

SPECIALTY GUIDELINE MANAGEMENT

Tyvaso (treprostinil inhalation solution) Tyvaso DPI (treprostinil inhalation powder)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indications

- A. Treatment of Pulmonary arterial hypertension (PAH; World Health Organization [WHO] Group 1) to improve exercise ability. Studies establishing effectiveness predominately included patients with New York Heart Association (NYHA) Functional Class III symptoms and etiologies of idiopathic or heritable PAH or PAH associated with connective tissue diseases.
- B. Treatment of Pulmonary hypertension associated with interstitial lung disease (PH-ILD; WHO Group 3) to improve exercise ability. The study establishing effectiveness predominately included patients with etiologies of idiopathic interstitial pneumonia (IIP) inclusive of idiopathic pulmonary fibrosis (IPF), combined pulmonary fibrosis and emphysema (CPFE), and WHO Group 3 connective tissue disease.

All other indications are considered experimental/investigational and not medically necessary.

II. PRESCRIBER SPECIALTIES

This medication must be prescribed by or in consultation with a pulmonologist or cardiologist.

III. CRITERIA FOR INITIAL APPROVAL

Pulmonary Hypertension (PH)

Authorization of 12 months may be granted for treatment of PH when both of the following criteria are met:

- A. Member has either of the following:
 1. WHO Group 1 class of pulmonary hypertension (refer to Appendix)
 2. Pulmonary hypertension associated with interstitial lung disease (WHO Group 3)
- B. PH was confirmed by either criterion (1) or criterion (2) below:
 1. Pretreatment right heart catheterization with all of the following results:
 - i. Mean pulmonary arterial pressure (mPAP) > 20 mmHg
 - ii. Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
 - iii. Pulmonary vascular resistance (PVR) ≥ 3 Wood units in adult patients or pulmonary vascular resistance index (PVRI) ≥ 3 Wood units x m² in pediatric patients
 2. For infants less than one year of age, PH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed.

IV. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for members with an indication listed in Section III who are currently receiving the requested medication through a paid pharmacy or medical benefit, and who are experiencing benefit from therapy as evidenced by disease stability or disease improvement.

V. APPENDIX

WHO Classification of Pulmonary Hypertension

1 PAH

- 1.1 Idiopathic (PAH)
- 1.2 Heritable PAH
- 1.3 Drug- and toxin-induced PAH
- 1.4. PAH associated with:
 - 1.4.1 Connective tissue diseases
 - 1.4.2 HIV infection
 - 1.4.3 Portal hypertension
 - 1.4.4 Congenital heart diseases
 - 1.4.5 Schistosomiasis
- 1.5 PAH long-term responders to calcium channel blockers
- 1.6 PAH with overt features of venous/capillaries (PVOD/PCH) involvement
- 1.7 Persistent PH of the newborn syndrome

2 PH due to left heart disease

- 2.1 PH due to heart failure with preserved LVEF
- 2.2 PH due to heart failure with reduced LVEF
- 2.3 Valvular heart disease
- 2.4 Congenital/acquired cardiovascular conditions leading to post-capillary PH

3 PH due to lung diseases and/or hypoxia

- 3.1 Obstructive lung disease
- 3.2 Restrictive lung disease
- 3.3 Other lung disease with mixed restrictive/obstructive pattern
- 3.4 Hypoxia without lung disease
- 3.5 Developmental lung disorders

4 PH due to pulmonary artery obstruction

- 4.1 Chronic thromboembolic PH
- 4.2 Other pulmonary artery obstructions
 - 4.2.1 Sarcoma (high or intermediate grade) or angiosarcoma
 - 4.2.2 Other malignant tumors
 - Renal carcinoma
 - Uterine carcinoma
 - Germ cell tumours of the testis
 - Other tumours
 - 4.2.3 Non-malignant tumours
 - Uterine leiomyoma
 - 4.2.4 Arteritis without connective tissue disease
 - 4.2.5 Congenital pulmonary artery stenosis
 - 4.2.6 Parasites
 - Hydatidosis

5 PH with unclear and/or multifactorial mechanisms

- 5.1 Hematologic disorders: Chronic hemolytic anemia, myeloproliferative disorders

- 5.2 Systemic and metabolic disorders: Pulmonary Langerhans cell histiocytosis, Gaucher disease, glycogen storage disease, neurofibromatosis, sarcoidosis
- 5.3 Others: chronic renal failure with or without hemodialysis, fibrosing mediastinitis
- 5.4 Complex congenital heart disease

VI. REFERENCES

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