Policy:
Lung transplantation is indicated for patients with progressive and irreversible chronic lung disease, who have severe exercise intolerance, are less than 65 years old and are oxygen dependent. Single, double and lobar-lung transplantation have been successful for various pulmonary diseases. Single lung transplantation is most effective for patients with end-stage pulmonary fibrosis, while double lung transplantation is effective for patients with end stage chronic obstructive pulmonary disease and cystic fibrosis patients with preserved cardiac function. Lobar lung transplantation is best suited for children or adolescents who are appropriate candidates for lung transplantation and will not survive waiting for cadaver lungs.

Procedure:
A. Qualifying Conditions for Lung Transplantation
1. Restrictive Lung Diseases (Idiopathic Pulmonary Fibrosis, Eosinophilic Granuloma and Sarcoidosis- -Must meet the disease specific criteria below, Collagen vascular disease, post-chemotherapy, and Scleroderma)
2. Primary Pulmonary Hypertension --Must meet the disease specific criteria below
3. Advanced Cystic Fibrosis—Must meet the disease specific criteria below
4. Obstructive Lung Disease (Bronchiectasis, Bronchiolitis Obliterans, COPD, Emphysema)—For emphysema, COPD, and alpha 1 antitrypsin deficiency must meet disease specific criteria below
5. Bronchopulmonary dysplasia
6. Congenital Heart disease (Eisenmenger’s defect or complex) Must meet the disease specific criteria below
7. Lymphangioleiomyomatosis (LAM)
8. Retransplantation:
   a. Usually due to non-function of the grafted organ, rejection refractory to Immunosuppressive therapy, bronchiolitis obliterans (chronic rejection) and airway complications not correctable by other measures. Outcomes following early emergent retransplantation for primary graft dysfunction are poor, and consequently, its use in this setting is discouraged. Experience with retransplantation for refractory airway complications (dehiscence, strictures) is limited, and results have been conflicting. Retransplantation of carefully selected patients with chronic graft failure due to bronchiolitis obliterans syndrome results in survival rates that approach that of initial transplantation. Such patients should ideally be ventilator-independent and ambulatory at the time of retransplantation and should otherwise meet all standard criteria for suitability, including absence of significant medical comorbidities.

B. Diagnostic Investigations
   1. Full pulmonary function tests
   2. Exercise performance measured by a standardized test, e.g., six-minute walk
   3. Electrocardiogram
   4. Echocardiogram
   5. Right and left heart cath
   6. High resolution computed tomography (HRCT) of the thorax in patients with parenchymal disease, pleural disease, or previous thoracic surgical procedures
   7. Stress echocardiogram—e.g., dobutamine, dobutamine PET, sestamibi, etc.—and/or coronary angiograms in patients at high risk for coronary artery disease
   8. LABS: Liver function studies, CBC, Cr, Chemistry panel, cotinine level and drug screen, HIV, hepatitis B, hepatitis C, toxoplasmosis, CMV, and EBV serologies, 24-hour creatinine clearance Mammogram
   9. Pap Smear
   10. PSA
   11. Colonoscopy
   12. Bone density study
   13. Bronchoscopy

C. Relative Contraindications:
   1. Symptomatic osteoporosis should be assessed with bone densitometry
   2. Severe musculoskeletal disease or neuromuscular disease affecting the thorax, e.g., kyphoscoliosis
   3. Current use of corticosteroids—should try to discontinue these drugs or at least reduce the dose to \( \leq 20 \) mg/d prednisolone or prednisone
   4. Requirement for invasive ventilation Ventilator dependence: The limited data suggest that patients who are dependent on a ventilator prior to the transplant have higher mortality rates but may be candidates for lung transplantation. Singer et al found that ventilator dependence was associated with decreased overall survival; risk of death was highest in the first 6 months post-transplant. A prolonged wait while the patient is on a mechanical ventilator may lead to various complications such as infections,
cardiovascular deconditioning, and muscle atrophy, all of which further compromise the outcome of the transplant.

5. Colonization with fungi or atypical mycobacteria
6. Adequately treated M. tuberculosis

D. Current Contraindications

1. Nutritional issues--Patients with a BMI > 30 kg/m² require weight loss to become eligible for transplant. Patients who have cachexia (BMI < 16) likely have poor nutritional status and would have a poor outcome following transplantation. Obesity (BMI >30) also may be a concern because of postoperative atelectasis and pneumonia.

