



AETNA BETTER HEALTH®  
Coverage Policy/Guideline

Name: Opsumit

Page: 1 of 4

Effective Date: 12/21/2023

Last Review Date: 11/2023

Applies to:	<input checked="" type="checkbox"/> Illinois	<input type="checkbox"/> Florida	<input type="checkbox"/> Florida Kids
	<input type="checkbox"/> New Jersey	<input type="checkbox"/> Maryland	<input type="checkbox"/> Michigan
	<input type="checkbox"/> Pennsylvania Kids	<input type="checkbox"/> Virginia	<input type="checkbox"/> Kentucky PRMD

### Intent:

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

### Description:

#### FDA-Approved Indication

Opsumit is an endothelin receptor antagonist (ERA) indicated for the treatment of pulmonary arterial hypertension (PAH) (World Health Organization [WHO] Group 1) to reduce the risks of disease progression and hospitalization for PAH. Effectiveness was established in a long-term study in PAH patients with predominantly WHO Functional Class II-III symptoms treated for an average of 2 years. Patients had idiopathic and heritable PAH, PAH caused by connective tissue disorders, and PAH caused by congenital heart disease with repaired shunts.

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

### Applicable Drug List:

Opsumit

### Policy/Guideline:

#### Prescriber Specialties:

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

### 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. mPAP > 20 mmHg
    - ii. PCWP ≤ 15 mmHg



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- iii. Pulmonary vascular resistance (PVR)  $\geq 3$  Wood units in adult patients or pulmonary vascular resistance index (PVRI)  $\geq 3$  Wood units  $\times m^2$  in pediatric patients
- 2. For infants less than one year of age, PAH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed.
- C. 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



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### 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:** Opsumit 10 mg tablets: 30 per 30 days

#### References:

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11. Simonneau G, Montani D, Celermajer DS, et al. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. *Eur Respir J.* 2019;53:1801913; doi:10.1183/13993003.01913-2018.