Intent:

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

Description:

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]

Uptravi is indicated for the treatment of pulmonary arterial hypertension (PAH, World Health Organization [WHO] Group I) to delay disease progression and reduce the risk of hospitalization for PAH. Effectiveness of Uptravi tablets was established in a long-term study in PAH patients with WHO Functional Class II-III symptoms. Patients had idiopathic and heritable PAH, PAH associated with connective tissue disease, and PAH associated with congenital heart disease with repaired shunts.

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

Applicable Drug List:

Uptravi

Policy/Guideline:

Prescriber Specialties

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

Coverage Criteria

Pulmonary Arterial Hypertension (PAH)[1-6]

Authorization of 12 months may be granted for treatment of PAH 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 either of the following criteria:

  - Pretreatment right heart catheterization with all of the following results:
    - Mean pulmonary arterial pressure (mPAP) > 20 mmHg
    - Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
    - Pulmonary vascular resistance (PVR) > 2 Wood units. For pediatric members, pulmonary vascular resistance index (PVRI) > 3 Wood units x m2 is also acceptable.
  - For infants less than one year of age, PAH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed.
- Patient is unable to take the required number of formulary alternatives (2) for the given diagnosis due to a trial and inadequate treatment response or intolerance, or a contraindication.

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)[4]

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

Approval Duration and Quantity Restrictions:

Approval: 12 months

Quantity Level Limit:

- Uptravi (selexipag) 200 mcg tablets: 140 per 28 days
- Uptravi (selexipag) 400 mcg tablets: 60 per 30 days
- Uptravi (selexipag) 600 mcg tablets: 60 per 30 days
- Uptravi (selexipag) 800 mcg tablets: 60 per 30 days
- Uptravi (selexipag) 1000 mcg tablets: 60 per 30 days
- Uptravi (selexipag) 1200 mcg tablets: 60 per 30 days
- Uptravi (selexipag) 1400 mcg tablets: 60 per 30 days
- Uptravi (selexipag) 1600 mcg tablets: 60 per 30 days
- Uptravi 1800 mcg single-dose vials: 60 vials per 30 days
- Uptravi (selexipag) titration pack (200 mcg/800 mcg tablets): 1 pack (140- 200mcg tablets, 60- 800 mcg tablets) per 28 days

References:

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4. 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
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