Medical Necessity Guidelines

Medical Benefit Drugs

Pulmonary Hypertension Medications: Epoprostenol Products, Remodulin® (trepostinil), Tyvaso® (treprostinil), Ventavis® (iloprost)

Effective: January 1, 2026

Guideline Type	☒ Prior Authorization ☐ Non-Formulary ☐ Step-Therapy ☐ Administrative

Applies to:

☒ CarePartners of Connecticut Medicare Advantage HMO plans, Fax 617-673-0956

☒ CarePartners of Connecticut Medicare Advantage PPO plans, Fax 617-673-0956

Note: While you may not be the provider responsible for obtaining prior authorization, as a condition of payment you will need to ensure that prior authorization has been obtained.

Overview

Pulmonary arterial hypertension (PAH) is a rare but life-threatening disorder characterized by hypertension in the arteries of the lungs and shortness of breath and fatigue following exertion. Interstitial lung disease (ILD) is a group of diseases characterized by marked scarring or fibrosis of the lungs.

Food and Drug Administration - Approved Indications

Flolan (epoprostenol) is a prostacyclin vasodilator indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to improve exercise capacity. Studies establishing effectiveness included predominantly (97%) patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH (49%) or PAH associated with connective tissue diseases (51%).

Remodulin (treprostinil) is a prostacyclin vasodilator indicated for:

- Treatment of PAH (WHO Group 1) to diminish symptoms associated with exercise. Studies establishing effectiveness included patients with NYHA Functional Class II-IV symptoms and etiologies of idiopathic or heritable PAH (58%), PAH associated with congenital systemic-to- pulmonary shunts (23%), or PAH associated with connective tissue diseases (19%).
- In patients with PAH requiring transition from epoprostenol to diminish the rate of clinical deterioration. The risks and benefits of each drug should be carefully considered prior to transition.

Tyvaso (treprostinil) is a prostacyclin mimetic indicated for the treatment of:

- PAH (WHO Group 1) to improve exercise ability. Studies establishing effectiveness predominately included patients with NYHA Functional Class III symptoms and etiologies of idiopathic or heritable PAH (56%) or PAH associated with connective tissue diseases (33%).
- Pulmonary hypertension associated with ILD (WHO Group 3) to improve exercise ability. The study establishing effectiveness predominately included patients with etiologies of idiopathic interstitial pneumonia (IIP) (45%) inclusive of idiopathic pulmonary fibrosis, combined pulmonary fibrosis and emphysema (CPFE) (25%), and WHO Group 3 connective tissue disease (22%).

Veletri (epoprostenol) is a prostacyclin vasodilator indicated for the treatment of PAH (WHO Group 1) to improve exercise capacity. Studies establishing effectiveness included predominantly patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH or PAH associated with connective tissue diseases.

Ventavis (iloprost) is a synthetic analog of prostacyclin indicated for the treatment of PAH (WHO Group 1) to improve a composite endpoint consisting of exercise tolerance, symptoms (NYHA Class), and lack of deterioration. Studies establishing effectiveness included predominately patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH (65%) or PAH associated with connective tissue diseases (23%).

Clinical Guideline Coverage Criteria

The plan may authorize a Pulmonary Hypertension Medication for Members when all the following criteria is met:

1. Documentation of **one (1)** of the following:
    a. Documented diagnosis of pulmonary arterial hypertension (PAH) (WHO Group 1)

b. If the request is for Tyvaso, documented diagnosis of pulmonary arterial hypertension associated with interstitial lung disease (WHO Group 3)

AND

2. Prescribed by a cardiologist or pulmonologist

Limitations

- None

Codes

The following code(s) require prior authorization:

Table 1: HCPCS Codes

HCPCS Codes	Description
J1325	Injection, epoprostenol, 0.5 mg
J3285	Injection treprostinil, 1 mg
J7686	Treprostinil, inhalation solution, FDA-approved final product, non-compounded, administered through DME, unit dose form, 1.74 mg
Q4074	Iloprost, inhalation solution, administered through DME, up to 20mcg

