

Subject: Lung Transplantation		Original Effective Date: 8/30/12
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PREFACE

This Medical Guidance is intended to facilitate the Utilization Management process. 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 (i.e., 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 exclusions 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 following website: <http://www.cms.hhs.gov/center/coverage.asp>.

FDA INDICATIONS

Lung transplantation is a procedure not subject to FDA regulation.

CENTERS FOR MEDICARE AND MEDICAID SERVICES (CMS)

The coverage directive(s) and criteria from an existing National Coverage Determination (NCD) or Local Coverage Determination (LCD) will supersede the contents of this Molina medical coverage guidance (MCG) document and provide the directive for all Medicare members. The directives from this MCG document may be followed if there are no available NCD or LCD documents available and outlined below.

CMS does not have a NCD or LCD for lung transplantation¹. According to CMS, a transplant program is defined as a component within a transplant hospital that provides transplantation of a particular type of organ. Medicare Conditions of Participation for organ transplant programs were established on March 30, 2007, and became effective on June 28, 2007. 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.

INITIAL COVERAGE CRITERIA

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. If the criteria are met using appropriate NCD and/or LCD guidelines, state regulations and/or MCG policies the Corporate Senior Medical Director's designee can approve the requested transplant. Members under age 12 must meet UNOS guidelines for Priority 1 status or members over age 12 must meet UNOS guidelines for lung allocation score (LAS) and the diagnosis must be made by a Pulmonologist and or Transplant Surgeon.

Pre-Transplant Evaluation:

General requirements for transplant evaluation include all of the following^{2,4,5,16}:

- History and physical examination
- Psychosocial evaluation and clearance:** This must be completed and documentation submitted for review before any additional transplant work-up or testing is initiated.
- Dietary consult and clearance for transplant
- Chest x-ray
- ECG
- Echocardiogram
- Pulmonary function tests
- Ventilation perfusion scan
- CT scan chest and thorax
- Right heart catheterization
- Coronary artery disease screening: chronic smokers, > 50 years age, and those with a clinical or family history of heart disease or diabetes should be evaluated with testing in the following order:
 1. dobutamine stress echocardiography and if positive
 2. cardiac catheterization
- Physical therapy assessment including 6 minute walk test
- Lab studies:
 - Complete blood cell count, comprehensive metabolic panel, coagulation profile
 - HIV testing
 - Hepatitis A (HAV), Hepatitis B (HBV), Hepatitis C (HCV), Hepatitis D, Epstein-Barr virus (EBV), cytomegalovirus (CMV) serology, syphilis, toxoplasmosis, herpes simplex virus
 - Arterial blood gases
 - Sputum culture
 - HLA Antibody
 - Blood type
 - Urinalysis, 24 hour creatinine clearance

Within the last 12 months the following may be required:

- Colonoscopy (if indicated or > age 50) with removal of any polyps
- Dental examination within last 12 months: Contact Plan for Coverage Criteria
- GYN examination with Pap smear (if indicated or > age 18) with complete workup and treatment of abnormal results as indicated
- Immunizations up to date when indicated: Hepatitis A and Hepatitis B, pneumococcal vaccine, influenza vaccine, tetanus booster)
- Mammogram (if indicated or > age 40) with complete workup and treatment of abnormal results as indicated
- Osteoporosis screening with DEXA scan: [ONE]

- indicated for cholestatic disorders
- prolonged corticosteroid therapy
- postmenopausal women
- > age 65
- ❑ Peripheral artery disease (PAD) screening with doppler-recorded ankle-brachial index: [ONE]
 - age > 50
 - history of diabetes or smoking
- ❑ Testicular examination > age 50

Adult and Pediatric Criteria:

1. Single, double, or donor lobar lung transplantation may be authorized in adults and children when all of the following criteria are met^{3 8 10}:

