ dysfunction and extrapulmonary manifestations of a systemic disease. Guidelines for recipient selection have been developed by the American Thoracic Society and the International Society of Heart and Lung Transplantation.

Emphysema is a form of COPD defined by abnormal and permanent enlargement of the air spaces distal to the terminal bronchioles. It is associated with the destruction of the alveolar walls. Emphysema causes dyspnea through airflow limitation, hyperinflation and loss of gas exchanging surfaces in the lungs (also known as increased physiologic dead space).

LVRS (also called reduction pneumoplasty or bilateral pneumectomy) is a surgical technique that may be beneficial for some patients with advanced emphysema who have poor control of their disease despite maximal medical therapy. LVRS entails reducing the lung volume by wedge excision of emphysematous tissue.

The mechanisms by which LVRS might provide benefit are not known with certainty. It has been suggested that LVRS reduces the size mismatching between the hyperinflated lungs and the chest cavity, thereby restoring the outward circumferential pull on the bronchioles (i.e., increasing elastic recoil) and improving expiratory airflow. As an example, in a study of 20 patients undergoing volume reduction surgery, 16 experienced an increase in elastic recoil. The patients with improved elastic recoil had a significantly greater increase in exercise capacity than the four without increased elastic recoil.

**Lung transplant types and the condition:**
- Bilateral lung transplant (BLT):
  - Cystic fibrosis.
  - Bronchiectasis.
  - Pulmonary hypertension.
  - Emphysema.
  - Pulmonary fibrosis (idiopathic or secondary to scleroderma or other disease states).
- Single lung transplant (SLT):
  - Emphysema.
  - Pulmonary fibrosis (idiopathic or secondary to scleroderma or other disease states).
- Heart and lung transplant:
  - Same as SLT and BLT with:
    - Poor left ventricular function or irreversible right ventricular function.
    - Surgically irreparable congenital heart defects.

Deceased donor lung transplantation:
- A deceased donor, also known as cadaveric donor, is the most common donor source used for lung transplantation. In 1995, the United Network for Organ Sharing (UNOS) changed the method for allocating donated cadaver lungs for individuals over age 12 by assigning each candidate a lung allocation score based on survival benefit and urgency rather than waiting time (Mulligan, 2008). In contrast, allocation to children under age 12 continues to be based on waiting time. Preferential transplantation of sicker patients has not resulted in an increase in early mortality following transplantation (Kotloff, 2010).
- According to the Organ Procurement and Transplantation Network (OPTN, 2014) national data for deceased donor primary lung transplantation performed between 1997 and 2004, graft survival rates were 83.1 percent, 62.1 percent, and 46.2 percent, respectively, at one, three, and five years (based on OPTN data as of July 4, 2014).

Living donor lung transplantation (LDLT):
- Use of a live donor as a source for lung transplantation was initiated in 1993 due to the higher demand than supply for patients waiting for lung transplantation. Although LDLT may be appropriate for a highly selected individual who likely would not survive waiting times for a deceased donor, it is now rarely performed. According to the OPTN annual report (2012), only one LDLT was performed in 2012, with four LDLTs performed between 2007 and 2012. Survival data for LDLT performed in 2012 were not published in the annual report.
- This procedure requires the donation of one lung lobe from each of two living donors. Major complications have included pleural effusion, bronchial stump fistula, bilobectomy, hemorrhage, phrenic nerve injury, pulmonary artery thrombosis, and bronchial stricture. Minor complications include persistent air leak, arrhythmia, and pneumonia (Solomon, 2010). Deceased donor transplantation is preferred to avoid the risk to two healthy donors (Solomon, 2010).

