

## SPECIALTY GUIDELINE MANAGEMENT

### TRACLEER (bosentan) bosentan

#### 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 Indication

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

- A. 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.
- B. 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.

#### II. PRESCRIBER SPECIALTIES

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

#### III. CRITERIA FOR INITIAL APPROVAL

##### **Pulmonary arterial hypertension (PAH)**

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

- A. Member has PAH defined as WHO Group 1 class of pulmonary hypertension (refer to Appendix).
- B. PAH 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 members or pulmonary vascular resistance index (PVRI) ≥ 3 Wood units x m<sup>2</sup> in pediatric members
  2. For infants less than one year of age, PAH 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 (PH)

#### 1 Pulmonary arterial hypertension (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 disease
  - 1.4.2 Human immunodeficiency virus (HIV) infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease
  - 1.4.5 Schistosomiasis
- 1.5 PAH long-term responders to calcium channel blockers
- 1.6 PAH with overt features of venous/capillaries (pulmonary veno-occlusive disease [PVOD]/pulmonary capillary hemangiomatosis [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 left ventricular ejection fraction (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 obstructions

- 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 tumors of the testis
    - Other tumors
  - 4.2.3 Non-malignant tumors
    - 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

Reference number(s)
1649-A

- 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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