2. Social and Psychiatric Issues - Refer for psychosocial evaluation and/or psychiatry consultation for guidance. Psychosocial problems that are unable to be resolved and that have a high likelihood of impacting negatively on the patient's outcome, e.g., poorly controlled major psychoaffective disorder, inability to comply with complex medication regimen, are a relative contraindication. A documented history of noncompliance with medical care or treatment plans even in the absence of documented psychiatric problem is a relative contraindication as per Member Compliance Policy I.7.
   a. Active alcoholism and substance abuse. Requires 6 months of documented abstinence through participation in a structured alcohol/substance abuse program with regular meeting attendance and negative random drug testing as per Member Compliance Policy I.7.
   b. Emotional instability, significant depression or other psychiatric illness that cannot be controlled that would impact ability to comply with a complex evaluation process, surgical procedure and post-transplant plan of care and/or ability to give informed consent (and does not have a representative/guardian/conservator).
   c. Limited cognitive ability (memory loss, dementia, etc.) that would impact ability to comply with a complex evaluation process, surgical procedure and post-transplant plan of care and/or ability to give informed consent (and does not have a representative/guardian/conservator).
   d. Lack of psychosocial support as indicated by either no identified caregiver or an uncommitted caregiver. This would include the lack of transportation to and from transplant related appointments, patient and/or caregiver is unable to adhere to the requirements of transplant related treatment plan. A care contract may be needed.
   e. History of non-adherence that has not been successfully remediated
   f. Inability to give informed consent. If the patient has an authorized representative/guardian/conservator or parent in the case of a minor, that individual must understand and support the ongoing health care needs of the patient.

3. Severe end stage organ damage other than the lung including irreversible severe renal dysfunction—creatinine clearance of < 50 mg/ml/min, severe diabetes with end organ damage, irreversible severe hepatic disease (total bilirubin level of greater than 2 mg/dL, is associated with an unfavorable outcome following transplant). A patient with severe left ventricular systolic or diastolic dysfunction is not a candidate for lung only transplantation (but may be a candidate for a Heart/Lung transplant). No angiographic evidence of significant coronary artery disease, ejection fraction greater than 40 %, no myocardial infarction in last 6 months, negative stress test). Persons with any cardiac symptoms may require heart catheterization to rule out significant heart disease;
4. Infections
   a. Acquired Immunodeficiency Syndrome. A member must have CD4 counts greater than 200 cells/mm³ for greater than 6 months, HIV 1 RNA viral load undetectable, no other complications from AIDS such as opportunistic infection, and on stable antiviral therapy greater than 3 months.
   b. Systemic or uncontrolled infection including sepsis.

5. Active *malignancy* within the past two years with the exception of basal cell and squamous cell carcinoma of skin. In addition a waiting period of at least 5 years is needed for extracapsular renal cell tumors, breast cancer stage 2 or higher, colon cancer staged higher than Dukes A, and melanoma, level III or higher. Requires oncology clearance.

6. Hepatitis B antigen positivity.

7. Hepatitis C with biopsy-proven histologic evidence of liver disease.


9. Age limits:
   a. Heart–lung transplants ~ 55 years
   b. Single lung transplants ~ 65 years
   c. Bilateral lung transplants ~ 60 years

10. Irreversible, severe brain damage

11. Post-transplant lymphoproliferative disease (PTLD) unless no active disease demonstrated by negative positron emission tomography (PET) scan and resolved adenopathy on computed tomography (CT) and/or magnetic resonance imaging.

12. Limited irreversible rehabilitative potential. Under established guidelines, active rehabilitation is considered important to the success of transplantation. Mechanically-ventilated or otherwise immobile persons are considered poor candidates for transplantation; however, short-term mechanical ventilation (less than 2 weeks) or bridge to transplant with ambulatory ECMO does not, in itself, rule out candidacy for lung transplantation.

**Additional Selection Criteria:** Transplant will not be approved if any one of the following indicators of non-compliance are observed or documented:

1. For members with a history of tobacco use, if the member fails or refuses to submit to monthly cotinine testing for the preceding 6 months prior to the transplant and while listed, or refuses to actively and continuously participate in an accepted smoking cessation program.

2. For members with a history of alcohol abuse, member fails or refuses to submit to testing for alcohol use. Absence of documentation showing member has not engaged in alcohol use for at least six months prior to transplant and monthly while listed.

