# Molina Clinical Policy
# Lung Transplantation: Policy No. 115
**Last Approval: 10/09/2024**
**Next Review Due By: October 2025**

## DISCLAIMER

This Molina Clinical Policy (MCP) is intended to facilitate the Utilization Management process. Policies are not a supplementation or recommendation for treatment; Providers are solely responsible for the diagnosis, treatment, and clinical recommendations for the Member. It expresses Molina's determination as to whether certain services or supplies are medically necessary, experimental, investigational, or cosmetic for purposes of determining appropriateness of payment. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered (e.g., will be paid for by Molina) for a particular Member. The Member's benefit plan determines coverage – each benefit plan defines which services are covered, which are excluded, and which are subject to dollar caps or other limits. Members and their Providers will need to consult the Member's benefit plan to determine if there are any exclusion(s) or other benefit limitations applicable to this service or supply. If there is a discrepancy between this policy and a Member's plan of benefits, the benefits plan will govern. In addition, coverage may be mandated by applicable legal requirements of a State, the Federal government or CMS for Medicare and Medicaid Members. CMS's Coverage Database can be found on the CMS website. The coverage directive(s) and criteria from an existing National Coverage Determination (NCD) or Local Coverage Determination (LCD) will supersede the contents of this MCP and provide the directive for all Medicare members. References included were accurate at the time of policy approval and publication.

## OVERVIEW

**Lung transplantation** is a surgical procedure performed to replace a diseased lung with a healthy donor lung. Transplant may be required for those with lung issues that do not respond to other treatments or those with a life expectancy of 12 to 24 months if a transplant is not performed ([1]Hachem 2024; [1]OPTN 2023). Previously, the number of completed lung transplants were limited due to high mortality rates from intra- and post-surgical complications; however, advances in surgical technique, post-operative management, recipient selection, and the availability of cyclosporine have improved lung transplant outcomes and made lung transplant an option for patients of all ages ([1]Hachem 2024). The primary limiting factors for the number of lung transplants performed is a lack of suitable healthy lung donors and potential complications before and after donor brain death that affect the viability of potential donor lungs ([1]Hachem 2024). The number of lung transplants in the United States peaked in 2019 at 2,759 and subsequently declined due to the COVID-19 pandemic; however, the number of lung transplants is steadily increasing and approximately 2,743 lung transplants were performed in 2022 (SRTR 2024). The number of individuals added to the transplant waiting list continues to increase which has led to the development of the "Continuous Distribution Framework" to improve lung allocation ([1-2]OPTN 2023; SRTR 2024). There is also a focus on the management of patients with end-stage lung disease who are waiting for a suitable donor. Management depends on the cause of lung disease and includes, but is not limited to ([2]Hachem 2024):

- Lung volume reduction surgery
- Oxygen therapy
- Pulmonary rehabilitation
- Treatment of reversible airway disease
- Vasodilators
- Pulmonary thromboendarterectomy in patients with chronic pulmonary thromboembolic disease

### Continuous Distribution Framework and Lung Composite Allocation Score

The Organ Procurement and Transplantation Network (OPTN) developed the "continuous distribution" framework in December 2018 to improve the process of matching organ donors with recipients and make the donation process more equitable and improve waitlist mortality. Each organ will have a "composite allocation score (CAS)" that assigns a score based on attributes such as medical urgency, pediatric age group, post-transplant survival, blood type, sensitization, candidate size, prior living donor status, and placement efficiency. Each attribute has a specific weight that indicates the effect that attribute will have on the total score ([1]OPTN 2023).

The lung CAS for lung transplantation was developed and implemented in March 2023 by the OPTN to replace the previously utilized "lung allocation score." The lung CAS has a maximum possible score of 100 points based on waiting list urgency (1-year survival without a transplant), expected 5-year post-transplant survival, biologic compatibility (blood type, calculated panel reactive antigen, and height), patient access, and placement efficiency (travel and proximity to transplant center). The lung CAS "will extend to decimal point values, meaning there should not be many instances where two or more candidates will have the exact same score ([3]OPTN 2023)." If there are two candidates with the same score, the candidate that was registered first will be prioritized to receive the donor lung ([2]OPTN 2023). Regarding candidates awaiting multi-organ transplants, "if a heart-lung, lung-liver, or lung-kidney candidate has a lung

CAS that is equal to or greater than the threshold for multi-organ transplantation including the lung, the [organ procurement organization] must offer both organs to that candidate ([2]OPTN 2023)."

Pediatric patients awaiting lung transplant are given preset scores for post-transplant outcomes and patient access goals. Pediatric patients <12 years of age awaiting a lung transplant are assigned either "priority 1" or "priority 2" to indicate medical urgency based on clinical criteria. Additionally, all pediatric patients <12 years of age "are assigned 18.6336 post-transplant outcome points based on their post-transplant survival probability…and all candidates registered before the age of 18, including those younger than age 12, receive an additional 20 points under the patient access goal for pediatric status ([2]OPTN 2023)." If a pediatric patient is placed on the waiting list prior to age 12 and remains on the waiting list on their 12th birthday, that candidate will receive a calculated waiting list survival and post-transplant survival scores based on scored clinical values rather than the predetermined value based on priority. Those that are placed on the waiting list prior to age 18 and turn 18 years old while still on the waiting list "will continue to receive the 20 points for pediatric status after they turn 18 ([2]OPTN 2023)."

**Conditions Associated with Lung Transplantation**

**Interstitial Lung Disease (ILD)** is a term used to describe lung diseases that begin in the pulmonary interstitium. The diseases are classified together due to similar clinical, radiographic, physiologic, and/or pathologic manifestations ([1]King 2024). The lung diseases are further grouped according to whether the cause of the disease is known or unknown. Unknown causes of ILD are referred to as "idiopathic." Diseases classified as idiopathic ILD include cryptogenic organizing pneumonia, sarcoidosis, and idiopathic interstitial pneumonias (acute, subacute, smoking-related, and chronic fibrosing) ([1]King 2024). Causes of ILD may include infectious organisms (bacterial, fungal, or viral), exposure to occupational or environmental agents, drug-induced pulmonary toxicity, or radiation-induced lung injury ([1]King 2024). Most of the symptoms for each disease process are nonspecific, making it important to determine the 1) duration and severity of symptoms, 2) past medical, family, and social history, 3) results of radiographic imaging of the chest, and 4) pulmonary function test (PFT) results ([1]King 2024). PFT trending can help determine the severity of reduction and is also used as an indicator for lung transplant referral ([2]King 2024; Leard et al. 2021). ILD accounts for approximately 40% of all lung transplants and interstitial pulmonary fibrosis is the most common form of ILD that is referred for lung transplantation ([2]King 2024; Leard et al. 2021).

**Chronic Obstructive Pulmonary Disease (COPD)** is the world's third leading cause of death according to the World Health Organization (WHO 2020). In the United States, COPD was the fourth leading cause of death. Approximately 15.7 million Americans, or roughly 6.4% of the population, have been diagnosed with COPD. However, there may be a significantly higher number of cases yet to be diagnosed as COPD in the United States as more than 50% of adults with low pulmonary function were not aware that they had COPD when receiving an official diagnosis. Women are typically diagnosed with COPD during the advanced stages of the disease while men are typically diagnosed in the earlier stages of the disease, leading to a decline in age-adjusted death rates for men while the death rates for women have remained the same. Treatment for COPD in women tends to be less effective due to the advanced stages of disease on diagnosis. COPD is typically diagnosed using PFT trending, specifically spirometry. The BODE index is typically used in conjunction with PFTs to assess a patient for referral for transplant evaluation at a transplant center ([2]Hachem 2024). The BODE index provides a score from 0-10 based on **b**ody mass index, airflow **o**bstruction, **d**yspnea, and **e**xercise capacity ([2]Hachem 2024). A higher score indicates a lower 4-year survival rate ([2]Hachem 2024). Approximately 30.6% of all lung transplants occur in individuals with COPD ([1]CDC 2024; GOLD 2023).

**Cystic Fibrosis (CF)** is an inherited genetic disorder resulting from mutations in the cystic fibrosis transmembrane conductance regulator gene. According to Simon (2024), "CF is a multisystem disease" that causes dysfunction of multiple organs. The most common cause of mortality in patients with CF is pulmonary disease as CF causes thick, sticky mucus that blocks the airways and can lead to lung damage. In addition, CF can increase the risk of recurrent respiratory infections due to germs becoming trapped in the thick, sticky mucus. Those with CF have a decreased ability to absorb nutrients from food due to proteins needed for digestion not reaching the intestines. Various organs can be affected by CF which may lead to other health conditions such as diabetes, cirrhosis of the liver, arthritis, reflux, hypersplenism, and osteoporosis. Due to being inherited, genetic counseling and testing is recommended if there is a family history of CF. Screening for CF is part of newborn screening panels in the United States as early detection allows for early treatment. Regular treatments for CF consist of airway clearance therapies, bronchodilators, pancreatic enzyme replacement therapies, anti-inflammatory therapies, and infection prevention. Routine PFTs are also performed to assess and trend lung function. Lung transplantation may be indicated if the disease progresses and symptoms such as massive hemoptysis and recurrent pneumothorax develop. Other indications for referral for transplant evaluation include rapidly declining PFT results, more than 2 exacerbations per year requiring intravenous

antibiotics, a body mass index < 18 despite improving nutritional status, and hypoxemia or hypercarbia (Ramos et al. 2019). Referral for a transplant evaluation is recommended earlier in the disease process "to allow patients and their families to better prepare for transplantation medically and psychologically ([2]Hachem 2024)." Approximately 35,000 people in the United States have cystic fibrosis ([2]CDC 2024; [2]Hachem 2024; Simon 2024).

