COMMON GROUND
HEALTHCARE COOPERATIVE

PHARMACY POLICY STATEMENT
Common Ground Healthcare Cooperative (CGHC)

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).
9. **Dosage allowed/Quantity limit:**
   a) 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.
   b) 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.

COMMON GROUND
HEALTHCARE COOPERATIVE

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.
04/30/2024	Updated references; removed PAH diagnosis from appendix.

Appendix:

World Health Organization Functional Assessment Classification	
Class I	Patients without resulting limitation of physical activity. Ordinary physical activity does not cause undue dyspnea, fatigue, chest pain or near syncope.
Class II	Patients with 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 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 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.

References:

1. Orenitram [package insert]. Research Triangle Park, NC: United Therapeutics Corp; 2023.
2. Uptravi [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US, Inc.; 2022.
3. Coons JC, Pogue K, Kolodziej AR, Hirsch GA, George MP. Pulmonary Arterial Hypertension: a Pharmacotherapeutic Update. *Curr Cardiol Rep.* 2019;21(11):141. Published 2019 Nov 22. doi:10.1007/s11886-019-1235-4

4. Klinger JR, Elliott CG, Levine DJ, et al. Therapy for Pulmonary Arterial Hypertension in Adults: Update of the CHEST Guideline and Expert Panel Report [published correction appears in Chest. 2021 Jan;159(1):457]. *Chest*. 2019;155(3):565-586. doi:10.1016/j.chest.2018.11.03
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: 01/01/2025
Revised date: 04/30/2024