- ❑ Documentation that all medical, pharmaceutical and surgical alternatives to lung transplant have been utilized that includes but is not limited to the following:
 - Oxygen therapy
 - Pulmonary rehabilitation (Refer to Pulmonary Rehabilitation for Chronic Pulmonary Diseases MCG-086)
 - Lung volume reduction surgery for patients with chronic obstructive lung disease
- ❑ Donor lobar lung transplant requests require documentation supporting the member's inability to survive the wait for a deceased donor allograft
 - If donor lobar lung transplant is not performed they may become ineligible for lung transplantation due to clinical deterioration
 - Ambulatory with meeting requirements for receiving pulmonary rehab
 - End stage pulmonary disease with a life expectancy < 18 months without a transplant
 - No other serious systemic disease or condition affecting long term survival
 - No documented history of non-compliance
- ❑ If HIV positive all of the following are met¹⁵:
 - CD4 count >200 cells/mm³ for >6 months
 - HIV-1 RNA undetectable
 - On stable anti-retroviral therapy >3 months
 - No other complications from AIDS (e.g., opportunistic infection, including aspergillus, tuberculosis, coccidioides mycosis, resistant fungal infections, Kaposi's sarcoma, or other neoplasm)
 - Meeting all other criteria for transplantation
- ❑ None of the following **absolute contraindications** are present:
 - Active alcohol and/or other substance abuse including smoking (to remove as a contraindication there must be 6 months of documented abstinence through participation in a structured alcohol/substance abuse program with regular meeting attendance and negative random drug testing)
 - Active Malignancy within 2 years of the evaluation not including skin cancers
 - AIDS (CD4 count < 200cells/mm³)

- Documented history of non-compliance or inability to follow through with medication adherence or office follow-up
- Irreversible brain damage
- Non-ambulatory with limited rehabilitation potential
- Primary or metastatic malignancies of the respiratory or intrathoracic organs
- Significant or advanced heart, liver, kidney, gastrointestinal or other systemic or multi-system disease that is likely to contribute to a poor outcome after lung transplantation
- Systemic and/or uncontrolled infection including chronic active viral hepatitis B, hepatitis C
- Significant chest wall or spinal deformity requires transplant surgeon acknowledgement and clearance
- No behavioral health disorder by history or psychosocial issues: [One]
 - ◇ if history of behavioral health disorder, no severe psychosis or personality disorder
 - ◇ mood/anxiety disorder must be excluded or treated
 - ◇ member has understanding of surgical risk and post procedure compliance and follow-up required

Note: Patient's need to understand the importance of adherence to medication schedules and follow-up appointments/noncompliance is a major cause of graft failure.

- No adequate social/family support
- For multi-organ heart and lung transplant requests, criteria must be met for each organ requested see individual policy for heart transplantation criteria.

2. **Disease specific criteria includes all of the following**^{3 8 10 12 13 21 22 23}.

- **Chronic Obstructive Lung Disease:** (e.g., COPD, Emphysema, Alpha-1 antitrypsin disease, Bronchiolitis obliterans syndrome (BOS), Bronchiectasis): Single or double lung transplantation is indicated and guidelines for transplantation include:
 - BODE index* score of > 5 measured by a six-minute walk test should be referred for transplant evaluation and **ONE** of the following;
 - ◇ History of hospitalization for exacerbation of COPD associated with acute hypercapnia ($PCO_2 \geq 50$ mmHg);
 - ◇ FEV1 (i.e., forced expiratory volume in the first second) < 25% of predicted, without reversibility;
 - ◇ elevated $PaCO_2 > 50$ mm hg with progressive deterioration requiring long term oxygen therapy (defined as ≥ 6 months¹⁸);
 - ◇ Pulmonary hypertension (mean pulmonary artery pressure > 20 mm Hg) and/or cor pulmonale, despite oxygen therapy

*BODE index²¹ 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. These factors are calculated together and the approximate 4 year survival interpretation is:

0-2 = 80%
3-4 = 67%
5-6 = 57%
7-10 = 18%

- ❑ **Cystic fibrosis:** Only double lung transplantation is indicated and guidelines for transplantation include ANY of the following:
 - FEV1 \leq 30% of predicted value;
 - increasing frequency of exacerbations requiring cycling antibiotic therapy;
 - refractory and/or recurrent pneumothorax;
 - oxygen-dependent respiratory failure;
 - hypercapnia (defined as $\text{PCO}_2 \geq 50$ mm hg);
 - pulmonary hypertension (mean pulmonary artery pressure > 20 mm hg);
 - rapid respiratory deterioration with FEV1 $> 30\%$ with **one** of the following despite medical management: [ONE]
 - ◇ Increasing numbers of hospitalizations
 - ◇ rapid fall in FEV1
 - ◇ recurrent massive hemoptysis
 - ◇ increasing cachexia

