PHARMACY PRIOR AUTHORIZATION
Clinical Guideline – Pulmonary Arterial Hypertension Clinical Guideline

Adcirca (tadalafil)  Opsumit (macitentan)  Uptravi (selexipag)
Revatio (sildenafil)  Tracleer (bosentan)  Veletri (epoprostenol)
Adempas (riociguat)  Remodulin (treprostinil)  Ventavis (Iloprost)
epoprostenol  Tyvaso (treprostinil)
Letairis (ambrisentan)  Orenitram (treprostinil)

Preferred Agents: sildenafil, Adcirca, Tracleer, Letairis, and epoprostenol

Authorization Guidelines:

1. General Coverage Criteria for ALL Medications:
   - Prescribed by, or in consultation with a pulmonologist or cardiologist
   - Evidence of right heart catheterization (RHC) with a mean PAP ≥ 25 mm Hg
   - Medical records supporting diagnosis of Pulmonary Arterial Hypertension (PAH) WHO Group I with NYHA Functional Class II to IV symptoms.
   - Inadequate response, or intolerance/contraindication to, a calcium channel blocker (CCB)

   Note: Adempas may include WHO Group IV and does not require a trial of CCB

2. Additional Coverage Criteria Based on Medication Requested:
   - **Brand Revatio** (sildenafil) oral suspension
     - Documentation to support the difficulty or inability to swallow and the necessity of the brand suspension formulation.
   - **Adcirca** (tadalafil)
     - Documentation to support trial and failure or has contraindication/intolerance to or intolerance to sildenafil
   - **Opsumit** (macitentan)
     - Member has tried and failed or has contraindication/intolerance to 2 preferred oral agents
       - One PDE-5 inhibitor (e.g., sildenafil or Adcirca)
       - One Endothelin Receptor Antagonist (e.g., Tracleer or Letairis)
   - **Adempas** (riociguat)
     - Diagnosis of WHO (PAH) Group I (as described above) and
     - Member has tried and failed or has contraindication/intolerance to 2 preferred oral agents
       - One PDE-5 inhibitor (e.g., sildenafil or Adcirca)
       - One Endothelin Receptor Antagonist (e.g., Tracleer or Letairis)
   - **OR**
     - Diagnosis of CTEPH, WHO Group IV and one of the following:
       - Recurrent or persistent CTEPH, after surgical treatment
• Inoperable CTEPH
  o Uptravi (selexipag), Orenitram (treprostinil)
    ▪ Member has tried and failed or has contraindication/intolerance to 2 preferred oral agents
      • One PDE-5 Inhibitor (e.g., sildenafil or Adcirca)
      • One Endothelin Receptor Antagonist (e.g., Tracleer or Letairis)
  o Tyvaso (trepostinil), Ventavis (iloprost), Remodulin (trepostinil)
    ▪ Member has tried and failed or has contraindication/intolerance 2 preferred oral agents
      • One PDE-5 inhibitor (e.g., sildenafil or Adcirca)
      • One Endothelin Receptor Antagonist (e.g., Tracleer or Letairis)

Non Coverage Criteria:
• Use not approved by the FDA; AND
• The use is unapproved and not supported by the literature or evidence as an accepted off-label use.
• Any contraindications to treatment including but not limited to the following:
  o Pregnancy: Endothelin Receptor Antagonist (ERA’s) and Adempas
  o Concurrent use of organic nitrates (i.e., isosorbide mononitrate, isosorbide dinitrate, nitroglycerin): PDE-5 inhibitors and Adempas
  o Child Pugh class C hepatic impairment: Orenitram
  o HF with severe left ventricular dysfunction: Veletri/epoprostenol
  o Pulmonary veno-occlusive disease (PVOD): Adcirca, sildenafil, Letairis, Opsumit, epoprostenol, and Tracleer

Initial Approval: 6 months

Renewal:
• Medical records and lab results to support response to therapy; to maintain or achieve a low risk profile (e.g., improvement in 6 min walk distance, functional class, or reducing time to clinical worsening)

• Approve for 1 year

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(^*), 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(^*) with ordinary physical activity</td>
</tr>
<tr>
<td>III</td>
<td>Persons who are comfortable at rest but have symptoms(^*) with less-than-ordinary effort</td>
</tr>
<tr>
<td>IV</td>
<td>Persons who have symptoms(^*) at rest</td>
</tr>
</tbody>
</table>
AETNA BETTER HEALTH OF PENNSYLVANIA
Clinical Practice Guideline

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

Dosing Recommendations per Manufacturer:

<table>
<thead>
<tr>
<th>Medications</th>
<th>Dosage Strength</th>
<th>Maximum Quantity Limits</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adcirca</td>
<td>20 mg</td>
<td>60 tablets per 30 days</td>
</tr>
<tr>
<td>Adempas</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>30 tablets per 30 days</td>
</tr>
<tr>
<td>Orenitram</td>
<td>0.125 mg, 0.25 mg, 1 mg, 2.5 mg</td>
<td>Determine by tolerability</td>
</tr>
<tr>
<td>Sildenafil tabs</td>
<td>20 mg</td>
<td>90 tablets per 30 days</td>
</tr>
<tr>
<td>Tracleer</td>
<td>62.5mg, 125mg</td>
<td>60 tablets per 30 days</td>
</tr>
<tr>
<td>Letairis</td>
<td>5mg, 10mg</td>
<td>30 tablets per 30 days</td>
</tr>
<tr>
<td>Uptravi</td>
<td>200 mcg, 400 mcg, 600 mcg, 800 mcg, 1000 mcg, 1200 mcg, 1400 mcg, 1600 mcg</td>
<td>60 tablets per 30 days following titration quantities</td>
</tr>
<tr>
<td>Tyvaso</td>
<td>18 mcg (3 breaths) per sessions</td>
<td>54 mcg (9 breaths) per treatment session, 4 times daily</td>
</tr>
</tbody>
</table>

References: