AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name:	Tyvaso	Page:	1 of 5
Effective Date:	2/28/2025	Last Review Date:	1/2025

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:

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[1,2]

- 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.
- 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.

Coverage Criteria

Pulmonary Hypertension (PH)[1-7]

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

- Member has either of the following criteria:
  - WHO Group 1 class of pulmonary hypertension (refer to Appendix)
  - Pulmonary hypertension associated with interstitial lung disease (PH-ILD; WHO Group 3)
- PH was confirmed by either of the following:
  - Pretreatment right heart catheterization with all of the following results:
- Mean pulmonary arterial pressure (mPAP) > 20 mmHg
- Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
- Pulmonary vascular resistance (PVR) > 2 Wood units. For pediatric members, pulmonary vascular resistance index (PVRI) > 3 Wood units x m^2 is also acceptable.
  - For infants less than one year of age, PH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed.
- 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.

Continuation of Therapy

Authorization of 12 months may be granted for members with an indication listed in the coverage criteria section 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 (PH)[4]

Note: Patients with heritable PAH or PAH associated with drugs and toxins might be long-term responders to calcium channel blockers.

Group 1: Pulmonary Arterial Hypertension (PAH)

- Idiopathic
  - Long-term responders to calcium channel blockers
- Heritable
- Associated with drugs and toxins
- Associated with:

  - Connective tissue disease
  - Human immunodeficiency virus (HIV) infection
  - Portal hypertension
  - Congenital heart disease
  - Schistosomiasis
- PAH with features of venous/capillary (pulmonary veno-occlusive disease [PVOD]/pulmonary capillary hemangiomatosis [PCH]) involvement
- Persistent PH of the newborn

Group 2: PH associated with Left Heart Disease

- Heart failure:
  - With preserved ejection fraction
  - With reduced or mildly reduced ejection fraction
  - Cardiomyopathies with specific etiologies (i.e., hypertrophic, amyloid, Fabry disease, and Chagas disease)
- Valvular heart disease:
  - Aortic valve disease
  - Mitral valve disease
  - Mixed valvular disease
- Congenital/acquired cardiovascular conditions leading to post-capillary PH

Group 3: PH associated with Lung Diseases and/or Hypoxia

- Chronic obstructive pulmonary disease (COPD) and/or emphysema
- Interstitial lung disease
- Combined pulmonary fibrosis and emphysema
- Other parenchymal lung diseases (i.e., parenchymal lung diseases not included in Group 5)
- Nonparenchymal restrictive diseases:
  - Hypoventilation syndromes
  - Pneumonectomy
- Hypoxia without lung disease (e.g., high altitude)
- Developmental lung diseases

Group 4: PH associated with Pulmonary Artery Obstructions

- Chronic thromboembolic PH
- Other pulmonary artery obstructions:
  - Sarcomas (high- or intermediate-grade or angiosarcoma)

- Other malignant tumors (e.g., renal carcinoma, uterine carcinoma, germ-cell tumors of the testis)
- Non-malignant tumors (e.g., uterine leiomyoma)
- Arteritis without connective tissue disease
- Congenital pulmonary artery stenoses
- Hydatidosis

Group 5: PH with Unclear and/or Multifactorial Mechanisms

- Hematological disorders, including inherited and acquired chronic hemolytic anemia and chronic myeloproliferative disorders
- Systemic disorders: Sarcoidosis, pulmonary Langerhans cell histiocytosis, and neurofibromatosis type 1
- Metabolic disorders, including glycogen storage diseases and Gaucher disease
- Chronic renal failure with or without hemodialysis
- Pulmonary tumor thrombotic microangiopathy
- Fibrosing mediastinitis
- 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:

1. Tyvaso [package insert]. Research Triangle Park, NC: United Therapeutics Corp.; May 2022.

2. Tyvaso DPI [package insert]. Research Triangle Park, NC: United Therapeutics Corp.; November 2023.
3. Simonneau G, Montani D, Celermajer DS, et al. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. Eur Respir J. 2019;53(1):1801913. doi:10.1183/13993003.01913-2018.
4. Kovacs G, Bartolome S, Denton CP, et al. Definition, classification and diagnosis of pulmonary hypertension. Eur Respir J. 2024;64(4):2401324. doi: 10.1183/13993003.01324-2024
5. Chin KM, Gaine SP, Gerges C, et al. Treatment algorithm for pulmonary arterial hypertension. Eur Respir J. 2024;64(4):2401325. doi: 10.1183/13993003.01325-2024
6. Ivy D, Rosenzweig EB, Abman SH, et al. Embracing the challenges of neonatal and paediatric pulmonary hypertension. Eur Respir J. 2024;64(4):2401345. doi: 10.1183/13993003.01345-2024
7. Shlobin OA, Adir Y, Barbera JA, et al. Pulmonary hypertension associated with lung diseases. Eur Respir J. 2024;64(4):2401200. doi: 10.1183/13993003.01200-2024