- ❑ **Interstitial lung disease:** (e.g., Idiopathic pulmonary fibrosis (IPF) and Interstitial pneumonia): Single or double lung transplantation is indicated and guidelines for transplantation include:
 - Histologic or radiographic evidence of IPF and **ANY** of the following:
 - ◇ 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);
 - ◇ a 10% or greater decrease in FVC (i.e., forced vital capacity) during six months of follow-up;
 - ◇ diffusion capacity for carbon monoxide (DLCO) $< 39\%$ predicted;
 - ◇ honeycombing on computerized tomography;
 - ◇ decrease in pulse oximetry $< 88\%$ during a six-minute walk test

- ❑ **Sarcoidosis:** Single or double transplantation is indicated and guidelines for transplantation include:
 - New York Heart Association (NYHA) functional class III or IV* and **ANY** of the following:
 - ◇ pulmonary hypertension (mean pulmonary artery pressure > 20 mm Hg) ;
 - ◇ hypoxemia at rest ($\text{PaO}_2 < 55$ mm hg);
 - ◇ right atrial pressure > 15 mm Hg

- ❑ **Pulmonary Arterial Hypertension (PAH)** (includes idiopathic pulmonary hypertension or IPH): Double lung transplantation is preferred and guidelines for transplantation include:
 - No feasible pulmonary thromboendarterectomy for patients with chronic pulmonary thromboembolic disease **AND**;
 - No successful control of pulmonary arterial hypertension with pharmacogenic agents (e.g.; calcium channel blockers or endothelin receptor antagonists); **AND ANY** of the following:

- ◇ persistent NYHA functional class III or IV* on maximum medical therapy;
- ◇ low (350 meter) six-minute walk test;
- ◇ cardiac index < 2 liters per minute per square meter;
- ◇ right atrial pressure >15 mm Hg;
- ◇ mean pulmonary arterial pressure > 20 mm hg

- ❑ **Congenital Heart Disease** (e.g., Eisenmenger syndrome)²²: Single or double transplantation is indicated and guidelines for transplant include both of the following:
 - severe progression of symptoms despite optimal medical management (Refer to UpToDate for treatment options for specific indication);
 - NYHA functional class III or IV*
- ❑ **Lymphangioleiomyomatosis (LAM) and Eosinophilic Granuloma**: Single or double transplantation is indicated and guidelines for transplantation include:
 - NYHA functional class III or IV* and **ANY** of the following:
 - ◇ severe impairment in lung function and exercise capacity (VO₂ max < 50%);
 - ◇ hypoxemia at rest (PaO₂ < 55 mm hg)
- ❑ **Retransplantation**²⁴⁻²⁵: When retransplantation is being considered **ALL** of the following factors must be present:
 - the member must be ambulatory
 - ventilator independent
 - free of significant co-morbidities
 - meet all of the other requirements for transplantation outlined above **AND** have one of these indications:
 - ◇ non-function of the grafted organ
 - ◇ rejection refractory to immunosuppressive therapy
 - ◇ bronchiolitis obliterans (chronic rejection)
 - ◇ airway complications not correctable by other measures

NOTE: Requests for third or subsequent lung transplantation are not covered

***NYHA Functional Classification is defined as:**

I	Patients without resulting limitations of physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain.
II	Patients with a slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity results in fatigue, palpitation, dyspnea, or anginal pain.
III	Patients with marked limitation of physical activity. They are comfortable at rest. Less than ordinary physical activity causes fatigue, palpitation, dyspnea, or anginal pain.
IV	Patient with the inability to carry on any physical activity without discomfort. Symptoms of cardiac insufficiency or of the anginal syndrome may be present even at rest. If any physical activity is undertaken,

discomfort is increased.

3. **Heart and Lung Transplantation**²⁷: A simultaneous heart and lung transplantation may be authorized when all of the following criteria are met: **[ALL]**

- severe refractory end stage heart failure; AND
- end-stage lung disease; OR
- irreversible pulmonary hypertension; AND
- one of the following conditions: **[ONE]**
 - Congenital heart disease with Eisenmenger syndrome
 - Cystic fibrosis
 - end-stage parenchymal lung disease with severely compromised left ventricular function (e.g., sarcoidosis)

CONTINUATION OF THERAPY

When extension of a previously approved transplant authorization is requested, review using updated clinical information is appropriate.

