Name:	Tyvaso	Page:	1 of 4
Effective Date:	12/21/2023	Last Review Date:	11/2023

Applies to:	☒Illinois	☐Florida	☒Florida Kids
	☐New Jersey	☒Maryland	☐Michigan
	☒Pennsylvania Kids	☐Virginia	☐Kentucky PRMD

Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Tyvaso under the patient's prescription drug benefit.

Description:

FDA-Approved Indications

A. Treatment of Pulmonary arterial hypertension (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 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.

Applicable Drug List:

Tyvaso

Policy/Guideline:

Prescriber Specialty

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

Criteria for Initial Approval

Pulmonary Hypertension (PH)

Authorization of 12 months may be granted for treatment of PH when all 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. mPAP > 20 mmHg

ii. PCWP ≤ 15 mmHg
iii. Pulmonary vascular resistance (PVR) ≥ 3 Wood units in adult patients or pulmonary vascular resistance index (PVRI) ≥ 3 Wood units x m^2 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.

C. WHO Group 1 pulmonary hypertension: Patient is unable to take the required number of formulary alternatives (3) for the given diagnosis due to a trial and inadequate treatment response or intolerance, or a contraindication.

Criteria for Continuation of Therapy
Authorization of 12 months may be granted for members with an indication listed in criteria for initial approval 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.

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

Approval Duration and Quantity Restrictions:
Approval: 12 months

Quantity Level Limit:
- Tyvaso (treprostinil) 0.6 mg/mL inhalation solution: 81.2 mL (28 ampules) per 28 days
- Tyvaso (treprostinil) DPI 16 mcg and 32 mcg Inhalation Powder Titration Kit: 196 cartridges per 28 days
- Tyvaso (treprostinil) DPI 16 mcg, 32 mcg, and 48 mcg Inhalation Powder Titration Kit: 252 cartridges per 28 days

- Tyvaso (treprostinil) DPI 16 mcg Inhalation Powder Maintenance Kit: 112 cartridges per 28 days
- Tyvaso (treprostinil) DPI 32 mcg Inhalation Powder Maintenance Kit: 112 cartridges per 28 days
- Tyvaso (treprostinil) DPI 48 mcg Inhalation Powder Maintenance Kit: 112 cartridges per 28 days
- Tyvaso (treprostinil) DPI 64 mcg Inhalation Powder Maintenance Kit: 112 cartridges per 28 days
- Tyavso (treprostinil) DPI 32 mcg and 48 mcg Inhalation Powder Maintenance Kit: 224 cartridges per 28 days

References:

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10. Klinger JR, Elliott CG, Levine DJ, et al. Therapy for Pulmonary Arterial Hypertension in Adults: Update of the CHEST Guidelines and Expert Panel Report. *Chest*. 2019:155(3): 565-586.
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