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: Adcirca, epoprostenol, Letairis, Opsumit, sildenafil and Tracleer

Authorization Guidelines:

A. Authorization Criteria for All agents:
   1. Prescribed by, or in consultation with a pulmonologist or cardiologist
   2. Evidence of right heart catheterization (RHC) with a mean PAP equal to or greater than 25 mm Hg
   4. 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

B. Additional Drug Specific Criteria:
   1. Brand Revatio (sildenafil) oral suspension: Documentation to support the difficulty or inability to swallow
   2. Adcirca (tadalafil): Trial and failure or contraindication/intolerance to sildenafil
   3. Adempas (riociguat)
      a. Diagnosis of WHO (PAH) Group I (as described above)
         AND
      b. Trial and failure or contraindication/intolerance to two (2) preferred oral agents

Last Review: 8/2018
Previous PARP Approval 1/2018
Current PARP Approval: 1/2019
i. One Phosphodiesterase Type 5 Inhibitor (PDE-5) inhibitor (e.g., sildenafil or Adcirca)

ii. One Endothelin Receptor Antagonist (e.g., Tracleer, Letairis or Opsumit)

OR

c. Diagnosis of Chronic Thromboembolic Pulmonary Hypertension (CTEPH) WHO Group IV and one of the following:
   i. Recurrent or persistent CTEPH, after surgical treatment
   ii. Inoperable CTEPH

4. **Uptravi** (selexipag), **Orenitram** (treprostinil)
   a. Trial and failure or contraindication/intolerance to two (2) preferred oral agents
      i. One PDE-5 Inhibitor (e.g., sildenafil or Adcirca)
      ii. One Endothelin Receptor Antagonist (e.g., Tracleer, Letairis or Opsumit)

5. **Tyvaso** (trepostinil), **Ventavis** (Iloprost), **Remodulin** (trepostinil)
   a. Member must have NYHA Functional Class III-IV (class II-IV for trepostinil)
   b. Trial and failure or contraindication/intolerance two (2) preferred oral agents
      i. One PDE-5 inhibitor (e.g., sildenafil or Adcirca)
      ii. One Endothelin Receptor Antagonist (e.g., Tracleer, Letairis or Opsumit)

**Coverage Limitation:**

Any contraindications to treatment including but not limited to the following:

1. Pregnancy: Endothelin Receptor Antagonists (ERAs) and Adempas
2. Concurrent use of organic nitrates (for example, isosorbide mononitrate, isosorbide dinitrate, nitroglycerin): Phosphodiesterase Type 5 Inhibitors (PDE-5) and Adempas
3. Child Pugh class C hepatic impairment: Orenitram
4. Heart Failure (HF) with severe left ventricular dysfunction: Veletri/epoprostenol
5. 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

Quantity Limit

- Adcirca: 60 tabs per 30 days
- Adempas: 90 tabs per 30 days
- Opsumit: 30 tabs per 30 days
- Orenitram: Determine by tolerability: 90 tabs per 30 days
- Sildenafil tabs: 90 tabs per 30 days
- Brand Revatio (sildenafil) oral suspension: 180 ml per 30 days
- Tracleer: 60 tabs per 30 days
- Letairis: 30 tabs per 30 days
- Uptravi: 60 tabs per 30 days: (may be higher during titration phase)
- Tyvaso: 54 mcg (9 breaths) per treatment session, 4 times daily

Additional Information:

Pulmonary Arterial Hypertension (PAH) is a rare and complex disease with the risk of high morbidity and mortality. Diagnosis of Pulmonary Arterial Hypertension (PAH) is primarily based on right heart catheterization (RHC) with mean Pulmonary Arterial Pressure (PAP) greater than or equal to 25 mmHg, Pulmonary Artery Wedge Pressure (PAWP) less than or equal to 15mmHg and Pulmonary Vascular Resistance (PVR) greater than 3 wood units. Additional treatment options have recently increased within this disease and consists of three key drug classes which includes the Phosphodiesterase Type 5 Inhibitor (PDE-5) inhibitors (for example, sildenafil or tadalafil), endothelin receptor antagonists (ERAs) (for example, Tracleer, Letairis, and Opsumit), and Prostacyclin analogues (for example, treprostonil, epoprostenol, and iloprost). Treatment is considered in a stepwise approach often beginning with monotherapy followed by combination treatment such as with an endothelin receptor antagonist (ERA) and Phosphodiesterase Type 5
Inhibitor (PDE5) Inhibitor. However, severity of treatment such as rapid disease progression or worsening clinical prognosis may require initiation of treatment with a prostanoid before a Phosphodiesterase Type 5 (PDE-5) Inhibitor or endothelin receptor antagonist (ERA). Current national guidelines recommend prior to initiation of treatment patients should be referred to Expert Treatment Centers for Pulmonary Arterial Hypertension (PAH).

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

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

References:
