

**SPECIALTY GUIDELINE MANAGEMENT  
Pulmonary Arterial Hypertension (PAH)**

- Sildenafil tablet (generic Revatio)**
- Tadalafil tablet (generic Adcirca, Alyq)**
- Ambrisentan tablet (generic Letairis)**
- Bosentan tablet (generic Tracleer)**
- Opsumit tablet (macitentan)**
- Tracleer (bosentan tablet for oral susp)**
- Epoprostenol injection (generic Flolan and Veletri)**
- Treprostinil injection IV or SC (generic Remodulin)**
- Ventavis (iloprost) inhalation solution**
- Veletri (epoprostenol) injection**
- Orenitram (treprostinil) extended-release tablets and titration packs**
- Uptravi (selexipag) tablet**
- Winrevair (sotatercept-csrk) SC injection**

**POLICY**

**I. CRITERIA FOR INITIAL APPROVAL**

Authorization of 6 months may be granted for treatment of PAH when all the following criteria are met:

- A. Prescriber is either one of the following:
  - a. Prescriber is associated with an accredited Center of Comprehensive Care by the Pulmonary Hypertension Association for adult members
  - b. Prescriber is a cardiologist, pulmonologist or physician specializing in PAH for pediatric members
- B. Member has PAH defined as WHO Group 1 class of pulmonary hypertension (refer to Appendix)
- C. PAH was confirmed by either criterion (1) or criterion (2) below:
  - 1. Right heart catheterization with all of the following results:
    - i. Mean pulmonary arterial pressure (mPAP) > 20mmHg
    - ii. Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
    - iii. Pulmonary vascular resistance (PVR)
      - a. For Winrevair requests, pulmonary vascular resistance (PVR) ≥ 5 Wood units while stable on at least 2 PAH medications
      - b. For all other requests, pretreatment pulmonary vascular resistance (PVR) > 3 Wood units
  - 2. For infants less than one year of age with any of the following conditions, PAH was confirmed by Doppler echocardiogram if right heart catheterization cannot be performed.
- D. With the exception of members presenting in NYHA functional class IV, documentation that the member has undergone acute vasoreactivity testing and whether or not the results were positive; for those members who demonstrated a positive response to the acute vasoreactivity testing [defined as a fall in mean pulmonary arterial pressure (mPAP) of at least 10 mmHg to less than 40 mmHg with an

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increased or unchanged cardiac output], documentation must be submitted that PAH has progressed despite maximal medical treatment with a calcium channel blocker

- E. Dose and dosing regimen prescribed as well as medication therapy regimen is within FDA-approved guidelines and clinical guidelines
- F. If requesting Tracleer tablets for oral suspension, medical rationale must be submitted for reason why member is unable to trial bosentan tablets.
- G. If requesting Winrevair, the member must also meet all of the following criteria:
  - i. Member has WHO Functional Class II or III symptoms
  - ii. Member has a platelet count  $\geq 50,000/\text{mm}^3$
  - iii. Member is currently receiving at least two other PAH therapies from two different pharmacological classes for  $\geq 60$  days:
    - a. Phosphodiesterase type 5 inhibitors (PDE5i) [e.g., sildenafil, tadalafil]
    - b. Endothelin receptor antagonists (ERAs) [e.g., ambrisentan, bosentan, Opsumit]
    - c. Soluble guanylate cyclase stimulator (sGCs) [e.g., Adempas]
    - d. Prostacyclins (e.g., Flolan, Orenitram, Remodulin, Tyvaso, Veletri, Ventavis, Upravi)
  - iv. Winrevair will be used as add-on therapy and current PAH background therapies will be continued (e.g. ERA, PDE5i) unless not tolerated after addition of Winrevair

## II. CONTINUATION OF THERAPY

Authorization for 6 months may be granted for members with PAH who have experienced improved outcomes (e.g., sustained increase in six-minute walk distance from baseline, improvement in PAH symptoms/functional class, has not experienced clinical deterioration) with requested therapy.

## III. QUANTITY LIMITS

- A. Sildenafil 20mg tablet – 12 tablets per day
- B. Tadalafil 20mg tablet – 2 tablets per day
- C. Ambrisentan 5mg and 10mg tablets – 1 tablet per day
- D. Bosentan 62.5mg tablet and 125mg tablet – 2 tablets per day
- E. Opsumit 10mg tablet – 1 tablet per day
- F. Tracleer 32mg tablet – 4 tablets per day
- G. Ventavis 10mcg/ml and 20mcg/ml – 9 ampules per day
- H. Upravi – 2 tablets per day for all strengths except for titration pack (1 pack of 200 tablets per 30 days)
- I. Winrevair – 1 kit per 21 days (daily dose of 0.05)

## IV. 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

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