



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 type="checkbox"/> Illinois	<input type="checkbox"/> Florida	<input checked="" type="checkbox"/> Florida Kids
	<input checked="" type="checkbox"/> New Jersey	<input checked="" type="checkbox"/> Maryland	<input type="checkbox"/> Michigan
	<input checked="" 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



AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name: Opsumit

Page: 2 of 4

Effective Date: 12/21/2023

Last Review Date: 11/2023

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

- iii. Pulmonary vascular resistance (PVR) ≥ 3 Wood units in adult patients or pulmonary vascular resistance index (PVRI) ≥ 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.

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



AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name: Opsumit

Page: 3 of 4

Effective Date: 12/21/2023

Last Review Date: 11/2023

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

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:

1. Opsumit [package insert]. Titusville, NJ: Actelion Pharmaceuticals US, Inc.; July 2022.
2. Chin KM, Rubin LJ. Pulmonary arterial hypertension. *J Am Coll Cardiol.* 2008;51(16):1527-1538.
3. McLaughlin VV, Archer SL, Badesch DB, et al. ACCF/AHA 2009 expert consensus document on pulmonary hypertension a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association developed in collaboration with the American College of Chest Physicians, American Thoracic Society, Inc., and the Pulmonary Hypertension Association. *J Am Coll Cardiol.* 2009;53(17):1573-1619.
4. Badesch DB, Champion HC, Gomez-Sanchez MA, et al. Diagnosis and assessment of pulmonary arterial hypertension. *J Am Coll Cardiol.* 2009;54:S55-S66.



AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name: Opsumit Page: 4 of 4

Effective Date: 12/21/2023 Last Review Date: 11/2023

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

5. Rubin LJ. American College of Chest Physicians. Diagnosis and management of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. *Chest*. 2004;126(1 Suppl):7S-10S.
6. Barst RJ, Gibbs SR, Ghofrani HA, et al. Updated evidence-based treatment algorithm in pulmonary arterial hypertension. *J Am Coll Cardiol*. 2009;54:S78-S84.
7. Taichman DB, Ornelas J, Chung L, et al. Pharmacologic therapy for pulmonary arterial hypertension in adults. CHEST guideline and expert panel report. *Chest*. 2014;46(2):449-475.
8. Abman SH, Hansmann G, Archer SL, et al. Pediatric pulmonary hypertension: guidelines from the American Heart Association and American Thoracic Society. *Circulation*. 2015;132(21):2037-99.
9. 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.
10. Galie N, McLaughlin VV, Rubin LJ, Simonneau G. An overview of the 6th World Symposium on Pulmonary Hypertension. *Eur Respir J*. 2019; 53: 1802148; DOI: 10.1183/13993003.02148-2018. Published 24 January 2019.
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.