- If Molina Healthcare has authorized prior requests for transplantation, the following information is required for medical review: **[ALL]**
 - Presence of no absolute contraindications as listed above;
 - History and physical within the last 6 months;
 - Chest CT scan within the last 12 months;
 - ECHO within the last 12 months;
 - Right heart catheter (for members with any pulmonary hypertension at original listing and/or if ECHO results do not correlate with right heart cath pressure measurements) within the last 12 months;
 - Left heart catheter (for members with abnormalities identified on initial study or if clinically indicated) within the last 12 months;
 - LE Venous duplex (for members with history of DVT, current risk factors for DVT, and/or those unable to perform activities of daily living without total assistance);
 - DEXA scan within the last 12 months;
 - Laboratory studies: CBC, blood chemistries, Hep A, B, and C, HIV, RPR, 24 hour urine within the last 12 months;
 - Mammogram within the last 12 months;
 - Chest x-ray (if clinically indicated) within the last 12 months;
 - Dental examination within the last 12 months;
 - Social work evaluation within the last 12 months;
 - Psychosocial evaluation or update within the last 12 months;

- Per initial and updated history and physical, any other clinically indicated tests and/or scans as determined by transplant center physician or Molina Medical Director.

- If authorized prior requests for transplantation were obtained from another insurer, the following information is required for medical review: [ALL]
 - Authorization letter/documentation from previous insurer;
 - Presence of no absolute contraindications as listed above;
 - History and physical within the last 6 months;
 - Chest CT scan within the last 12 months;
 - ECHO within the last 12 months;
 - Right heart catheter (for members with any pulmonary hypertension at original listing and/or if ECHO results do not correlate with right heart cath pressure measurements) within the last 12 months;
 - Left heart catheter (for members with abnormalities identified on initial study or if clinically indicated) within the last 12 months;
 - LE Venous duplex (for members with history of DVT, current risk factors for DVT, and/or those unable to perform activities of daily living without total assistance);
 - DEXA scan within the last 12 months;
 - Laboratory studies: CBC, blood chemistries, Hep A, B, and C, HIV, RPR, 24 hour urine within the last 12 months;
 - Mammogram within the last 12 months;
 - Chest x-ray (if clinically indicated) within the last 12 months;
 - Dental examination within the last 12 months;
 - Social work evaluation within the last 12 months;
 - Psychosocial evaluation or update within the last 12 months;
 - Per initial and updated history and physical, any other clinically indicated tests and/or scans as determined by transplant center physician or Molina Medical Director.

COVERAGE EXCLUSIONS

The following **absolute** contraindications^{3 8 10} to lung transplantation are not covered:

- Active alcohol and/or other substance abuse including smoking (to remove as a contraindication there must be 6 months of documented abstinence through participation in a structured alcohol/substance abuse program with regular meeting attendance and negative random drug testing)
- Active Malignancy within 2 years of the evaluation not including skin cancers
- AIDS (CD4 count < 200cells/mm³)
- Documented history of non-compliance or inability to follow through with medication adherence or office follow-up
- Irreversible brain damage
- Non-ambulatory with limited rehabilitation potential
- Primary or metastatic malignancies of the respiratory or intrathoracic organs
- Significant or advanced heart, liver, kidney, gastrointestinal or other systemic or multi-system disease that is likely to contribute to a poor outcome after lung transplantation

- Systemic and/or uncontrolled infection including chronic active viral hepatitis B, hepatitis C
- Significant chest wall or spinal deformity requires transplant surgeon acknowledgement and clearance
- No behavioral health disorder by history or psychosocial issues: [One]
 - if history of behavioral health disorder, no severe psychosis or personality disorder
 - mood/anxiety disorder must be excluded or treated
 - member has understanding of surgical risk and post procedure compliance and follow-up required

Note: Patient's need to understand the importance of adherence to medication schedules and follow-up appointments/noncompliance is a major cause of graft failure.

