Subject: Lung Transplantation

Policy Number: MCP-115

Revision Date(s): 4/27/2015

Review Date: 12/16/15, 9/15/16, 05/02/17, 05/01/18

DISCLAIMER

This Molina Clinical Policy (MCP) 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 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 Molina Clinical Policy (MCP) document and provide the directive for all Medicare members.

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.

**Procedures:**

*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.

**Split lung bilateral lobar transplantation:** With this procedure, a single left lung from a donor who is approximately 15 percent 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.

Postoperatively, 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

**RECOMMENDATION**

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 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.
Members must meet UNOS guidelines for transplantation and the diagnosis must be made by a Specialist in the Disease and or Transplant Surgeon.

Pre-Transplant Evaluation:

Criteria for transplant evaluation include all of the following:

- History and physical examination
- Psychosocial evaluation and clearance:
  - No behavioral health disorder by history or psychosocial issues:
    - 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
  - Adequate family and social support
- EKG
- Chest x-ray
- Cardiac clearance in the presence of any of the following:
  - chronic smokers
  - > 50 years age
  - those with a clinical or family history of heart disease or diabetes
- Pulmonary clearance if evidence of pulmonary artery hypertension (PAH) or chronic pulmonary disease
- Pulmonary Function Tests
- Lab studies:
  - *Complete blood count, Kidney profile (blood urea nitrogen, creatinine), electrolytes, calcium, phosphorous, albumin, liver function tests, Coagulation profile (prothrombin time, and partial thromboplastin time)
  - *Serologic screening for HIV, Epstein Barr virus (EBV), Hepatitis virus B (HBV), and Hepatitis C(HCV), cytomegalovirus (CMV), RPR and/or FTA:
    - If HIV positive all of the following are met:
      - 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, coccidioides mycosis, resistant fungal infections, Kaposi’s sarcoma, or other neoplasm)
    - If abnormal serology need physician plan to address and/or treatment as indicated
  - UDS (urine drug screen) if patient is current or gives a history of past drug abuse
- *Colonoscopy (if indicated or if patient is 50 ≥ older should have had an initial screening colonoscopy, after initial negative screening requires follow up colonoscopy every ten years) with complete workup and treatment of abnormal results as indicated
- *GYN examination with Pap smear for women ≥ 21 to ≤ 65 years of age or indicated (not indicated in women who have had a TAH or TVH) with in the last three year with complete workup and treatment of abnormal results as indicated
Within the last 12 months:

- Dental examination or oral exam showing good dentition and oral care or no abnormality on panorex or plan for treatment of problems pre or post-transplant
- *Mammogram (if indicated or > age 40) with complete workup and treatment of abnormal results as indicated
- *PSA if history of prostate cancer or previously elevated PSA with complete workup and treatment of abnormal results as indicated

*Participating Centers of Excellence may waive these criteria

Adult and Pediatric Criteria: 5-13 28-36

1. Single, double, or donor lobar lung organ transplantation from a deceased or a living donor is considered medically necessary in adult and pediatric members that have met all of the following criteria: [ALL]

- All pre-transplant criteria are met; and
- Documentation that all medical, pharmaceutical and surgical alternatives to lung transplant have been utilized if applicable that includes but is not limited to the following:
  - Oxygen therapy
  - Pulmonary rehabilitation (Refer to Pulmonary Rehabilitation for Chronic Pulmonary Diseases MCP-086)
  - Lung volume reduction surgery for patients with chronic obstructive lung disease

- Living 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

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

AND

The requesting transplant recipient should not have any of the following absolute contraindications:

- Cardiac, pulmonary, and nervous system disease that cannot be corrected and is a prohibitive risk for surgery
- Malignant neoplasm with a high risk for reoccurrence, non-curable malignancy (excluding localized skin cancer)
- Systemic and/or uncontrolled infection
AIDS (CD4 count < 200 cells/mm³)
- Unwilling or unable to follow post-transplant regimen
  - Documented history of non-compliance
  - Inability to follow through with medication adherence or office follow-up
- Chronic illness with one year or less life expectancy
- Limited, irreversible rehabilitation potential
- Active untreated substance abuse issues, requires documentation supporting free from addiction for minimally 6 months if previous addiction was present
- No adequate social/family support