Lung transplantation should not be offered to adults with a recent history of malignancy:
- A two-year disease-free interval combined with a low predicted risk of recurrence after lung transplantation may be reasonable, for instance, in non-melanoma localized skin cancer that has been treated appropriately. However, a five-year disease-free interval is prudent in
most cases, particularly for patients with a history of hematologic malignancy; sarcoma; melanoma; or cancers of the breast, bladder, or kidney. Unfortunately, for a portion of patients with a history of cancer, the risk of recurrence may remain too high to proceed with lung transplantation even after a five-year disease-free interval.

Lung transplantation in children

Lung transplantation in children is evolving. Diseases that are potentially amenable to lung transplantation include primary pulmonary hypertension, pulmonary hypertension associated with structural heart disease, pulmonary vein stenosis, pulmonary hypertension associated with parenchymal lung disease, and congenital abnormalities of lung development or of lung adaptation to extrauterine life. As in adults, maximal medical therapy, including vasodilators and supplemental oxygen, should be instituted before children are considered for transplantation. Since the diagnoses are varied and the disease spectra diverse, prognostic indicators have been difficult to develop; thus empirical criteria are the primary means of selecting candidates.

Searches

Keystone First searched PubMed and the databases of:

- UK National Health Services Centre for Reviews and Dissemination.
- Agency for Healthcare Research and Quality’s National Guideline Clearinghouse and other evidence-based practice centers.
- The Centers for Medicare & Medicaid Services (CMS).

We conducted searches on September 14, 2015. Search terms were: “MeSH chronic obstructive pulmonary disease,” frailty,” “interstitial lung disease,” “lung transplantation,” “obesity,” “pulmonary arterial hypertension,” “sarcopenia,” and “cystic fibrosis.”

We included:

- **Systematic reviews**, which pool results from multiple studies to achieve larger sample sizes and greater precision of effect estimation than in smaller primary studies. Systematic reviews use predetermined transparent methods to minimize bias, effectively treating the review as a scientific endeavor, and are thus rated highest in evidence-grading hierarchies.
- **Guidelines based on systematic reviews**.
- **Economic analyses**, such as cost-effectiveness, and benefit or utility studies (but not simple cost studies), reporting both costs and outcomes — sometimes referred to as efficiency studies — which also rank near the top of evidence hierarchies.

**Findings**
According to the 2014 Registry report, the median survival for all adult recipients is 5.7 years, but bilateral lung recipients appear to have a better median survival than single lung recipients (7 versus 4.5 years, respectively.) (For a figure showing updated information, please see the International Society for Heart and Lung Transplantation slide set "Overall Lung and Adult Lung Transplantation Statistics," slide titled Number of Lung Transplants Performed by Year and Procedure Type at ISHLT Registry. However, it is unclear if this survival advantage is directly related to the type of operation or to the underlying recipient characteristics.

The biological needs and circumstances of candidates younger than age 12 are different from either adolescent or adult candidates. One key difference is the size and lung capacity of donors and patients among these age ranges. For this reason, lung allocation policy differs for these groups of candidates and is designed to suit their unique needs. Children younger than age 12 have priority for all donors of similar age and size within a 1,000-mile radius before any older candidates would be considered. In some circumstances, a transplant center may determine that a child's condition warrants a reduced size transplant from an adult donor. If the center wishes to consider this additional treatment option, these children will have access to adult organs once they are offered to adolescents and adults in the same allocation zone.

In 2012, there were 460 pediatric organ donors in the United States, including 114 between the ages of 6 and 10 years. Although there were only 11 lung donors in that age group, that number likely reflects low demand (two lung transplants in recipients aged 6 – 10) as much as organ availability.

Although definitive patient selection criteria for lung transplantation have not been established, there is evidence to indicate that potential recipients with chronic lung disease could be considered if they are not eligible for further medical or surgical therapy and if they have a < 50 percent chance of surviving for 24 to 36 months, when transplantation is expected to confer a survival advantage, and when there are no contraindications. International guidelines for selection of lung transplantation candidates, as set out by the ISHLT, contain general and disease-specific selection criteria, and absolute and relative contraindications. These guidelines aim to facilitate the selection process and to promote a fair distribution of donor organs. However, the final decision to place a candidate on the waiting list resides with the expertise and practice of individual transplant centers and will vary from country to country.

