Pharmacy Prior Authorization
Pulmonary Hypertension Agents Clinical Guidelines

Adcirca®
Adempas®
epoprostenol
Letairis®
Remodulin®
Opsumit®
Orenitram®
Revatio®
Tracleer®
Tyvaso®
Uptravi®
Veletri®
Ventavis®

Authorization Guidelines:
• General Coverage Criteria for ALL Medications:
  o Prescribed by, or in consultation with a pulmonologist or cardiologist with experience in treating pulmonary hypertension
  o Diagnosis of pulmonary hypertension with a mean pulmonary artery pressure (MPAP) \( \geq 25 \text{mmHg} \) at rest confirmed by right-heart catheterization (RHC)
    ▪  Adcirca, Letairis, Tracleer, Opsumit, Revatio (sildenafil), Adempas: World Health Organization (WHO) Class II to IV symptoms*
    ▪  Remodulin, epoprostenol, Tyvaso, Veletri, Ventavis, Orenitram: WHO Class III to IV symptoms*
  o Patient does not have fluid retention OR is receiving a diuretic

• Additional Coverage Criteria Based on Pulmonary Hypertension Type:
  o Type I Pulmonary ARTERIAL Hypertension (PAH):
    ▪  Patient is receiving anticoagulation; AND
    ▪  Patient remains symptomatic despite optimal treatment with a calcium channel blocker
    ▪  Patient had a negative vasoreactivity test
    ▪  Patient has PAH associated with connective tissue disease, congenital heart disease, HIV, portal hypertension, or schistosomiasis (this type is rarely vasoreactive)
  o Type II Pulmonary Hypertension (due to left heart disease): treatment with pulmonary hypertension medications is NOT recommended
  o Type III Pulmonary Hypertension (due to lung disease and/or hypoxia)
    ▪  Patient remains WHO Class III to IV despite optimal treatment of underlying cause (i.e., COPD, interstitial lung disease, sleep-disordered breathing)
    ▪  Patient is receiving supplemental oxygen
  o Type IV Pulmonary Hypertension (Chronic thromboembolic pulmonary hypertension, CTEPH):
    ▪  Patient is not a surgical candidate OR has persistent disease following thromboendarterectomy
    ▪  Patient is receiving anticoagulation
  o Type V Pulmonary Hypertension (due to unclear multifactorial mechanisms): benefit of pulmonary hypertension agents is unknown

• Additional Coverage Criteria Based on Medication Requested:
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- **Adcirca**: Member has a documented contraindication or intolerance or therapeutic failure of an adequate one-month trial of sildenafil. Patient is **NOT** currently taking a guanylate cyclase stimulator (i.e., Adempas)
- **Adempas, Letairis, Opsumit, and Tracleer**: Member is **NOT** pregnant
- **Adcirca, Revatio(sildenafil), Adempas**: Member is **NOT** concurrently on organic nitrates (i.e., isosorbide mononitrate, isosorbide dinitrate, nitroglycerin)
- **Adempas**: Member is **NOT** concurrently using PDE inhibitors (i.e., sildenafil, Adcirca, dipyridamole, or theophylline)
- **Orenitram**:
  - Member does not have severe hepatic impairment (Child Pugh Class C)
  - Member is not being converted from an inhaled or infused vasodilator (epoprostenol, treprostinil, or iloprost)
- **Adcirca, Revatio, Letairis, Opsumit, Flolan, Veletri, and Tracleer**: Member does not have Pulmonary veno-occlusive disease (PVOD)
- **Letairis**: Member does not have idiopathic pulmonary fibrosis
- **Tracleer**: Member is not taking glyburide or cyclosporine
- **Opsumit**: Member is not taking strong CYP3A4 inducers/inhibitors
- **Veletri**: Member does NOT have HF with severe left ventricular dysfunction
- **Flolan**: Member does NOT have HF with left ventricular dysfunction

**Initial Approval**: One year

**Renewal**: 3 years if responding to medication

**Additional Information**:

World Health Organization (WHO) functional classification of pulmonary artery hypertension:

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Persons with no symptoms(^1), and for whom ordinary physical activity does not cause fatigue, palpitation, dyspnea, or angina pain</td>
</tr>
<tr>
<td>II</td>
<td>Persons who are comfortable at rest but who have symptoms(^1) with ordinary physical activity</td>
</tr>
<tr>
<td>III</td>
<td>Persons who are comfortable at rest but have symptoms(^1) with less-than-ordinary effort</td>
</tr>
<tr>
<td>IV</td>
<td>Persons who have symptoms(^1) at rest</td>
</tr>
</tbody>
</table>

\(^1\) Key symptoms of PAH include fatigue, dizziness, and fainting (near syncope)

**Dosing Recommendations per Manufacturer**:

<table>
<thead>
<tr>
<th>Medications</th>
<th>Max Daily Dose</th>
<th>Dosage Strength</th>
<th>Quantity Limits</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adcirca</td>
<td>40 mg</td>
<td>20 mg</td>
<td>60 tablets per 30 days</td>
</tr>
<tr>
<td>Adempas</td>
<td>7.5 mg</td>
<td>0.5 mg, 1 mg, 1.5 mg, 2 mg, 2.5 mg</td>
<td>90 tablets per 30 days</td>
</tr>
<tr>
<td>Opsumit</td>
<td>10 mg</td>
<td>10 mg</td>
<td>30 tablets per 30 days</td>
</tr>
</tbody>
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<tbody>
<tr>
<td>Revatio</td>
<td>60 mg</td>
<td>10 mg/ml</td>
<td>180 ml per 30 days</td>
</tr>
<tr>
<td>suspension</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Revatio tabs</td>
<td>60 mg</td>
<td>20 mg</td>
<td>90 tablets per 30 days</td>
</tr>
</tbody>
</table>

References: