CareSource Rxinnovations

# PHARMACY POLICY STATEMENT
## Marketplace

| DRUG NAME | Oral Prostacyclins for Pulmonary Arterial Hypertension: Orenitram (treprostinil extended-release), Uptravi (selexipag tablets) |
|---|---|
| BENEFIT TYPE | Pharmacy |
| STATUS | Prior Authorization Required |

Pulmonary Arterial Hypertension (PAH) is a rare but serious condition characterized by elevated pulmonary arterial resistance. Orenitram and Uptravi are approved for the treatment of PAH World Health Organization (WHO) Group 1. Orenitram is indicated to delay disease progression and to improve exercise capacity. Uptravi is approved to delay disease progression and reduce the risk of hospitalization for PAH.

Oral Prostacyclins will be considered for coverage when the following criteria are met:

## Pulmonary Arterial Hypertension [WHO Group 1]

For **initial** authorization:

1. Member is at least 18 years of age or older; AND
2. Medication must be prescribed by or in consultation with a cardiologist or pulmonologist; AND
3. Member must have a diagnosis of WHO Group 1 PAH confirmed by right heart catheterization; AND
4. Member must have documentation of **one** of the following:
   a) Patient had an acute response to vasodilator testing AND has tried a calcium channel blocker (CCB) for at least 3 months;
   b) Patient did not have a response to vasodilator testing;
   c) Patient cannot undergo vasodilator testing;
   d) Patient cannot take CCB therapy; AND
5. Member has tried and failed an inhaled or injectable prostacyclin; AND
6. Member has tried and failed **one** of the following oral medications: phosphodiesterase type 5 inhibitor (ie. Sildenafil, Tadalafil), endothelin receptor antagonist (ie. Ambrisentan, Bosentan, Macitentan), or Soluble Guanylate Cyclase Stimulator (ie. Adempas); OR
7. Member has WHO functional class III symptoms with rapid progression of disease (see appendix); OR
8. Member has WHO functional class IV symptoms (see appendix).

**Dosage allowed/Quantity limit:**

Orenitram: Initiate 0.125 mg three times daily or 0.25 mg twice daily; Titrate by 0.125 mg three times daily or by 0.25 mg or 0.5 mg twice daily.

Uptravi: Initiate 200 mcg twice daily; Increase by 200 mcg twice daily usually at weekly intervals (maximum dose of 1600 mcg twice daily). Quantity Limit: 3,200 mcg per day.

***If all the above requirements are met, the medication will be approved for 6 months.***

For **reauthorization**:

1. Member has documentation of improvement in signs and symptoms of disease as evidenced by at least one of the following:
   a) Stabilization or improvement in functional class symptoms or quality of life;
   b) Stabilization or improvement in 6MWD (6-minute walk distance).

***If all the above requirements are met, the medication will be approved for an additional 12 months.***

**CareSource considers Oral Prostacyclins not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.**

| DATE | ACTION/DESCRIPTION |
|---|---|
| **06/15/2011** | Pulmonary Arterial Hypertension policy creation. |
| **05/13/2014** | Combined all PAH agents into one policy |
| **07/09/2015** | Revised guidelines for therapy aligning with CMS |
| **08/18/2015** | Revised guidelines to include diagnosis criteria |
| **10/13/2021** | Separated PAH agents by drug class; Updated guidelines; Added provider specialty |
| **05/04/2023** | Updated guidelines; Added quantity limits; updated trials to exclude WHO FC III with rapid progression and IV; Added trial of injectable/inhaled prostacyclin. |

References:


1. Orenitram [package insert]. Research Triangle Park, NC: United Therapeutics Corp; February 2023.
2. Uptravi [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US, Inc.; July 2022.
3. Coons, J.C., Pogue, K., Kolodziej, A.R. et al. Pulmonary Arterial Hypertension: a Pharmacotherapeutic Update. Curr Cardiol Rep. 2019; 21(141)
4. Klinger JR, Elliott CG et al. Therapy for Pulmonary Arterial Hypertension in Adults; Chest Journal. March 2019; 155(3): 565-586
5. Humbert M, Kovacs G, Hoeper MM, et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. Eur Respir J. 2023;61(1):2200879. Published 2023 Jan 6. doi:10.1183/13993003.00879-2022


Effective date: 10/01/2023
Revised date: 05/04/2023

**Appendix:**

| World Health Organization Functional Assessment Classification | |
|---|---|
| **Class I** | Patients with PAH but without resulting limitation of physical activity. Ordinary physical activity does not cause undue dyspnea, fatigue, chest pain or near syncope. |
| **Class II** | Patients with PAH resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity increases dyspnea, fatigue, chest pain, or near syncope. |
| **Class III** | Patients with PAH resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity increases dyspnea, fatigue, chest pain, or near syncope. |
| **Class IV** | Patients with PAH unable to carry out any physical activity without symptoms. These patients may have signs of right-heart failure. Dyspnea and/or fatigue may even be present at rest. Discomfort is increased by any physical activity. |