Reference number(s)
6454-A

SPECIALTY GUIDELINE MANAGEMENT

OPSYNVI (macitentan and tadalafil)

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
Opsynvi is the combination of macitentan and tadalafil indicated for the chronic treatment of adults with pulmonary arterial hypertension (PAH, World Health Organization [WHO] Group I and WHO Functional Class [FC] II-III). Individually, macitentan reduces the risk of clinical worsening events and hospitalization, and tadalafil improves 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:
1. Member has PAH defined as WHO Group 1 class of pulmonary hypertension (refer to Appendix).
2. PAH was confirmed by right heart catheterization with all of the following pretreatment 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

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
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

1. Opsynvi [package insert]. Titusville, NJ: Actelion Pharmaceuticals US, Inc.; March 2024.
2. Galie N, McLaughlin VV, Rubin LJ, Simonneau G. An overview of the 6th World Symposium on Pulmonary Hypertension. *Eur Respir J.* 2019;53(1):1802148. doi: 10.1183/13993003.02148-2018

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