- No adequate social/family support

Relative contraindications to lung transplantation include any of the following^{3 8 10}:

- Symptomatic osteoporosis
- Severe musculoskeletal disease
- Current use of corticosteroids >20 mg daily prednisone (or the equivalent)
- BMI <17 kg/m² or >30 kg/m²
- Colonization with highly resistant or highly virulent bacteria, fungi, or mycobacteria
- Psychosocial instability associated with the inability to cooperate or comply with medical therapy
- Other medical conditions that have not resulted in end-stage organ damage, such as diabetes mellitus, systemic hypertension, peptic ulcer disease, or gastroesophageal reflux should be optimally treated before transplantation. Patients with coronary artery disease may undergo percutaneous intervention before transplantation or coronary artery bypass grafting

DESCRIPTION OF PROCEDURE/SERVICE/PHARMACEUTICAL

Lung transplantation is a surgical procedure to replace one or both diseased lungs with a healthy lung or lungs from a donor. There are different types of lung transplantation, including a lobe transplant, a single lung transplant, a double lung transplant, or a heart-lung transplant. Lung transplantation has become a viable treatment option for selected patients with end-stage lung disease due to a wide variety of underlying disorders. Single, double, and lobar-lung transplantation have all been performed successfully. Living donor lobar lung transplantation has shown success and addresses the shortage of cadaveric organs. Single lung transplantation is most effective for patients with end-stage pulmonary fibrosis, double lung transplantation is most effective in patients with end-stage chronic obstructive pulmonary disease (COPD) and cystic fibrosis and lobar lung transplantation is usually reserved for pediatric patients who are not expected to survive the waiting time for cadaveric transplant. The most common indications for pediatric lung transplantation are cystic fibrosis with end stage lung disease, pulmonary hypertension and pulmonary fibrosis.

The goal of lung transplantation is to improve quality of life and long-term survival in patients with end-stage pulmonary disease. Advances in donor and recipient selection, new immunosuppressive medications, new and improved surgical techniques, and increased medical management of infections have improved the overall survival in patients after lung transplantation.

A heart-lung transplant is a procedure where the transplantation of one or both lungs and heart from a single cadaver donor is done. A combined heart-lung transplant is intended to prolong survival and improve function in recipients with end stage cardiopulmonary disease. The surgical technique requires a coordinated triple operative procedure that includes the procurement of a donor heart-lung block, surgical removal of the heart and lungs of a single cadaver donor, and implantation of the heart and lungs into the recipient.

The United Network for Organ Sharing (UNOS) has developed a system for prioritizing patients for lung transplants called the Lung Allocation Score System or LAS¹². Every transplant candidate age 12 and older will receive a lung allocation score from 0-100. The LAS is calculated from the following medical information: diagnosis, comorbidities such as diabetes, test results that include: forced vital capacity, pulmonary artery and capillary wedge pressures, oxygen at rest, age, body mass index, functional status, six minute walk, serum creatinine and if assisted ventilation is required. Waiting time for transplant plays a more significant role in lung transplant candidates under the age of 12. The candidate with the highest LAS in a particular age group will receive first priority for a donor lung offer. Because of unique circumstances in the pediatric population transplant candidates under the age of 12 are assigned a priority status. Those that meet UNOS criteria (respiratory failure or pulmonary hypertension) are assigned Priority 1 status and those that do not meet criteria are assigned Priority 2.

The goal of the pretransplant evaluation is to assess the ability of a patient to tolerate the surgery, post-operative immunosuppression, and transplant care. An extensive cardiopulmonary evaluation, screening for occult infection or cancer, and psychosocial evaluation is standard. Specific testing varies depending upon the patient's age, medical history, and transplant center practice. In addition, while a certain battery of tests may initiate the work up, more testing may be indicated depending upon the condition of the patient or the initial test results. The cardiopulmonary evaluation is intended to evaluate for any significant coronary artery or valvular disease, cardiomyopathy, obstructive or restrictive lung disease, and pulmonary hypertension. In addition to a standard medical evaluation the initial assessment should include a psychological and social support evaluation to identify issues that may impair a successful outcome after transplantation. These include a lack of information about the nature of the transplant procedure and post-transplant care, drug or alcohol dependence, compliance with complex medical and behavior regimens. The assessment includes education of the family and the support network of the patient because compliance with complex medical and behavior treatment is critical after any organ transplant procedure. Recipients must be able to incorporate complicated medications, follow-up appointments, and frequent laboratory visits into their schedules. Having an adequate support network aware of these requirements will encourage patient compliance and long-term success.

