Trillium Gift of Life Network

Ontario's Adult Referral Criteria for Lung Transplantation

Version 4.0

Ontario Health
Trillium Gift of Life Network

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PATIENT REFERRAL CRITERIA

The patient referral criteria are guidelines which a Health Care Provider would utilize to refer a patient to a transplant center for assessment. In general, referral for lung transplantation assessment is advisable for adults with chronic, end-stage lung disease who have high (>50%) risk of death from lung disease within 2 years if lung transplantation is not performed, AND have high (>80%) likelihood of 5-year post-transplant survival from a general medical perspective provided that there is adequate graft function.

The criteria identified below are the agreed upon conditions for which a patient should be referred for lung transplant assessment.

1) **Chronic Obstructive Pulmonary Disease (COPD):** Referral for lung transplantation should be considered for patients with chronic obstructive pulmonary disease (COPD). Such patients would meet the following criteria:
    - Clinical deterioration despite maximal treatment including medication, pulmonary rehabilitation, and oxygen therapy, and as appropriate, nocturnal non-invasive positive pressure ventilation;
    - For a patient who is a candidate for bronchoscopic or surgical lung volume reduction (LVR), simultaneous referral of patients with COPD for both lung transplant and LVRS evaluation is appropriate;
    - Body-Mass Index, Airflow Obstruction, Dyspnea, and Exercise (BODE) index of 5-6 with additional factor(s) present suggestive of increased risk of mortality:
        - Frequent acute exacerbations
        - Increase in BODE score >1 over past 24 months
        - Pulmonary artery to aorta diameter >1 on CT scan
        - Forced Expiratory Volume (FEV1) 20-25% predicted
    - Poor quality of life unacceptable to the patient

2) **Cystic Fibrosis and Other Causes of Bronchiectasis:** Referral for lung transplantation should occur for an individual with cystic fibrosis meeting any of the following criteria, despite optimal medical management including a trial of elexacaftor/ tezacaftor/ ivacaftor if eligible:
    - FEV1 below 30% predicted
    - FEV1 <40% predicted in adults and any of the following:
        - 6-minute walk distance <400 m;
        - P_aCO_2 > 50 mmHg
        - Hypoxemia at rest or with exertion
        - Pulmonary hypertension (systolic pulmonary arterial pressure (PAP) >50 mmHg on echocardiography or evidence of right ventricular dysfunction)
        - 2 exacerbations per year requiring intravenous antibiotics
        - Worsening nutritional status despite supplementation
        - Pneumothorax
        - Massive hemoptysis (>240mL) despite bronchial artery embolization

- FEV_1 < 50% predicted and rapidly declining based on pulmonary function testing or progressive symptoms
- Any exacerbation requiring positive pressure ventilation

3) **Interstitial Lung Disease:** Referral for lung transplantation should be made at time of diagnosis, even if a patient is being initiated on therapy, for histopathological Usual Interstitial Pneumonia (UIP) or radiographic evidence of a probable or definite UIP pattern.
   - Any form of pulmonary fibrosis with forced Vital Capacity (FVC) <80% predicted or diffusion capacity of the lung for carbon monoxide (DLCO) <40% predicted;
   - Any form of pulmonary fibrosis with one of the following in the past 2 years:
     - Relative decline in FVC ≥10%
     - Relative decline in DLCO ≥15%
     - Relative decline in FVC ≥ 5% in combination with worsening of respiratory symptoms or radiographic progression
   - Supplemental oxygen requirement, either at rest or on exertion;
   - For inflammatory interstitial lung disease (ILD), progression of disease (either on imaging or pulmonary function) despite treatment;
   - For patients with connective tissue disease or familial pulmonary fibrosis, early referral is recommended as extrapulmonary manifestations may require special consideration.

4) **Pulmonary Arterial Hypertension:** Referral for lung transplantation should be considered for patients with pulmonary arterial hypertension (PAH). Such patients would have one of the following:
   - ESC/ERS intermediate or high risk or REVEAL risk score ≥ 8 despite appropriate PAH therapy
   - Significant RV dysfunction despite appropriate the use of PAH therapy
   - Need for IV or SC prostacyclin therapy
   - Progressive disease despite appropriate therapy or recent hospitalization for worsening of PAH;
   - Known or suspected high-risk variants such as pulmonary veno-occlusive disease (PVOD) or pulmonary capillary hemangiomatosis (PCH), scleroderma, large and progressive pulmonary artery aneurysms.
   - Signs of secondary liver or kidney dysfunction due to PAH
   - Potentially life-threatening complications such as recurrent hemoptysis

5) **Sarcoidosis:** Referral for lung transplantation should be considered for patients with sarcoidosis if they are NYHA functional class III or IV.

6) **Lymphangioleio-myomatosis (LAM):** Referral for lung transplantation should occur for patients with lymphangioleio-myomatosis who has any of the following criteria, despite mTOR inhibitor therapy:
   - Severely abnormal lung function (e.g. FEV1 < 30% predicted)
   - Exertional dyspnea (NYHA class III or IV)
   - Hypoxemia at rest
   - Pulmonary hypertension
   - Refractory pneumothorax

7) **Pulmonary Langerhans Cell Histiocytosis (Eosinophilic Granuloma):** Referral for lung transplantation should be considered for patients with pulmonary Langerhans cell histiocytosis if they are NYHA functional class III or IV.

8) **Acute Respiratory Distress Syndrome (ARDS):** Persistent requirement for mechanical ventilatory support and /or ECLS without expectation of clinical recovery and with evidence of irreversible lung destruction. Such patient would be required to meet ALL of the following criteria:
   - Meet other criteria for lung transplantation
   - At least 6 weeks on mechanical ventilation or ECLS
   - The potential to undergo rehabilitation after transplantation
   - Be able to provide first person consent for transplantation
   - Negative COVID-PCR test
   - Clinical and Radiological evidence of irreversible lung disease
   - Age less than 65

9) **Thoracic Malignancy:** Lung transplant should be limited to very select cases of lung-limited adenocarcinoma in situ, minimally invasive adenocarcinoma, or lepidic predominant adenocarcinoma for patients with the following conditions. Such transplant should only be performed in selected centres:
   - Surgical resection is not feasible either because of multifocal disease or significant underlying pulmonary disease;
   - Multifocal disease has resulted in significant lung restriction and respiratory compromise;
   - Medical oncology therapies have failed or are contraindicated; and
   - Lung transplant is expected to be curative.

10) **Immunization Status:** Vaccination history to be reviewed with the patient and vaccines to be updated pre-transplant where possible.
   - **Note:* Patient referral, listing or transplantation processes will not be impeded due to incomplete vaccinations. *Please refer to **ON Adult Lung Listing Criteria** document to see COVID-19 vaccination specific information.*

Version Control

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