**Bronchopulmonary dysplasia** is a chronic lung disease that is most common in infants born prematurely due to underdeveloped and immature lungs, but it may also affect older infants that are born with conditions that affect normal lung development. Bronchopulmonary dysplasia develops after birth and affects normal breathing with some infants requiring supplemental oxygen therapy following discharge from the hospital. While many children recover, bronchopulmonary dysplasia may cause long-term breathing difficulties due to progressive pulmonary deterioration which could warrant a lung transplant. The disease affects 10,000-15,000 infants annually in the United States. The risk of developing bronchopulmonary dysplasia increases with a lower birth weight (< 2.2 pounds) and a lower gestational age at birth. The incidence of bronchopulmonary dysplasia has continued to increase as medical teams are able to keep infants with much lower gestational ages and birth weights alive compared to the past due to advances in medicine, such as surfactant and improved mechanical ventilation and non-invasive ventilation strategies (Dani et al. 2023; NORD 2023).

Other conditions that may warrant lung transplantation include pulmonary hypertension, and heart disease or heart defects. Hereditary conditions and other diseases causing severe lung damage such as sarcoidosis, histiocytosis, and lymphangioleiomyomatosis may also necessitate transplantation. *Lung cancer is not a condition for which transplant is generally recommended.*

## Transplant Risks

Risks of lung transplantation include bleeding, infection, blockage of the arteries and/or blood vessels to the new lung(s), blockage of the airways, severe pulmonary edema, and blood clots. A major risk of transplant is allograft rejection which is the body's normal reaction to a foreign object or tissue. Immunosuppressive agents are typically used to reduce the risk of allograft rejection; however, there are several side effects associated with immunosuppression that may complicate the recovery period. Each transplant program is responsible for assessing each recipient's individual risk factors to determine if there are contraindications to transplant (Ahya & Kawut 2024; Pilewski 2023; Leard et al. 2021).

## Types of Lung Transplantation

**Single Lung Transplant:** This type of transplant "accounts for approximately 15-20% of adult lung transplantations (Hartwig & Klapper 2023)." Single lung transplant is commonly performed in patients with idiopathic pulmonary fibrosis and may also be performed in patients with other disease processes, such as COPD and mild to moderate pulmonary hypertension, to extend the limited supply of donor lungs ([3]Hachem 2024; Hartwig & Klapper 2023). The lung with worse pulmonary function is chosen for replacement (Hartwig & Klapper 2023). If both lungs function equally, then the right lung is usually favored for removal, as this avoids having to maneuver around the heart which is required for excision of the left lung (Hartwig & Klapper 2023).

**Bilateral Lung Transplant:** A bilateral lung transplant can be performed sequentially, en bloc, or simultaneously. Removal of both lungs is mandatory in patients with end stage CF as single lung transplant "provides less lung function as a buffer for late complications." Bilateral lung transplant is also preferred for patients with severe pulmonary hypertension and bronchiectasis (Hartwig & Klapper 2023).

**Heart-Lung Transplant:** A heart-lung transplant refers to transplantation of the heart and one or both lungs from a single cadaver donor. A heart-lung transplant is intended to prolong survival and improve function in recipients with end stage cardiopulmonary disease. Indications for a heart-lung transplant include complex congenital heart disease with Eisenmenger syndrome, nonidiopathic pulmonary hypertension due to congenital heart disease, CF, cardiomyopathy, and idiopathic pulmonary hypertension. Advancements in the medical management of pulmonary hypertension and congenital heart disease (including Eisenmenger syndrome) have decreased the need for heart-lung transplant. In addition, heart-lung transplant is less preferred to isolated lung and heart transplantation as there are disadvantages to combined heart-lung transplant including "the need to procure a heart-lung block [leading] to increased waiting time and increased mortality among patients awaiting combined heart-lung transplant" and "exposure of the recipient to the risks of both graft coronary artery vasculopathy and chronic lung allograft dysfunction (Singer & Mooney 2024)."

**Living Donor Lobar Transplant:** Living donor lobar lung transplantation involves the transplantation of a lung lobe or lobes from one or two healthy donors to replace the diseased lung(s) of a recipient with end-stage lung disease. The indications for this type of transplant include improvement of functional status and quality of life as well as to prolong survival in a patient who requires lung transplantation but whose deteriorating condition will likely lead to death before a cadaveric organ becomes available. Each donor donates only one lung lobe. The decision concerning which lobe should be donated is based on an optimal size match between the potential donor and the recipient. This transplant option is "limited to children and adults with small stature." Most living donor lung recipients are patients with CF and most lung donors are first-degree relatives who are compatible in terms of size and ABO blood group. Living donation is an alternative to cadaveric organ donation particularly when cadaveric transplantation is unavailable, or in patients who are deteriorating clinically to the point of transplant ineligibility while waiting for a cadaveric donor. Living donation may also be an option for critically ill children, due to a shortage of suitable cadaveric donors for this patient population ([3]Hachem 2024).