The requesting transplant recipient should be evaluated carefully and potentially treated if the following relative contraindications are present:
- Irreversible lung disease patients require consultation and clearance by a Pulmonologist prior to consideration of transplantation, this includes the following:
  - Smoking, documentation supporting free from smoking for 6 months
  - Active peptic ulcer disease
  - Active gastroesophageal reflux disease
  - CVA with long term impairment that is not amendable to rehabilitation or a patient with CVA/transient ischemic attack within past 6 months
  - Obesity with body mass index of >30 kg/m² may increase surgical risk
- Chronic liver disease such as Hepatitis B/C/D, or cirrhosis which increases the risk of death from sepsis and hepatic failure requires consultation by a gastroenterologist or hepatologist
- Gall bladder disease requires ultrasound of the gall bladder with treatment prior to transplantation

2. **Disease specific criteria includes all of the following:**

- **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 (PCO₂ ≥50 mmHg);
    - FEV₁ (i.e., forced expiratory volume in the first second) < 25% of predicted, without reversibility;
    - Elevated PaCO₂ > 50 mm Hg with progressive deterioration requiring long term oxygen therapy (defined as ≥ 6 months);
    - Pulmonary hypertension (mean pulmonary artery pressure > 20 mm Hg) and/or cor pulmonale, despite oxygen therapy
*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. 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 ≤ 30% of predicted value;
- increasing frequency of exacerbations requiring cycling antibiotic therapy;
- refractory and/or recurrent pneumothorax;
- oxygen-dependent respiratory failure;
- hypercapnia (defined as PCO2 ≥ 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 (PaO2 < 55 mm Hg);
  - right atrial pressure > 15 mm Hg

**Scleroderma:** Single or double transplantation is indicated and guidelines for transplant include the following:

- FVC below 70% to 80% predicted at the time of diagnosis
Pulmonary Arterial Hypertension (PAH) (includes idiopathic pulmonary hypertension or IPH): Double lung transplantation is preferred and guidelines for transplantation include:
  o No feasible pulmonary thromboendarterectomy for patients with chronic pulmonary thromboembolic disease AND;
  o 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): Single or double transplantation is indicated and guidelines for transplant include both of the following:
  o severe progression of symptoms despite optimal medical management (Refer to UpToDate for treatment options for specific indication);
  o NYHA functional class III or IV*

Lymphangioleiomyomatosis (LAM) and Eosinophilic Granuloma: Single or double transplantation is indicated and guidelines for transplantation include:
  o 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: When retransplantation is being considered ALL of the following factors must be present:
  o the member must be ambulatory
  o ventilator independent
  o free of significant co-morbidities
  o 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:

<table>
<thead>
<tr>
<th>NYHA Functional Classification</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Patients without resulting limitations of physical activity. Ordinary physical activity does not cause undue fatigue, palpitation, dyspnea, or anginal pain.</td>
</tr>
</tbody>
</table>
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**: 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 contraindication as listed above;
  - History and physical within the last 12 months;
  - Kidney profile within the last 12 months;
  - Cardiac update if history of cardiac disease within two years (≥ 50 years of age);
  - 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 contraindication as listed above;
- History and physical within the last 12 months;
- Cardiac update if history of cardiac disease within two years (> 50 years of age);
- 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**

Single, double, or donor lobar lung organ and heart-lung transplantation is considered not medically necessary when the above criteria are not met.

