

## SPECIALTY GUIDELINE MANAGEMENT

### Ventavis (iloprost inhalation solution)

#### 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

##### **Pulmonary Arterial Hypertension**

Ventavis is indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to improve a composite endpoint consisting of exercise tolerance, symptoms (NYHA Class), and lack of deterioration.

All other indications are considered experimental/investigational and are not a covered benefit.

##### II. CRITERIA FOR INITIAL APPROVAL

Indefinite authorization 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:
    - mPAP  $\geq$  25 mmHg
    - PCWP  $\leq$  15 mmHg
    - 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:
    - Post cardiac surgery
    - Chronic heart disease
    - Chronic lung disease associated with prematurity
    - Congenital diaphragmatic hernia

##### III. CONTINUATION OF THERAPY

Indefinite authorization may be granted for members with PAH who are currently receiving Ventavis therapy through a paid pharmacy or medical benefit.

##### IV. APPENDIX

##### **WHO Classification of Pulmonary Hypertension**

##### WHO Group 1. Pulmonary Arterial Hypertension (PAH)

##### 1.1 Idiopathic (IPAH)

##### 1.2 Heritable PAH

- 1.2.1 Germline mutations in the bone morphogenetic protein receptor type 2 (BMP2)
- 1.2.2 Activin receptor-like kinase type 1 (ALK1), endoglin (with or without hereditary hemorrhagic telangiectasia), Smad 9, caveolin-1 (CAV1), potassium channel super family K member-3 (KCNK3)
- 1.2.3 Unknown
- 1.3 Drug- and toxin-induced
- 1.4. 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'. Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)
- 1". Persistent pulmonary hypertension of the newborn (PPHN)

#### WHO Group 2. Pulmonary Hypertension Owing to Left Heart Disease

- 2.1 Systolic dysfunction
- 2.2 Diastolic dysfunction
- 2.3 Valvular disease
- 2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies

#### WHO Group 3. Pulmonary Hypertension Owing to Lung Disease and/or Hypoxia

- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental abnormalities

#### WHO Group 4. Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

#### WHO Group 5. Pulmonary Hypertension with Unclear Multifactorial Mechanisms

- 5.1 Hematologic disorders: Chronic hemolytic anemia, myeloproliferative disorders, splenectomy
- 5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis: lymphangioleiomyomatosis, neurofibromatosis, vasculitis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis, segmental PH

## **V. REFERENCES**

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4. Badesch DB, Champion HC, Gomez-Sanchez MA, et al. Diagnosis and assessment of pulmonary arterial hypertension. *J Am Coll Cardiol.* 2009;54:S55-S66.
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