Missouri Pharmacy Program – Preferred Drug List

Pulmonary Arterial Hypertension (PAH) Agents (Oral)

Effective 01/10/2013
Revised 01/10/2019

Preferred Agents

- Letairis®
- Sildenafil (gen Revatio)
- Tracleer®

Non-Preferred Agents

- Adcirca®
- Adempas®
- Opsumit®
- Orenitram® ER
- Revatio® Tabs / Susp
- Tadalafil
- Uptravi®

Approval Criteria

- Approved Diagnosis

<table>
<thead>
<tr>
<th>ICD-9 Diagnosis</th>
<th>Description</th>
<th>ICD-10 Diagnosis</th>
<th>Description</th>
<th>Date Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>416.0</td>
<td>Primary pulmonary hypertension</td>
<td>I27.0</td>
<td>Primary pulmonary hypertension</td>
<td>730 days</td>
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<tr>
<td>416.1</td>
<td>Kyphoscoliotic heart disease</td>
<td>I27</td>
<td>Kyphoscoliotic heart disease</td>
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<tr>
<td>416.2</td>
<td>Chronic pulmonary embolism</td>
<td>I27.82</td>
<td>Chronic pulmonary embolism</td>
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<tr>
<td>416.8</td>
<td>Other chronic pulmonary heart diseases</td>
<td>I27.2</td>
<td>Other secondary pulmonary hypertension</td>
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<tr>
<td>416.9</td>
<td>Chronic pulmonary heart disease, unspecified</td>
<td>I27.89</td>
<td>Other specified pulmonary heart diseases</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>ICD-9 Diagnosis</th>
<th>Description</th>
<th>ICD-10 Diagnosis</th>
<th>Description</th>
<th>Date Range</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>I27.81</td>
<td>Cor pulmonale (chronic)</td>
<td>730 days</td>
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<td></td>
<td></td>
<td>I27.9</td>
<td>Pulmonary heart disease, unspecified</td>
<td>730 days</td>
</tr>
</tbody>
</table>

- Revatio dosing
  o 20mg three times daily
- Failure to achieve desired therapeutic outcomes with trial on 2 or more preferred agents
  o Documented trial period for preferred agents
  o Documented ADE/ADR to preferred agents
- Documented compliance on current therapy regimen
Denial Criteria

- Therapy will be denied if no approval criteria are met
- Lack of adequate trial on required preferred agents
- Nitrate therapy or Norvir therapy or Kaletra therapy in last 30 days
  - Revatio
- Pregnancy Diagnosis
  - Tracleer
  - Letairis
  - Opsumit
  - Adempas
- Drug Prior Authorization Hotline: (800) 392-8030