**Split Lung Bilateral Lobar Transplant:** With this procedure, a single left lung from a donor who is approximately 15% taller than the recipient is divided such that the left upper and lower lobes are implanted into the recipient's right and left hemithorax, respectively. The principal advantage of split lung bilateral lobar transplantation is that it permits single lung transplantation from a donor with a large size discrepancy with the recipient, such as a small adult or child ([3]Hachem 2024).

**Retransplantation**: Retransplantation refers to replacing a previously transplanted lung or lungs (primary transplant) with a new donor lung or lungs following graft failure. Retransplantation is only considered in carefully selected patients due to the difficulties associated with retransplantation. The surgical procedure is much more complicated due to the thoracic adhesions present from the primary transplantation. In addition, survival following retransplantation is significantly worse with median survival post-transplant currently 3.1 years versus 6.2 years for primary transplantation ([3]Hachem 2024).

## COVERAGE POLICY

**All transplants require prior authorization from the Corporate Transplant Department. Solid organ transplant requests will be reviewed by the Corporate Senior Medical Director or qualified clinical designee. All other transplants will be reviewed by the Corporate Senior Medical Director or covering Medical Director. If the criteria are met using appropriate NCD and/or LCD guidelines, State regulations, and/or MCP policies the Corporate Senior Medical Director's designee can approve the requested transplant.**

*Office visits with participating Providers do NOT require prior authorization. Providers should see the Member in office visits as soon as possible and without delay. Failure to see the Member in office visits may be considered a serious quality of care concern.*

**Please see *MCP-459 Pre-Transplant and Transplant Evaluation* for pre-transplant criteria and transplant evaluation criteria that must be met prior to solid organ transplant.**

### Adult and Pediatric Criteria for Lung Transplantation

Lung transplantation is **considered medically necessary** when the Member meets **ALL** the following criteria:

1. Single, bilateral, or donor lobar lung organ transplantation from a deceased or a living donor **is considered medically necessary** in adult and pediatric members when **ALL** the following criteria are met:
    a. All transplant evaluation criteria are met
    b. Documentation that all medical, pharmaceutical, and surgical alternatives to lung transplant have been utilized if applicable that includes, but is not limited to:
        i. Oxygen therapy
        ii. Pulmonary rehabilitation
        iii. Lung volume reduction surgery for patients with chronic obstructive lung disease
    c. Living Donor lobar lung transplant requests require documentation supporting the Member's inability to survive the wait for a deceased donor allograft:

i. If donor lobar lung transplant is not performed, they may become ineligible for lung transplantation due to clinical deterioration
ii. Ambulatory with meeting requirements for receiving pulmonary rehab
iii. End stage pulmonary disease with a life expectancy < 18 months without a transplant
iv. No other serious systemic disease or condition affecting long term survival
v. No documented history of non-compliance

d. For multi-organ heart and lung transplant requests, criteria must be met for each organ requested (see individual policy for heart transplantation criteria)

2. Member meets disease specific criteria for **ONE** of the following conditions:
   a. **Chronic Obstructive Lung Disease:** (e.g., COPD, emphysema, alpha-1 antitrypsin disease, Bronchiolitis Obliterans Syndrome, bronchiectasis). Single or bilateral lung transplantation is indicated and guidelines for transplantation include:
      i. BODE index score of ≥ 7 measured by a six-minute walk test and the presence of **ONE** of the following:
         1. History of hospitalization for exacerbation of COPD associated with acute hypercapnia (partial pressure of carbon dioxide in arterial blood [$PaCO_2$] ≥ 50 mmHg)
         2. Three or more severe exacerbations within the preceding year
         3. Refractory dependence on noninvasive ventilatory assistance
         4. Forced expiratory volume in one second ($FEV_1$) < 20% of predicted, without reversibility
         5. Elevated $PaCO_2$ > 50 mmHg with progressive deterioration requiring long term oxygen therapy (defined as ≥ 6 months)
         6. Moderate to severe pulmonary hypertension (e.g., mean pulmonary artery pressure > 35 mmHg or mean right atrial pressure greater than 15 mmHg) or cor pulmonale despite oxygen therapy
   b. **Cystic Fibrosis:** Only bilateral lung transplantation is indicated and guidelines for transplantation include **ANY** of the following:
      i. Congenital pulmonary disease (e.g., pulmonary hypoplasia, bronchopulmonary dysplasia, surfactant disorders, hereditary hemorrhagic telangiectasia)
      ii. $FEV_1$ ≤ 30% of predicted value
      iii. Hypercapnia (defined as $PaCO_2$ ≥ 50 mmHg)
      iv. Increasing frequency of exacerbations requiring cycling antibiotic therapy
      v. Oxygen-dependent respiratory failure
      vi. Pulmonary hypertension (mean pulmonary artery pressure > 20 mmHg)
      vii. Refractory and/or recurrent pneumothorax
      viii. Refractory dependence on noninvasive ventilatory assistance
      ix. Rapid respiratory deterioration with $FEV_1$ < 30% with **ONE** of the following despite medical management:
         1. Increasing numbers of hospitalizations
         2. Exacerbation requiring ICU stay or mechanical ventilation
         3. Refractory or recurrent pneumothorax
         4. Recurrent hemoptysis not controlled by embolization
         5. Ongoing weight loss despite aggressive nutritional supplementation
   c. **Interstitial Lung Disease:** (e.g., interstitial pulmonary fibrosis and interstitial pneumonia). Single or bilateral lung transplantation is indicated and guidelines for transplantation include:
      i. Histologic or radiographic evidence of interstitial pulmonary fibrosis and **ANY** of the following:
         1. Symptomatic (e.g., oxygen desaturation with rest or exercise), progressive disease with failure to improve or maintain lung function despite standardized optimal therapy (e.g., supplemental oxygen, pulmonary rehabilitation)
         2. A 10% or greater decrease in forced vital capacity (FVC) during 6 months of follow-up
         3. Diffusion capacity for carbon monoxide (DLCO) < 40% of predicted or decline of 15% or more over 6 months
         4. Extensive reticulation or honeycomb change on CT scan
         5. Pulse oximetry less than 88% or distance less than 250 meters (820 feet) during 6-minute walk test
         6. Pulmonary hypertension (mean pulmonary artery pressure > 20 mmHg)
         7. Decrease in 6-minute walk test distance by more than 50 meters (165 feet) over 6 months

d. **Sarcoidosis:** Single or bilateral lung transplantation is indicated and guidelines for transplantation include**:**
   i. New York Heart Association (NYHA) functional class III or IV and **ANY** of the following:
      1. Pulmonary hypertension (mean pulmonary artery pressure > 20 mmHg)
      2. Hypoxemia at rest (partial pressure of oxygen in arterial blood [$PaO_2$] < 55 mmHg)
      3. Right atrial pressure > 15 mmHg
e. **Scleroderma:** Single or bilateral lung transplantation is indicated and guidelines for transplant include:
   i. FVC below 70% to 80% predicted at the time of diagnosis.
f. **Pulmonary Hypertension:** (e.g., idiopathic pulmonary hypertension). Bilateral lung transplantation is preferred and guidelines for transplantation include:
   i. No feasible pulmonary thromboendarterectomy for patients with chronic pulmonary thromboembolic disease
   ii. No successful control of pulmonary hypertension with pharmacogenic agents (e.g., calcium channel blockers or endothelin receptor antagonists)
   iii. **ANY** of the following:
      1. Persistent NYHA functional class III or IV despite maximal medical therapy for 3 months (e.g., combination therapy including prostanoids)
      2. Low (350 meter) six-minute walk test
      3. Cardiac index < 2 liters per minute per square meter
      4. Right atrial pressure >15 mmHg
      5. Mean pulmonary arterial pressure > 20 mmHg
      6. Refractory right heart failure (progressive renal insufficiency, increasing bilirubin, refractory ascites, increasing brain natriuretic peptide levels)
g. **Congenital Heart Disease:** (e.g., Eisenmenger syndrome). Single or bilateral lung transplantation is indicated and guidelines for transplant include **ALL** the following:
   i. NYHA functional class III or IV
   ii. Pulmonary hypertension
   iii. Severe progression of symptoms despite optimal medical management
h. **Pulmonary Langerhans Cell Histiocytosis, Lymphangioleiomyomatosis, and Eosinophilic Granuloma:** Single or bilateral lung transplantation is indicated, and guidelines include:
   i. NYHA functional class III or IV and **ANY** of the following:
      1. Severe impairment in lung function and exercise capacity ($VO_2$ max < 50%)
      2. Hypoxemia at rest ($PaO_2$ < 55 mmHg)
i. **Graft versus host disease** and **ANY** of the following:
   i. Progressive lung damage resulting in severe compromise of activities of daily living
   ii. Life expectancy limited by lung disease
j. **Other end-stage lung disease with Member not expected to recover without transplant (e.g., prolonged acute respiratory distress syndrome [ARDS] and acute fulminant lung damage including post-coronavirus 2019 [COVID-19] infection):** Bilateral lung transplantation is indicated.

