

# SPECIALTY GUIDELINE MANAGEMENT

## Uptravi (selexipag)

### 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

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, PAH associated with congenital heart disease with repaired shunts.

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:

- 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. Mean pulmonary arterial pressure (mPAP) > 20 mmHg
    - ii. Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
    - iii. Pulmonary vascular resistance (PVR) ≥ 3 Wood units in adult patients or pulmonary vascular resistance index (PVRI) ≥ 3 Wood units x m<sup>2</sup> 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.

#### 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****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
- 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

**VI. REFERENCES**

1. Upravi [package insert]. Titusville, NJ: Actelion Pharmaceuticals US, Inc.; July 2022.

<b>Reference number(s)</b>
1645-A

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3. 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.
4. McLaughlin V, et al. ACCF/AHA 2009 Expert Consensus Document on Pulmonary Hypertension. *J Am Coll Cardiol*. 2009;53:1573-1619.
5. 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.
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7. 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.
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.