AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name:	Bosentan	Page:	1 of 4
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 Tracleer (bosentan) 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]

Indicated for the treatment of pulmonary arterial hypertension (PAH) (World Health Organization [WHO] Group 1):

- In adults to improve exercise ability and to decrease clinical worsening. Studies establishing effectiveness included predominantly patients with WHO Functional Class II-IV symptoms and etiologies of idiopathic or heritable PAH, PAH associated with connective tissue diseases, and PAH associated with congenital heart disease with left-to-right shunts.
- In pediatric patients aged 3 years and older with idiopathic or congenital PAH to improve pulmonary vascular resistance (PVR), which is expected to result in an improvement in exercise ability.

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

Applicable Drug List:

Bosentan 125 mg tablets
Bosentan 62.5 mg tablets
Tracleer (bosentan) 32 mg tablets for suspension

Policy/Guideline:

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

Coverage Criteria:
Pulmonary Arterial Hypertension (PAH)[1-6]
Authorization of 12 months may be granted for treatment of PAH when ALL of the following criteria are met:

- Member has PAH defined as WHO Group 1 class of pulmonary hypertension (refer to Appendix).
- PAH was confirmed by either of the following criteria:
  - 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, PAH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed.

Criteria for 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:

- Bosentan 125 mg tablets: 60 per 30 days
- Bosentan 62.5mg tablets: 60 per 30 days
- Tracleer 32 mg tablets: 112 per 28 days

References:

1. Tracleer [package insert]. Titusville, NJ: Actelion Pharmaceuticals US, Inc.; February 2024.
2. Bosentan [package insert]. Parsippany, NJ: Teva Pharmaceuticals. March 2024.
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