Evidence from registry and cohort studies on deceased donor lung transplantation indicated that a substantial number of the recipients will derive a survival advantage from the procedure, together with improvement in quality of life (QOL). Evidence was controversial as to which diagnostic group of patients benefited the most in survival or health-related quality of life (HRQOL) outcomes, how long after transplantation the survival benefit was reached, and which type of transplantation (single or bilateral) was associated with longer survival. Evidence indicated that lung transplantation was associated with life-threatening complications, such as rejection to the allograft and infection. In addition, lifelong commitment to immunosuppressive medications was associated with inherent adverse effects. Thus, transplantation will improve survival and QOL, but at the same time, it will introduce new restrictions and complications.
Policy updates:

2016 — Added citation updated links and references.

Summary of clinical evidence:

<table>
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<tr>
<th>Citation</th>
<th>Content, Methods, Recommendations</th>
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| Stephenson AL, et al. (May 2015). Clinical and demographic factors associated with post-lung transplantation survival in individuals with cystic fibrosis | Key points:  
- Contemporary studies evaluating post-transplant survival are limited and often include data from single centers or selected sub-groups. The purpose of this study was to evaluate overall transplant survival and to identify risk factors associated with death after transplant.  
- After lung transplantation, five-year survival in Canadians with cystic fibrosis is 67%, and 50% of patients live > 10 years.  
- Despite these impressive probabilities, age at transplant, pancreatic sufficiency, and B cepacia infection remain important determinants of survival after lung transplantation. |
| Kaltman JK (2007) Pediatric Cardiomyopathy Registry (PCMR) from 1990 to 2007 | Key points:  
- This population-based study used data from the Pediatric Cardiomyopathy Registry (PCMR). From 1990 to 2007, the PCMR, led by the University of Miami Leonard M. Miller School of Medicine, enrolled 1,731 children (18 years of age or younger) diagnosed with pediatric dilated cardiomyopathy, the most common heart muscle disease.  
- Dilated cardiomyopathy can lead to heart valve problems, arrhythmias (irregular heartbeats), blood clots in the heart and even heart failure. |
| Robertson et al. (2012) Evaluating the safety of fundoplication in LTX recipients and its effects on quality of life. | Key points:  
- Between June 1, 2008, and December 31, 2010, a prospective study of LTX recipients undergoing fundoplication was undertaken.  
- Quality of life was assessed before and after surgery. Body mass index (BMI) and pulmonary function were followed-up.  
- A total of 16 patients, mean +/- SD age of 38 +/-11.9 yrs, underwent laparoscopic Nissen fundoplication.  
- There was no peri-operative mortality or major complications.  
- Mean +/- SD hospital stay was 2.6 +/- 0.9 days; 15 out of 16 patients were satisfied with the results of surgery post-fundoplication.  
- There was a significant improvement in reflux symptom index and DeMeester questionnaires and gastro-intestinal quality of life index scores at six months. |
| Snell GI, et al. (2000) Outcomes from paired single-lung transplants from the same donor | Key points:  
- Simultaneous, paired SLTs from a single organ donor are one way to maximize lung transplant opportunities.  
- Paired transplants allow comparison between left and right SLTs and also provide insight into the relevance of donor vs. recipient factors in rejection outcomes.  
- The general outcomes of right and left transplants are similar, although we observed increased six-month to two-year mortality associated with left lung transplantation.  
- The lack of correlation between the incidence of acute rejection episodes or the severity of BOS in paired allograft recipients suggests that "donor factors" are not the dominant cause. |
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<th>Citation</th>
<th>Content, Methods, Recommendations</th>
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</thead>
</table>
| Black MC, et al. (2014)  
Double lung transplants have significantly improved survival compared with single lung transplants in high lung allocation score patients | **Key points:**  
- The UNOS Thoracic Transplant Database for lung transplants from January 2005 to June 2012 was used for analysis.  
- Propensity matching was used to minimize differences between the high and low LAS groups and between SLTs and BLTs in the high LAS group.  
- Despite a higher operative morbidity, patients who had a high LAS did substantially better in survival if two lungs were transplanted rather than only one, with a larger difference in survival than for patients with a lower LAS. |
| Kirshbom PM, et al. (2002)  
Use of extracorporeal membrane oxygenation in pediatric thoracic organ transplantation | **Key points:**  
- Mechanical cardiorespiratory support is occasionally required before or after pediatric thoracic organ transplantation.  
- Extracorporeal membrane oxygenation is the most commonly used mechanical support technique in children.  
- The goal of this study was to examine the indications for initiation and outcomes after peri-transplant use of extracorporeal membrane oxygenation. |
| Berry G J (2016). Lung Transplantation | **Key points:**  
- The therapeutic options for patients with advanced pulmonary parenchymal or vascular disorders are currently limited.  
- Lung transplantation remains one of the few viable interventions, but on account of the insufficient donor pool only a minority of these patients actually undergo the procedure each year.  
- Following transplantation, there are a number of early and late allograft complications, such as primary graft dysfunction, allograft rejection, infection, post-transplant lymphoproliferative disorder, and late injury that is now classified as chronic lung allograft dysfunction.  
- The pathologist plays an essential role in the diagnosis and classification of these myriad complications. |