**Summary of Medical Evidence**

The published medical evidence and outcomes for lung and heart-lung transplantation in children and adults in the United States consists of registry data obtained from transplant centers that perform adult and pediatric transplantation and is available from the United Network for Organ Sharing (UNOS) database. Registry data demonstrates graft survival rates and outcomes comparable to other organ transplants. 2-3 14-27

**Coding Information**

The codes listed in this policy are for reference purposes only. Listing of a service or device code in this policy does not imply that the service described by this code is covered or non-covered. Coverage is determined by the benefit document. This list of codes may not be all inclusive.

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<tr>
<th>CPT</th>
<th>Description</th>
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<tbody>
<tr>
<td>32850</td>
<td>Donor pneumonectomy(s) (including cold preservation), from cadaver donor</td>
</tr>
<tr>
<td>32851</td>
<td>Lung transplant, single; without cardiopulmonary bypass</td>
</tr>
<tr>
<td>32852</td>
<td>Lung transplant, single; with cardiopulmonary bypass</td>
</tr>
<tr>
<td>32853</td>
<td>Lung transplant, double (bilateral sequential or en bloc); without cardiopulmonary bypass</td>
</tr>
<tr>
<td>32854</td>
<td>Lung transplant, double (bilateral sequential or en bloc); with cardiopulmonary bypass</td>
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<tr>
<td>32855</td>
<td>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</td>
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<tr>
<td>32856</td>
<td>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</td>
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<tr>
<td>33930</td>
<td>Donor cardiectomy-pneumonectomy (including cold preservation)</td>
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<tr>
<td>33933</td>
<td>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</td>
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<tr>
<td>33935</td>
<td>Heart-lung transplant with recipient cardiectomy-pneumonectomy</td>
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<tr>
<th>HCPCS</th>
<th>Description</th>
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<tr>
<td>S2060</td>
<td>Lobar lung transplantation</td>
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<tr>
<td>S2061</td>
<td>Donor lobectomy (lung) for transplantation, living donor</td>
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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

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<td>33.5-33.52</td>
<td>Lung transplant</td>
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<td>Other chronic pulmonary heart diseases</td>
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<td>Unspecified chronic pulmonary heart disease</td>
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<td>Mucopurulent chronic bronchitis</td>
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<td>Congenital cystic lung</td>
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<td>Congenital agenesis, hypoplasia, and dysplasia of lung</td>
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<td>Congenital bronchiectasis</td>
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<td>Alpha-1-antitrypsin deficiency</td>
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<tr>
<td>J68.4</td>
<td>Chronic respiratory conditions due to chemicals, gases, fumes and vapors</td>
</tr>
<tr>
<td>J84.10</td>
<td>Pulmonary fibrosis, unspecified</td>
</tr>
<tr>
<td>J84.11-J84.112</td>
<td>Idiopathic interstitial pneumonia, idiopathic pulmonary fibrosis</td>
</tr>
<tr>
<td>D86.0</td>
<td>Sarcoidosis, lung</td>
</tr>
<tr>
<td>J96.10</td>
<td>Chronic respiratory failure, unspecified whether with hypoxia or hypercapnia</td>
</tr>
<tr>
<td>I27.89</td>
<td>Eisenmenger’s syndrome</td>
</tr>
<tr>
<td>M34-M34.9</td>
<td>Systemic sclerosis [scleroderma]</td>
</tr>
<tr>
<td>Q33.0</td>
<td>Congenital cystic lung</td>
</tr>
<tr>
<td>Q33.3-Q33.6</td>
<td>Congenital agenesis, hypoplasia, and dysplasia of lung</td>
</tr>
<tr>
<td>Q33.4</td>
<td>Congenital bronchiectasis</td>
</tr>
<tr>
<td>P27.81</td>
<td>Bronchopulmonary dysplasia originating in the perinatal period</td>
</tr>
<tr>
<td>R23.0</td>
<td>Cyanosis</td>
</tr>
<tr>
<td>R06.89</td>
<td>Other abnormalities of breathing</td>
</tr>
<tr>
<td>R09.02</td>
<td>Hypoxemia</td>
</tr>
</tbody>
</table>

**Resources References**

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