References

1. Flolan (epoprostenol sodium) [package insert]. Research Triangle Park, NC: GlaxoSmithKline; August 2021.
2. Humbert M, et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. European Heart Journal. 2022 Oct;43(38):3618–31.
3. Klinger JR, et al. Therapy for Pulmonary Arterial Hypertension in Adults 2018: Update of the CHEST Guideline and Expert Panel Report. Chest. 2019 Mar;155(3).
4. McLaughlin VV, et al. Results of an Expert Consensus Survey on the Treatment of Pulmonary Arterial Hypertension With Oral Prostacyclin Pathway Agents. Chest. 2020;157(4):955-965
5. McLaughlin VV, et al. ACCF/AHA 2009 Expert Consensus Document on Pulmonary Hypertension. JACC. 2009;53(17):1573–619.
6. Remodulin (treprostinil) injection [package insert]. Research Triangle Park, NC: United Therapeutics, Corp; February 2021.
7. Tyvaso (treprostinil) [package insert]. Research Triangle Park, NC: United Therapeutics, Corp; May 2022.
8. Veletri (epoprostenol for injection) [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US, Inc.; October 2020.
9. Ventavis (iloprost) [package insert]. South San Francisco: Actelion Pharmaceuticals US, Inc. March 2022.

Approval And Revision History

September 13, 2022: Reviewed by Pharmacy and Therapeutics Committee (P&T)

Subsequent endorsement date(s) and changes made:

- September 21, 2022: Reviewed by the Medical Policy Approval Committee (MPAC)
- December 12, 2023: Updated the title of the Medical Necessity Guideline from Ventavis to Pulmonary Hypertension Medications: Epoprostenol products, Remodulin, Tyvaso, Veletri, Ventavis. Added epoprostenol products, Remodulin, Tyvaso, and Veletri. For Remodulin, removed “The Member is transitioning from epoprostenol to Remodulin (treprostinil) to reduce the rate of clinical deterioration.” For Flolan, removed Flolan® (epoprostenol) is being prescribed to improve exercise capacity. Removed the Limitation Any indications other than FDA-approved indications are considered experimental or investigational and will not be approved by the health plan. Added provider specialty requirements. Administrative Update in support of calendar year 2024 Medicare Advantage and PDP Final Rule (eff 3/1/24).
- November 12, 2024: No changes (eff 1/1/25)
- December 2024: Joint Medical Policy and Health Care Services UM Committee review (eff 1/1/25)
- December 9, 2025: No changes (eff 1/1/26)
- December 2025: Joint Medical Policy and Health Care Services UM Committee review (effective 1/1/26)

Background, Product and Disclaimer Information

Point32Health prior authorization criteria to be applied to Medicare Advantage plan members is based on guidance from Medicare laws, National Coverage Determinations (NCDs) or Local Coverage Determinations (LCDs). When no guidance is

provided, Point32Health uses clinical practice guidance published by relevant medical societies, relevant medical literature, Food and Drug Administration (FDA)-approved package labeling, and drug compendia to develop prior authorization criteria to apply to Medicare Advantage plan members. Medications that require prior authorization generally meet one or more of the following criteria: Drug product has the potential to be used for cosmetic purposes; drug product is not considered as first-line treatment by medically accepted practice guidelines, evidence to support the safety and efficacy of a drug product is poor, or drug product has the potential to be used for indications outside of the indications approved by the FDA. Prior authorization and use of the coverage criteria within this Medical Necessity Guideline will ensure drug therapy is medically necessary, clinically appropriate, and aligns with evidence-based guidelines. We revise and update Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests revisions.

Treating providers are solely responsible for the medical advice and treatment of Members. The use of this guideline is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to eligibility and benefits on the date of service, coordination of benefits, referral/authorization, utilization management guidelines when applicable, and adherence to plan policies, plan procedures, and claims editing logic.