**Glossary**

**Burkholderia cepacia complex (B. cepacia)** — Consists of different species of bacteria that are found in the natural environment. Some of these species pose serious risks to the health of a person with cystic fibrosis.

**Chronic obstructive pulmonary disease (COPD)** — A progressive disease that makes it hard to breathe. There are two main forms of COPD: chronic bronchitis, which involves a long-term cough with mucus, and emphysema, which involves damage to the lungs over time. Most people with COPD have a combination of both conditions.

**Extracorporeal life support (ECLS)** — ECLS systems are mechanical devices to temporarily support the failing heart and lung. It is a further development of a conventional heart-lung machine (HLM). Compared to the HLM, it is smaller and has been reduced to only main components, such as the centrifugal pump and a membrane oxygenator. It is highly mobile and can be used both in and outside of the hospital.

**Extracorporeal membrane oxygenation (ECMO)** — A treatment that uses a pump to circulate blood through an artificial lung back into the bloodstream of a very ill baby. This system provides heart-lung
bypass support outside of the baby's body. It may help support a child who is awaiting a heart or lung transplant.

**Eisenmenger's defect or complex** — A condition that affects blood flow from the heart to the lungs in some people who were born with structural problems of the heart.

**References**

**Professional society guidelines/other:**


**Peer-reviewed references:**


Clinical trials:

Searched clinicaltrials.gov on September 30, 2016 using terms lung transplants | Open Studies. 228 studies found, relevant two cited below:


CMS National Coverage Determinations (NCDs):

No NCDs identified as of the writing of this policy.

On March 23, 2007, Medicare issued a final rule setting forth requirements that transplant centers must meet to participate in the Medicare program that moves Medicare-covered transplant programs toward an outcome-focused system (CMS, 2007). The rule became effective on June 28, 2007. Transplant organ programs were defined as a component within a transplant hospital that provides transplantation of a particular type of organ. All organ transplant programs must be located in a hospital that has a Medicare provider agreement. In addition to meeting the transplant Conditions of Participation, the transplant program must also comply with the hospital Conditions of Participation (CMS, 2009).

Local Coverage Determinations (LCDs):

No LCDs identified as of the writing of this policy.
**Commonly submitted codes**

Below are the most commonly submitted codes for the service(s)/item(s) subject to this policy. This is not an exhaustive list of codes. Providers are expected to consult the appropriate coding manuals and bill accordingly.

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