## Adult and Pediatric Criteria for Heart - Lung Transplantation

A simultaneous heart and lung transplantation may be **considered medically necessary** when ALL the following criteria are met:

1. Severe refractory end stage heart failure

2. **ONE** of the following conditions:
   a. End-stage lung disease
   b. Irreversible pulmonary hypertension

3. **ONE** of the following conditions:
   a. Congenital heart disease with Eisenmenger syndrome
   b. Cystic fibrosis
   c. End-stage parenchymal lung disease with severely compromised left ventricular function (e.g., sarcoidosis)

## Adult and Pediatric Criteria for Retransplantation

Retransplantation **may be considered medically necessary** when **ALL** the following criteria are met:

1. Member is ambulatory
2. Member is ventilator independent
3. Member is free of significant co-morbidities
4. Member meets all other requirements for transplantation outlined above and have **ONE** of these indications:
    a. Non-function of the grafted organ
    b. Rejection refractory to immunosuppressive therapy
    c. Bronchiolitis obliterans (chronic rejection)
    d. Airway complications not correctable by other measures

## Limitations and Exclusions

**Requests for third or subsequent lung transplantation are considered not medically necessary and may not be authorized.**

**DOCUMENTATION REQUIREMENTS.** Molina Healthcare reserves the right to require that additional documentation be made available as part of its coverage determination; quality improvement; and fraud; waste and abuse prevention processes. Documentation required may include, but is not limited to, patient records, test results and credentials of the provider ordering or performing a drug or service. Molina Healthcare may deny reimbursement or take additional appropriate action if the documentation provided does not support the initial determination that the drugs or services were medically necessary, not investigational, or experimental, and otherwise within the scope of benefits afforded to the member, and/or the documentation demonstrates a pattern of billing or other practice that is inappropriate or excessive.

## SUMMARY OF MEDICAL EVIDENCE

### National and Specialty Organizations

The **Global Initiative for Chronic Obstructive Lung Disease (GOLD)** published *Global strategy for prevention, diagnosis and management of COPD: 2024 report* (GOLD 2024) for the diagnosis, management, and prevention of COPD. Recommendations specific to lung transplant referral include patients with COPD who "have progressive disease despite maximal medical treatment, are not candidates for lung volume reduction surgery, have a BODE index of 5 to 6, a $PaCO_2$ > 50 mg and/or $PaO_2$ < 60 mmHg and $FEV_1$ < 25%." GOLD recommends that patients be listed for transplant "when the BODE index is > 7, $FEV_1$ is < 15-20%, and they have had three or more severe exacerbations during the previous year, one severe exacerbation with hypercapnic respiratory failure, or have moderate to severe pulmonary hypertension." GOLD notes that over 70% of lung transplants in patients with COPD are bilateral lung transplants. Single lung transplants may be performed in patients with COPD but there are potential complications for the native lung, including hyperinflation (15-30% occurrence rate) and lung cancer (occurrence rate 5.2-6.1%). Additionally, patients with advanced emphysema may require lung volume reduction surgery or endoscopic lung volume reduction in the native lung to treat hyperinflation following single lung transplantation. Patients with advanced emphysema may also undergo lung volume reduction surgery or endoscopic lung volume reduction prior to lung transplantation to delay the need for transplant or optimize their condition prior to lung transplantation. Lung volume reduction surgery may be performed in one or both lungs.

The **European Society of Cardiology and European Respiratory Society (ESC/ERS)** published the *2022 ESC/ERS Guidelines for the Diagnosis and Treatment of Pulmonary Hypertension to propose management strategies for patients with pulmonary hypertension* (Humbert et al. 2022). The guidelines recommend evaluation for lung transplant referral in patients that are refractory to optimized medical therapy, present with an intermediate-high or high-risk of death and have a disease variant that responds poorly to medical therapy. The guidelines also recommend referral for progressive disease or recent hospitalization for worsening pulmonary hypertension, a need for intravenous or subcutaneous prostacyclin therapy, and signs of secondary liver or kidney dysfunction due to pulmonary hypertension or other potentially life-threatening complications. Heart-lung transplantation remains an option for those with significant cardiac and pulmonary dysfunction; however, heart-lung transplantation remains limited by the availability of organs and the complexities of a combined heart-lung transplantation. The ESC/ERS states that connective tissue disease is not a

contraindication for lung transplantation.

The **International Society for Heart and Lung Transplantation (ISHLT)** published the *Consensus Document for the Selection of Lung Transplant Candidates* (Leard et al. 2021) which provides guidance regarding timely referral, assessment, optimization, and listing of potential lung transplant candidates. The 2021 report highlights how comorbidities and other risk factors interact to affect post-transplant survival benefit. The guidelines also list absolute contraindications and notes that "most lung transplant programs should not transplant patients with [those] risk factors except under very exceptional or extenuating circumstances." The ISHLT agrees that lung transplantation improves survival and quality of life. However, the panel acknowledges that when making recommendations about allocating a scarce resource, survival benefit should be prioritized.