3. For members with a history of illicit drug use, member fails or refuses to submit to testing for illegal drug use. Absence of documentation showing member has not engaged in illegal drug use for at least six months prior to transplant and monthly testing while listed.

4. Tobacco, Alcohol and Drug Addiction: Refusal or failure to participate in available addiction interventions for actively using members must be documented in non-compliance determinations.
5. Documentation of non-impactable social issues that substantially increase the risks of an adverse outcome of the medical therapy or transplant at issue

6. Severely Mentally Ill Adults and Severely Emotionally Disturbed Minors: Non-adherence to psychotropic medications or medical regimen in SMI or SED members for whom core symptoms include lack of insight into illness, must be assessed for adequacy of and engagement with psychosocial resource supports in Care Coordination prior to non-compliance determinations.

7. Developmental or Acquired Cognitive Impairment and Dementia: Psychosocial and guardianship support as well as reversibility of impairment must be assessed and documented prior to non-compliance determinations.

E. Disease Specific Guidelines

1. Nonbronchiectatic Chronic Obstructive Lung Disease (emphysema, chronic bronchitis, alpha 1 antitrypsin deficiency and bronchiolitis obliterans). COPD patients are considered if they have a BODE index* of 7 to 10 (on a scale of 0 to 10) or at least one of the following criteria:
   a. FEV$_1$ < 20% of predicted and either DLCO of less than 20% or homogeneous distribution of emphysema
   b. Hospitalizations for COPD exacerbation associated with hypercapnia defined as Pa$_{CO2}$ > 50 mm Hg and/or the following:
      - Declining body mass
      - Increasing oxygen requirements
      - Reduced serum albumin
   c. Presence of Cor Pulmonale (enlarged pulmonary arteries on chest Xray, mean pulmonary artery pressure greater than 25 mm HG at rest, pedal edema or JVD, or RVH or Right atrial enlargement on EKG) or pulmonary hypertension despite oxygen therapy

*(BODE takes into account the following factors: BMI, degree of airflow assessed by the percent predicted FEV$_1$, degree of dyspnea assessed by the MMRC dyspnea scale, and the exercise capacity as assessed with the 6 MWT)

2. Cystic Fibrosis and other Bronchiectatic Diseases: FEV1 <30% of predicted, or rapidly declining lung function if FEV1 >30% (females and patients <18 years of age have a poorer prognosis; consider earlier listing) and any 2 of the following:
   a. Oxygen dependent respiratory failure
   b. Pulmonary hypertension
   c. Hypercapnia Pa$_{CO2}$ > 55 mm Hg

3. Idiopathic Pulmonary Fibrosis (also known as UIP usual interstitial pneumonia). Member must have histologic or radiographic evidence of UIP and any of the following:
   a. A decrease in pulse oximetry below 88% during a six minute walk test (6-MWT)
   b. A 10% or greater decrement in FVC during 6 months follow up
   c. Diffusing capacity of the lung for carbon monoxide (DLCO) <39% of predicted value
   d. Honeycombing on HRCT (fibrosis score of >2)

4. Member with histologic evidence of NSIP (nonspecific interstitial pneumonia) and any of the following:
   a. A DLCO of less than 35% predicted
   b. A 10% or greater decrease in FVC or 15% decrease in DLCO during 6 months of follow up.

5. Systemic Disease with Pulmonary Fibrosis
a. Pulmonary fibrosis is a common lung pathology in a number of systemic diseases, e.g., scleroderma, rheumatoid arthritis. Evidence that the systemic disease is not active is required. Should be considered on a case by case basis.

6. Sarcoidosis --Member must have NYHA class III or IV and meet one of the following:
   a. Hypoxemia at rest
   b. Presence of cor pulmonale or pulmonary hypertension
   c. Elevated right atrial pressure exceeding 15 mm Hg

7. Pulmonary Hypertension without Congenital Heart Disease (Idiopathic pulmonary arterial hypertension)
   a. Symptomatic, progressive disease which, despite optimal medical and/or surgical treatment, leaves the patient in NYHA III or NYHA IV.
   b. Cardiac index of less than 2 L/min/m²
   c. Right atrial pressure of more than 15 mm Hg
   d. Low (< 350 meter or <1,148 feet) or declining 6 MWT
   e. Failing therapy with iv epoprostenol or equivalent

