Reference number(s)
6455-A

Specialty Guideline Management Winrevair

Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Winrevair	sotatercept-csrk

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]

Winrevair is indicated for the treatment of adults with pulmonary arterial hypertension (PAH, World Health Organization [WHO] Group 1) to increase exercise capacity, improve WHO functional class (FC), and reduce the risk of clinical worsening events.

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

Documentation

Submission of the following information is necessary to initiate the prior authorization review for initial requests: Chart notes, medical record documentation, or claims history supporting current pulmonary arterial hypertension (PAH) therapy.

Reference number(s)
6455-A

Prescriber Specialties

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

Coverage Criteria

Pulmonary Arterial Hypertension (PAH)[1-7]

Authorization of 12 months may be granted for treatment of PAH in members 18 years of age and older 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 right heart catheterization with all of the following pretreatment (before any PAH therapy) results:
    - Mean pulmonary arterial pressure (mPAP) > 20 mmHg
    - Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
    - Pulmonary vascular resistance (PVR) > 2 Wood units
- The requested medication will be used as add-on therapy.
- Member is currently receiving PAH therapy with medications from at least two of the following drug classes:
    - Endothelin receptor antagonist (e.g., Letairis, Opsumit, Tracleer)
    - Phosphodiesterase-5 inhibitor (e.g., Adcirca, Revatio)
    - Soluble guanylate cyclase stimulator (e.g., Adempas)
    - Prostacyclin analog (e.g., Flolan, Orenitram, Remodulin, Tyvaso, Veletri, Ventavis)
    - Prostacyclin receptor agonist (e.g., Uptravi)

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)[6]

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

References

1. Winrevair [package insert]. Rahway, NJ: Merck Sharp & Dohme LLC; March 2024.
2. Hoeper MM, Badesch DB, Ghofrani HA, et al. Phase 3 trial of sotatercept for treatment of pulmonary arterial hypertension. N Engl J Med. 2023;388(16):1478-1490. doi: 10.1056/NEJMoa2213558
3. Hoeper MM, Badesch DB, Ghofrani HA, et al. Phase 3 trial of sotatercept for treatment of pulmonary arterial hypertension. Supplementary appendix. N Engl J Med. 2023;Suppl Appendix.
4. 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
5. Acceleron Pharma, Inc. A Study of Sotatercept for the Treatment of Pulmonary Arterial Hypertension (MK-7962-003/A011-11)(STELLAR). In: ClinicalTrials.gov [Internet]. Bethesda (MD): National Library of Medicine (US). 2000- [4/25/2024]. Available from: https://clinicaltrials.gov/study/NCT04576988. NLM Identifier: NCT04576988.
6. 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
7. 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