

SPECIALTY GUIDELINE MANAGEMENT

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

POLICY

I. CRITERIA FOR INITIAL APPROVAL

Pulmonary Hypertension (PH)

Authorization of 6 months may be granted for treatment of PH when both of the following criteria are met:

- A. Prescriber is either one of the following:
 - 1. Prescriber is associated with an accredited Center of Comprehensive Care by the Pulmonary Hypertension Association for adult members
 - 2. Prescriber is a cardiologist, pulmonologist or physician specializing in PAH for pediatric members
- B. 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. mPAP > 20 mmHg
 - ii. PCWP ≤ 15 mmHg
 - iii. PVR ≥ 3 Wood units
 - 2. For infants less than one year of age, PH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed.

II. CONTINUATION OF THERAPY

Authorization of 6 months may be granted for members with an indication listed in Section I who are currently receiving the requested medication through a paid pharmacy benefit, and who are experiencing benefit from therapy as evidenced by disease stability or disease improvement.

III. QUANTITY LIMITS

- a. Tyvaso 0.6mg/ml – 2.9 ml (1 amp) per day
- b. Tyvaso DPI 16mcg, 32mcg, 48mcg and 64mcg – 4 cartridges per day
- c. Tyvaso DPI Titration Kit 16-32mcg – 7 cartridges per day
- d. Tyvaso DPI Titration Kit 32-48mcg – 8 cartridges per day
- e. Tyvaso DPI Titration Kit 16-32-48mcg – 9 cartridges per day

IV. 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

V. REFERENCES

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