The **Cystic Fibrosis Foundation** published consensus guidelines for lung transplant referral for individuals with CF (Ramos et al. 2019). The following are indications for lung transplant referral:

- Patients that have an $FEV_1$ < 50% of predicted.
- Patients that have an $FEV_1$ < 40% predicted and > 2 exacerbations per year requiring intravenous antibiotics or 1 exacerbation requiring positive pressure ventilation regardless of $FEV_1$.
- Those ≥ 18 years of age who have an $FEV_1$ < 50% of predicted and rapidly declining (>20% relative decline within 12 months) **or** $FEV_1$ < 40% of predicted with markers of shortened survival **or** $FEV_1$ < 30% of predicted.
- Those < 18 years of age who have an $FEV_1$ < 50% of predicted and rapidly declining (>20 relative decline within 12 months) **or** $FEV_1$ is < 50% of predicted with markers of shortened survival **or** $FEV_1$ < 40% of predicted.
- Adults with a body mass index < 18 and FEV1 < 40% predicted while concurrently working to improve nutritional state.
- Patients with an $FEV_1$ < 40% predicted and massive hemoptysis requiring intensive care admission or bronchial artery embolization.
- Patients with an $FEV_1$ < 40% and a pneumothorax.

Markers of shortened survival include a 6-minute walk test distance < 400 meters, hypoxemia at rest or with exertion, hypercarbia ($PaCO_2$ > 50 mmHg) on arterial blood gas, and pulmonary hypertension on echocardiogram (PA systolic pressure > 50 mmHg) or evidence of right ventricular dysfunction in the absence of a tricuspid regurgitant jet. The Cystic Fibrosis Foundation also recommends ensuring modifiable barriers, such as nutritional status, diabetes management, physical inactivity or deconditioning, adherence behaviors, mental health issues, substance use, and psychosocial factors be addressed preemptively to optimize transplant candidacy.

**National Institute for Health and Clinical Excellence (NICE)**

The 2018 guidance *Chronic Obstructive Pulmonary Disease in Over 16s: Diagnosis and Management (NG115)* provides guidance for COPD as well as emphysema and chronic bronchitis in individuals aged 16 and older. A goal is to help providers diagnose patients earlier to obtain optimal benefit from treatments and improve quality of life.

Two major recommendations are also included (NICE 2018). First, referrals should be considered to a multidisciplinary team including lung transplantation specialists for patients who:

- Have severe COPD with $FEV_1$ less than 50% and breathlessness that affects their quality of life despite optimal medical treatment
- Are non-smokers
- Have completed pulmonary rehabilitation
- Do not have contraindications for transplantation such as comorbidities or frailty

Secondly, NICE recommends that providers do not use previous lung volume reduction procedures as a reason not to refer a person for assessment for lung transplantation. This was modified previously as evidence demonstrated that those with severe COPD had improvements in lung function, exercise capacity, quality of life, and long-term mortality as a result of lung volume reduction surgery.

NICE also published guidelines in 2017 for the diagnosis and management of interstitial pulmonary fibrosis (NICE 2017). Recommendations specific to lung transplantation include discussing transplantation as a treatment option between 3-6 months after diagnosis unless clinical condition warrants earlier referral and provided there are no absolute contraindications to transplant.

## SUPPLEMENTAL INFORMATION

### Pulmonary Function Tests

PFTs are performed to assess pulmonary function in patients that have a history, risk factors, or symptoms of lung disease. There are numerous forms of PFTs that may be performed in outpatient or inpatient settings. The outpatient setting is generally preferred as a greater range of testing may be performed with better accuracy. Results from PFTs are interpreted to determine the degree and type of physiologic impairment (restrictive or obstructive). The most common types of PFTs are spirometry, pre- and post-bronchodilator spirometry, lung volumes, and diffusing capacity for carbon monoxide. The most common measurements are (Kaminsky 2023):

- **FVC**: total exhaled volume after maximum inspiration.
- **$FEV_1$**: the total volume exhaled in the first second of forceful and complete exhalation.
- **$FEV_1$/FVC**: the proportion of a person's vital capacity they can expire in the first second of forced expiration to the full FVC.
- **DLCO**: measurement of the diffusing capacity of carbon monoxide in a single breath.

