Executive Summary

Purpose: The MO HealthNet Pharmacy Program will implement a state-specific preferred drug list. Pulmonary arterial hypertension (PAH) is a severe, progressive, and often fatal condition that occurs idiopathically as well as in association with pulmonary, cardiac, and other thoracic conditions. Reliable estimates of the total prevalence of this condition are difficult to obtain because of the diversity of identifiable causes. PAH, formerly known as primary pulmonary hypertension, is characterized by elevations in pulmonary arterial pressure (PAP) to greater than 25 mmHg at rest and greater than 30 mmHg with exercise. The disease occurs when the PAP is abnormally elevated and forces the right side of the heart to progressively work harder when it pumps blood to the lungs. Symptoms include dyspnea, fatigue, chest pain, palpitations, syncope, and edema. Prognosis varies based on the severity of disease, whether right heart failure is present, and response to vasodilator therapy. If left untreated, the disease produces increases in PAP that may lead to right ventricular failure and death. Despite recent developments in the symptomatic treatment of PAH, there is still no cure.

PAH should be differentiated from pulmonary hypertension secondary to diseases of the heart and lung based on both pathology of the underlying disease and accepted treatments.

Why Issue Selected: Total program savings for the PDL classes will be regularly reviewed.

Program-Specific Information:

<table>
<thead>
<tr>
<th>Preferred Agents</th>
<th>Non-Preferred Agents</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Epoprostenol Injection</td>
<td>• Flolan™ Injection</td>
</tr>
<tr>
<td>• Treprostinil Infusion/SQ</td>
<td>• Remodulin® Infusion/SQ</td>
</tr>
<tr>
<td>• Veletri™ Injection</td>
<td>• Veletri™ Injection</td>
</tr>
</tbody>
</table>

Type of Criteria: ☒ Preferred Drug List  ☐ Increased risk of ADE  ☐ Appropriate Indications  ☐ Clinical Edit

Data Sources: ☐ Only Administrative Databases  ☒ Databases + Prescriber-Supplied
**Setting & Population**

- Drug class for review: Pulmonary Arterial Hypertension (PAH) Agents, Prostacyclin Pathway Agonists, Injectable
- Age range: All appropriate MO HealthNet participants

**Approval Criteria**

- Documented diagnosis of pulmonary hypertension AND
- Documented compliance on current therapy regimen OR
- Failure to achieve desired therapeutic outcomes with trial on 2 or more preferred agents, with one being generic epoprostenol:
  - Documented trial period for preferred agents OR
  - Documented ADE/ADR to preferred agents

**Denial Criteria**

- Lack of adequate trial on required preferred agents
- Therapy will be denied if all approval criteria are not met

**Required Documentation**

<table>
<thead>
<tr>
<th>Laboratory Results:</th>
<th>Progress Notes:</th>
</tr>
</thead>
<tbody>
<tr>
<td>MedWatch Form:</td>
<td>Other:</td>
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</table>

**Disposition of Edit**

Denial: Exception Code "0160" (Preferred Drug List)
Rule Type: PDL

**Default Approval Period**

1 year

**References**

3. USPDI, Micromedex; 2021.