8. Pulmonary Hypertension Secondary to Congenital Heart Disease (Eisenmenger's Syndrome) Member must meet one of the following:
   a. Severe, progressive symptoms with function at NYHA III or NYHA IV level despite optimal medical management
   b. Pulmonary hypertension with mean pulmonary artery pressure greater than 25 mm Hg
   c. Signs of right ventricular failure—hepatomegaly, ascites

9. Lymphangioleiomyomatosis (LAM)
   a. Severe impairment in lung function and exercise capacity (VO₂ < 50% predicted)
   b. Hypoxemia at rest

10. Pulmonary Langerhans Cell Histiocytosis (Eosinophilic Granuloma)
    a. Severe impairment in lung function and exercise capacity (VO₂ < 50% predicted)
    b. Hypoxemia at rest

**Pediatric Lung Transplantation**

Cardiopulmonary Vascular Disease (Qualifying Conditions):
1. Primary pulmonary hypertension,
2. Pulmonary hypertension associated with structural heart disease
3. Pulmonary vein stenosis
4. Pulmonary hypertension associated with parenchymal lung disease
5. Congenital abnormalities of lung development or of lung adaptation to extrauterine life.

**Criteria:**
1. Disease no longer responding to maximum medical and surgical treatment
2. Moderately severe or severe functional impairment (NYHA Class III or IV)
3. Right ventricular failure, severe cyanosis, and low cardiac output

**When to Refer for Lung Transplantation Evaluation**
"In general, referral for transplantation assessment is advisable when patients have a less than 50%, 2 to 3 year predicted survival or New York Heart Association (NYHA) class III or IV level of
The Lung Allocation Score (LAS) is used to place patients on the lung waiting list. This is similar to the MELD system for liver transplantation. The LAS takes into account the severity of the illness pre-transplant including the likelihood of death on the waiting list and the likelihood of survival one year post-transplant. The LAS is a dynamic measurement that is updated on a regular basis according to a follow-up schedule determined by UNOS. Raw LAS = net transplant benefit (1-year survival with transplant -- 1-year survival without transplant) minus medical urgency (1-year survival without transplant). This score is then normalized to a 0 to 100 scale for ease of use. Information about the LAS and the LAS Calculator can be found at: http://optn.transplant.hrsa.gov/resources/allocationcalculators.asp?index=88

Double lung transplantation is indicated for cystic fibrosis and other lung diseases characterized or complicated by chronic infections.

COPD and alpha 1-antitrypsin deficiency emphysema
- BODE index exceeding 5 (on a scale of 0 to 10) (BODE takes into account the following factors: BMI, degree of airflow assessed by the percent predicted FEV₁, degree of dyspnea assessed by the MMRC dyspnea scale, and the exercise capacity as assessed with the 6 MWT)Cystic Fibrosis and other Bronchiectasis
- Postbronchodilator FEV₁ less than 30%
- Exacerbation of pulmonary disease requiring ICU stay
- Increasing frequency of exacerbations requiring antibiotic therapy
- Refractory and/or recurrent pneumothorax
- Recurrent hemoptysis not controlled by embolization

Idiopathic Pulmonary Fibrosis (UIP usual interstitial pneumonia) and Non-Specific Interstitial Pneumonia (NSIP)
- Histologic or radiographic evidence of UIP irrespective of vital capacity

Histologic evidence of fibrotic NSIP
- Pulmonary Hypertension without congenital heart disease
  - NYHA class III or IV irrespective of ongoing therapy
  - Rapidly progressive disease

Sarcoidosis
- NYHA class III or IV

Lymphangioleiomyomatosis
- NYHA class III or IV

Pulmonary Langerhans Cell Histiocytosis (Eosinophilic Granuloma)
- NYHA class III or IV

Special Instructions: N/A

Line of Business Applicability:
For Medicaid/Medicaid Expansion Plan members, this policy will apply. Coverage is based on medical necessity criteria being met and the codes being submitted and considered for review being included on either the Michigan Medicaid Fee Schedule (located at: http://www.michigan.gov/mdch/0,1607,7-132-2945_42542_42543_42546_42551-159815--,00.html), the Illinois Medicaid Fee Schedule (located
For **Medicare** members, coverage is determined by the Centers for Medicare and Medicaid Services (CMS). If a coverage determination has not been adopted by CMS, this policy applies. Medicare Fee Schedules can be found on the CMS website ([https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/FeeScheduleGenInfo/index.html](https://www.cms.gov/Medicare/Medicare-Fee-for-Service-Payment/FeeScheduleGenInfo/index.html)).


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