

	
AETNA BETTER HEALTH® Coverage Policy/Guideline	
Name: Uptravi	Page: 1 of 4
Effective Date: 2/28/2025	Last Review Date: 1/2025
Applies to: <div> <input checked="" type="checkbox"/> Illinois <input type="checkbox"/> New Jersey <input checked="" type="checkbox"/> Pennsylvania Kids </div>	<div> <input type="checkbox"/> Florida <input checked="" type="checkbox"/> Maryland <input type="checkbox"/> Virginia </div> <div> <input checked="" type="checkbox"/> Florida Kids <input type="checkbox"/> Michigan <input type="checkbox"/> Kentucky PRMD </div>

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:

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 Indications¹

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 Specialty

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:



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- 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 (3) 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



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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



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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 (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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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
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