The donor selection criteria may vary from center to center, but the general criteria include:

- Cytomegalovirus (CMV) compatibility
- Age < 65 years
- Absence of severe chest trauma or infection
- Minimal pulmonary secretions
- Negative screens for human immunodeficiency virus (HIV) and hepatitis C and B
- Blood type compatibility
- Partial pressure of oxygen in arterial blood (PaO₂) > 300 mm Hg (millimeters of mercury) on 100% fraction of inspired oxygen
- Clear chest radiograph

- No history of a malignant neoplasm
- A donor lung within 25% to 30% of the predicted size of the recipient's lungs

Procedures^{4 5 17 20}:

Single Lung Transplantation: The operation begins when the donor lung arrives in the operating room. A single lung transplant requires 4 to 8 hours. A history of prior chest surgery may complicate the procedure and require additional time. The lung with worse pulmonary function is chosen for replacement. If both lungs function equally, then the right lung is usually favored for removal, because that avoids having to maneuver around the heart, as would be required for excision of the left lung. Single lung transplants are usually done through an incision extending from under the shoulder blade around the chest, ending near the sternum. An alternative method is an incision under the breastbone. Following excision of the native lung, the donor lung is wrapped in sponges soaked with a cold crystalloid solution and placed into the hemithorax. The bronchial anastomosis is performed first. The length of both the donor and recipient bronchi is minimized to preserve collateral blood supply and to achieve some degree of anastomotic overlap. After completion of the anastomoses, the lung is reinflated and a bronchoscopy is performed to clear remaining blood and mucus from the new lung. When the surgeon is satisfied with the performance of the lung, the chest incision is closed. Single lung transplants are most commonly performed in patients with idiopathic pulmonary fibrosis.

Double Lung Transplantation: A double lung transplant, also known as a bilateral transplant, can be performed sequentially, en bloc, or simultaneously. The most frequently performed double lung transplantation procedure is bilateral sequential single lung transplantation. The procedure requires 6 to 12 hours. For double lung transplants, an incision, known as a clamshell incision, is made across the entire chest, just below the breasts. Mobilization and pneumonectomy of the native lung and the implantation of the lung graft are conducted in the same manner as described for single lung transplantation. Removal of both lungs is mandatory in patients with end stage cystic fibrosis therefore double lung transplants are preferred. In patients with pulmonary artery hypertension double lung transplantation is preferred.

Living Donor Lobar Transplantation: 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 objectives of the surgery are to improve functional status and quality of life and 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. While the donor lobectomies are taking place, the recipient undergoes pneumonectomy in another operating suite. Surgery is usually performed through a unilateral or bilateral transverse thoracosternotomy incision, for implantation of one or two lobes, respectively. The majority of living donor lung recipients are patients with cystic fibrosis and the majority of 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, as there is a shortage of suitable cadaveric donors for this age group.

Post operatively, transplant rejection is a primary concern, both immediately after surgery and continuing throughout the patient's life. Signs of rejection are fever, flu-like symptoms, increased difficulty breathing, worsening pulmonary test results, increased chest pain or tenderness, or an increase or decrease in body weight > 2 kilograms (kg) per 24 hours. To prevent transplant rejection and subsequent damage to the new lung or lungs, patients must commit to a lifelong regimen of immunosuppressive drugs. Treatment of chronic rejection is the most difficult issue following lung transplantation. Transplant patients are vulnerable to infections. Antibiotics may be prescribed to treat or prevent infections. Certain medications may also have side effects or trigger allergic reactions. Close follow-up care is required to balance the benefits and potential risks of the drugs. The early postoperative period is the first 3 months following transplantation. Chest x-rays are performed according to the patient's clinical status. Spirometry is done after surgery, at predischarge, and periodically thereafter. Fiberoptic bronchoscopy and bronchoalveolar lavage are performed if the patient demonstrates new infiltrates on chest radiographs, a decrease in lung function on spirometry, or the presence of new symptoms. Depending on the center, routine transbronchial lung biopsy in asymptomatic patients with stable lung function is performed. Late monitoring begins after the third month following transplantation and includes mainly monitoring and follow-up of signs of chronic rejection.