### New York Heart Association (NYHA) Functional Classification

NYHA classification has served as a vital tool for risk stratification of heart failure and for determining clinical trial eligibility and medication and device candidate eligibility (AHA 2023):

- **Class I**: Individuals with cardiac disease but without resulting limitation of physical activity; ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain; symptoms only occur on severe exertion.
- **Class II**: Individuals with cardiac disease resulting in slight limitation of physical activity; they are comfortable at rest; ordinary physical activity (e.g., moderate physical exertion such as carrying shopping bags up several flights of stairs) results in fatigue, palpitation, dyspnea, or anginal pain.
- **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 or anginal pain.
- **Class IV**: Individuals with cardiac disease resulting in inability to carry on any physical activity without discomfort; symptoms of HF or the anginal syndrome may be present even at rest; if any physical activity is undertaken, discomfort is increased.

### BODE Index

The BODE index is a measurement to assess risk of mortality in patients with COPD and uses the following factors as indicators: weight (BMI), airway obstruction (FEV1), dyspnea, and exercise capacity. Factors are calculated together, and the approximate four-year survival interpretation is: 0-2 = 80%, 3-4 = 67%, 5-6 = 57%, 7-10 = 18% ([2]Hachem 2024).

## CODING & BILLING INFORMATION

**CPT (Current Procedural Terminology)**

| Code | Description |
|---|---|
| **32850** | Donor pneumonectomy(s) (including cold preservation), from cadaver donor |
| **32851** | Lung transplant, single; without cardiopulmonary bypass |
| **32852** | Lung transplant, single; with cardiopulmonary bypass |
| **32853** | Lung transplant, double (bilateral sequential or en bloc); without cardiopulmonary bypass |
| **32854** | Lung transplant, double (bilateral sequential or en bloc); with cardiopulmonary bypass |
| **32855** | Backbench standard preparation of cadaver donor lung allograft prior to transplantation, including dissection of allograft from surrounding soft tissues to prepare pulmonary venous/atrial cuff, pulmonary artery, and bronchus; unilateral |
| **32856** | Backbench standard preparation of cadaver donor lung allograft prior to transplantation, including dissection of allograft from surrounding soft tissues to prepare pulmonary venous/atrial cuff, pulmonary artery, and bronchus; bilateral |

| | |
|---|---|
| **33935** | Heart-lung transplant with recipient cardiectomy-pneumonectomy |
| **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 |

**HCPCS (Healthcare Common Procedure Coding System)**

| Code | Description |
|---|---|
| **S2060** | Lobar lung transplantation |
| **S2061** | Donor lobectomy (lung) for transplantation, living donor |
| **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 pre- and post-transplant care in the global definition |

**CODING DISCLAIMER.** Codes listed in this policy are for reference purposes only and may not be all-inclusive. Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement. Listing of a service or device code in this policy does not guarantee coverage. Coverage is determined by the benefit document. Molina adheres to Current Procedural Terminology (CPT®), a registered trademark of the American Medical Association (AMA). All CPT codes and descriptions are copyrighted by the AMA; this information is included for informational purposes only. Providers and facilities are expected to utilize industry standard coding practices for all submissions. When improper billing and coding is not followed, Molina has the right to reject/deny the claim and recover claim payment(s). Due to changing industry practices, Molina reserves the right to revise this policy as needed.

## APPROVAL HISTORY

| | |
|---|---|
| **10/09/2024** | Policy reviewed. No changes to coverage criteria. |
| **06/12/2024** | Coverage criteria revised with removal of transplant evaluation, continuation of therapy, and general contraindication coverage criteria as it is now stipulated in MCP 459 Pre-Transplant and Transplant Evaluation. |
| **10/12/2023** | Policy reviewed, changes to criteria include addition of other end-stage lung disease criteria, removed rapid fall in FEV1 for CF, age for colonoscopy reduced to 45 years, addition of non-life limiting neurological impairment criteria, removal of abnormal serology criteria and cannabis use section, and added substance use/vaping/smoking/inhaled cannabis to absolute contraindications. Updated Overview, Summary of Medical Evidence, and References. IRO Peer Review on September 22, 2023, by a practicing, board-certified physician with specialties in Surgery, Vascular Surgery, and Surgical Critical Care. |
| **10/12/2022** | Policy reviewed, no changes to criteria, included section on marijuana use, updated Overview and Summary of Medical Evidence. |
| **10/13/2021** | Policy reviewed, no criteria updates, updated references. Coding section updated; removed CPT codes 33930, 33933, 33935. |
| **09/16/2020** | Policy updated with additional disease specific criteria for COPD, cystic fibrosis, congenital heart disease, interstitial lung disease, PAH, PLCH, and graft vs. host disease, updated references. |
| **09/18/2019** | Policy reviewed, no changes. |
| **09/13/2018** | Policy reviewed, no changes. |
| **06/22/2017** | Policy reviewed, no changes. |
| **09/15/2016** | Policy reviewed, no changes. |
| **04/27/2015** | Policy updated with new pretransplant criteria; Summary of Medical Evidence section condensed. Added one new indication for individuals with scleroderma. |
| **08/30/2012** | New policy. |

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