AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name:	Uptravi	Page:	1 of 4
Effective Date:	12/21/2023	Last Review Date:	11/2023
Applies to:	☐Illinois ☒New Jersey ☐Pennsylvania Kids	☐Florida ☐Maryland ☐Virginia	☐Florida Kids ☐Michigan ☐Kentucky PRMD

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:

FDA-Approved Indication
Uptravi is indicated for the treatment of pulmonary arterial hypertension (PAH, 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.

Applicable Drug List:

Uptravi

Policy/Guideline:

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

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. mPAP > 20 mmHg
        ii. 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^2 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.

C. 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

Criteria for Continuation of Therapy

Authorization of 12 months may be granted for members with an indication listed in criteria for initial approval 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

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

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:

1. Uptravi [package insert]. Titusville, NJ: Actelion Pharmaceuticals US, Inc.; July 2022.
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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.
6. Galie N, McLaughlin VV, Rubin LJ, Simonneau G. An overview of the 6th World Symposium on Pulmonary Hypertension. *Eur Respir J.* 2019; 53: 1802148; DOI: 10.1183/13993003.02148-2018. Published 24 January 2019.
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.