Management of patients who have end-stage lung disease and who are waiting for a suitable donor depends on the cause of lung disease. Medical management includes the following:

- lung volume reduction surgery
- oxygen therapy
- pulmonary rehabilitation
- treatment of any reversible airway disease
- vasodilators
- pulmonary thromboendarterectomy in patients with chronic pulmonary thromboembolic disease

GENERAL INFORMATION

Summary of Medical Evidence

Hayes, Cochrane, UpToDate, MD Consult etc.

A Hayes Medical Technology Directory Report for lung transplantation⁴ indicates that a substantial number of the recipients will derive a survival advantage from lung transplant with improvement in quality of life (QOL). Evidence was controversial as to which diagnostic group of patients benefited the most in terms of 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 adverse effects. Therefore transplantation will improve survival and QOL, but at the same time, it will introduce new restrictions and complications.

Lung transplantation is indicated for carefully selected patients with end-stage lung disease from cystic fibrosis, obstructive lung diseases, or immunodeficiency disorders, who have no other medical or surgical treatment options, and who meet the established general and disease-specific criteria for cadaver lung transplantation. There is less evidence for lung transplantation in patients with end-stage lung disease from restrictive lung diseases and pulmonary vascular disease. Lung transplantation is not indicated in patients with contraindications to lung transplantation.

A Hayes Medical Directory Report for living donor lobar lung transplantation⁵ indicates evidence from the available, peer-reviewed published studies suggests that living donor lobar lung transplantation is a reasonable treatment option for carefully selected patients with end-stage lung disease who are unlikely to survive or who may deteriorate clinically to the point of transplant ineligibility during the wait for a compatible cadaveric donor but who are otherwise eligible candidates for unilateral or bilateral lung transplantation. The surgery provides health benefits by improving respiratory and cardiac function and quality of life, and by prolonging survival in patients who otherwise are likely to die. While a number of recipients experience complications or die, the likelihood of survival without transplant is extremely low. There is some evidence that living donor lobar lung transplants may be more efficacious than cadaveric lung transplants for certain patients, leading to greater improvement in respiratory function, and that the incidence of chronic rejection is lower than that for cadaveric transplantation.

Living donor lobar lung transplantation is indicated for patients with end-stage lung disease with no other medical or surgical treatment options and who meet the criteria for cadaveric lung transplantation but are unlikely to survive the wait for a cadaveric lung allograft or may become ineligible for lung transplantation due to clinical deterioration. Living donor lobar lung transplantation is not indicated for patients with contraindications to lung transplantation and for those whose prognosis is extremely poor despite transplantation, due to the risks to the healthy donor(s).

UpToDate² indicates that 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. The most common diseases that lead to lung transplantation are chronic obstructive pulmonary disease (COPD), idiopathic pulmonary fibrosis (IPF), cystic fibrosis (CF), emphysema due to alpha-1 antitrypsin deficiency, and pulmonary arterial hypertension. More rare diseases that may require lung transplantation include sarcoidosis, Eisenmenger syndrome, and Lymphangioleiomyomatosis (LAM).

Professional Organizations

The American Thoracic Society and the International Society of Heart and Lung Transplantation^{3 8 10} designate that lung transplantation is indicated for patients with chronic, end-stage lung disease who are failing maximal medical therapy, or in those where effective medical therapy does not exist. Potential candidates should be well informed and demonstrate adequate health behavior and a willingness to adhere to guidelines from health care professionals. The primary goal of lung transplantation is to provide a survival benefit particularly in patients with advanced cystic fibrosis, idiopathic pulmonary fibrosis, and primary pulmonary hypertension. Those with emphysema and Eisenmenger’s syndrome may have poorer outcomes.

The American Thoracic Society (ATS)³ international guidelines for the selection of lung transplant candidates indicate that age criteria includes:

- Heart-lung transplantation: approximately 55 years
- Single lung transplantation: approximately 65 years
- Bilateral lung transplantation: approximately 60 years

The American Society of Transplantation¹³ designates that pediatric lung transplantation should be considered in selected children with end stage or progressive lung disease and in life threatening pulmonary vascular disease when medical therapy is not available. For infants the most common indication is congenital heart disease. For children the most common diagnosis is cystic fibrosis. Other diagnoses in children include interstitial lung disease and bronchiolitis obliterans (BO).

