# Pulmonary Arterial Hypertension: Oral Prostacyclins Utilization Management Criteria

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|---|---|
| **Therapeutic Class:** | Pulmonary Arterial Hypertension: Oral Prostacyclins |
| **Non-Preferred Agents:** | Orenitram (treprostinil diolamine) tablet, Uptravi (selexipag) tablet |
| **Preferred Agents:** | None |
| **Implementation Date:** | 1/1/2026 |
| **Prepared For:** | CT |
| **PDL Status:** | Non-preferred |
| **Purpose:** | Pulmonary hypertension (PH) is a disease characterized by elevated pulmonary artery pressure. The World Health Organization (WHO) classifies patients with PH into five groups: Group 1 (pulmonary arterial hypertension [PAH]), Group 2 (PH due to left heart disease), Group 3 (PH due to chronic lung disease and/or hypoxemia), Group 4 (PH due to pulmonary artery obstructions), Group 5 (PH due to unclear mechanisms). PAH is a progressive disease characterized by dyspnea, fatigue, syncope, and edema. Patients with PAH are typically evaluated for baseline risk of disease progression and death prior to the selection of therapy; risk stratification determines initial treatment regimen selection. PAH-specific agents include prostacyclin pathway agents, endothelin-1 receptor antagonists, phosphodiesterase-5 inhibitors, a soluble guanylyl cyclase stimulator, an activin-signaling inhibitor, or, more rarely used, calcium channel blockers.<br><br>Oral prostacyclin pathway agents, treprostinil diolamine and selexipag, are approved therapies for use in PAH WHO Group 1 to delay disease progression. Selexipag is also indicated in patients with PAH WHO Group 1 to reduce risk of hospitalization. Selexipag is a selective prostacyclin IP receptor agonist that is metabolized to an active metabolite to induce vasodilation of the pulmonary vascular bed and inhibit platelet aggregation. Treprostinil is also indicated in patients with PAH WHO Group 1 to improve exercise capacity. Treprostinil diolamine is a prostacyclin analogue that directly vasodilates pulmonary and systemic arterial vascular beds and inhibits platelet aggregation. Common adverse reactions with the use of selexipag or oral treprostinil diolamine include headache, diarrhea, jaw pain, nausea, vomiting, and flushing. Oral treprostinil diolamine should not be abruptly discontinued and there is a warning in patients with diverticulosis, treprostinil diolamine tablets can become lodged in a diverticulum. Pulmonary edema in patients with pulmonary veno-occlusive disease can occur with use of selexipag; if confirmed, treatment should be discontinued. |

 

**Table 1. Pulmonary Arterial Hypertension: Oral Prostacyclins**

| Generic Name | Brand Name | Approved Indications | Route of Administration | Generic Availability |
|---|---|---|---|---|
| Selexipag | Uptravi® | PAH (WHO group 1) | PO, IV | N |
| Treprostinil diolamine | Orenitram® ER | PAH (WHO group 1) | PO | N |

*Abbreviations: IV, intravenous; PAH, pulmonary arterial hypertension; PO, orally; WHO, world health organization*

***All authorizations must be prescribed in accordance with FDA approved labeling. Use of samples to initiate therapy does not meet step therapy and/or continuation of therapy prior authorization requirements. Prior therapies will be verified through pharmacy claims and/or submitted chart notes.***

**General Approval Criteria:**

- Requested quantity in accordance with FDA approved product labelling

**Initial Therapy – All the following must be met:**

- Provider has expertise in treating patients with pulmonary hypertension
- Documented diagnosis of pulmonary hypertension
- Failure to achieve desired therapeutic outcomes with a trial of **ONE** oral endothelin receptor antagonist or oral phosphodiesterase-5 inhibitor (defined as 30 day trial) **OR** documentation of adverse drug event/adverse drug reaction or contraindication to both classes

**Additional Criteria for Orenitram**

- Patient does not have severe hepatic impairment (Child Pugh Class C)

**Additional Criteria for Uptravi**

- Patient is not taking concomitant strong CYP2C8 inhibitors (e.g. gemfibrozil)

**Initial PA length: 1 year**

**Exclusion Criteria:** Approval criteria not met

**Continuation Therapy:** Documented compliance on current therapy regimen **AND** Documented continued clinical benefit

**Continuation Length: 1 year**

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## Revision History

| Date | Version | Revisions |
|---|---|---|
| 11/7/2025 | V1 | Document approved by DSS |