The Clinical Practice Committee of the American Society of Transplantation¹⁵ proposed that the presence of AIDS could be considered a contraindication to kidney transplant unless the following criteria were present. These criteria may be extrapolated to other organs¹⁵:

- CD4 count >200 cells/mm-3 for >6 months
- HIV-1 RNA undetectable
- On stable anti-retroviral therapy >3 months
- No other complications from AIDS (e.g., opportunistic infection, including aspergillus, tuberculosis, coccidiosis mycosis, resistant fungal infections, Kaposi’s sarcoma, or other neoplasm)
- Meeting all other criteria for transplantation

CODING INFORMATION

CPT	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
33930	Donor cardiectomy-pneumonectomy (including cold preservation)
33933	Backbench standard preparation of cadaver donor heart/lung allograft prior to transplantation, including dissection of allograft from surrounding soft tissues to prepare aorta, superior vena cava, inferior vena cava, and trachea for implantation
33935	Heart-lung transplant with recipient cardiectomy-pneumonectomy

HCPCS	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
ICD-9	Description
V42.6	Organ tissue replaced by transplant, lung
33.5-33.52	Lung transplant
135	Sarcoidosis
238.1	Lymphangiomyomatosis
273.4	Alpha-1 antitrypsin deficiency
277.00-277.09	Cystic fibrosis
277.89	Eosinophilic granuloma
416.0	Primary pulmonary hypertension
416.8	Other chronic pulmonary heart diseases
416.9	Unspecified chronic pulmonary heart disease
490	Bronchitis, not specified as acute or chronic

491.1	Mucopurulent chronic bronchitis
491.20	Obstructive chronic bronchitis, without exacerbation
491.21	Obstructive chronic bronchitis, with (acute) exacerbation
491.22	With acute bronchitis
491.8	Other chronic bronchitis
491.9	Unspecified chronic bronchitis
492.8	Other emphysema
494.0- 494.1	Bronchiectasis
496	Chronic airway obstruction, not elsewhere classified
501	Asbestosis
506.4	Chronic respiratory conditions due to fumes and vapors
515	Postinflammatory pulmonary fibrosis
516.3	Idiopathic interstitial pneumonia
517.8	Sarcoidosis
518.83	Chronic respiratory failure
518.89	Other diseases of lung, not elsewhere classified
745.4	Eisenmenger's syndrome
748.4	Congenital cystic lung
748.5	Congenital agenesis, hypoplasia, and dysplasia of lung
748.61	Congenital bronchiectasis
770.7	Chronic respiratory disease arising in the perinatal period (Bronchopulmonary dysplasia)
782.5	Cyanosis
786.09	Symptoms involving respiratory system and other chest symptoms, Other
799.02	Hypoxemia
996.84	Complications of transplanted lung

ICD-10	Description
D86.0	Sarcoidosis, lung
J84.81	Lymphangliomyomatosis
E88.01	Alpha-1-antitrypsin deficiency
E84.0	Cystic Fibrosis
J82	Pulmonary eosinophilia
I27.0	Primary pulmonary hypertension
I27.89	Other specified pulmonary heart diseases
I27.89	Other specified pulmonary heart diseases
J44	Other chronic obstructive pulmonary disease
J43- J43.9	Emphysema
J47	Bronchiectasis
J44.9	Chronic obstructive pulmonary disease, unspecified
J61	Pneumoconiosis due to asbestos and other mineral fibers
J68.4	Chronic respiratory conditions due to chemicals, gases, fumes and vapors
J84.10	Pulmonary fibrosis, unspecified
J84.11- J84.112	Idiopathic interstitial pneumonia, idiopathic pulmonary fibrosis
D86.0	Sarcoidosis, lung
J96.10	Chronic respiratory failure, unspecified whether with hypoxia or hypercapnia
I27.89	Eisenmenger's syndrome
Q33.0	Congenital cystic lung
Q33.3- Q33.6	Congenital agenesis, hypoplasia, and dysplasia of lung
Q33.4	Congenital bronchiectasis
P27.81	Bronchopulmonary dysplasia originating in the perinatal period
R23.0	Cyanosis
R06.89	Other abnormalities of breathing
R09.02	Hypoxemia

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