

## SPECIALTY GUIDELINE MANAGEMENT

### sildenafil tablets (generic) Revatio (sildenafil tablets and oral suspension)

#### 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.

###### A. FDA-Approved Indication

Sildenafil/Revatio is indicated for the treatment of pulmonary arterial hypertension (WHO Group I) in adults to improve exercise ability and delay clinical worsening.

###### B. Compendial Use

Raynaud's phenomenon secondary to systemic sclerosis (*Tablets only*)

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

##### II. CRITERIA FOR INITIAL APPROVAL

###### A. **Pulmonary Arterial Hypertension**

Authorization of 12 months may be granted for treatment of PAH when ALL of the following criteria are met:

1. Member has PAH defined as WHO Group 1 class of pulmonary hypertension (refer to Appendix).
2. PAH was confirmed by either criterion (1) or criterion (2) below:
  - i. Pretreatment right heart catheterization with all of the following results:
    - mPAP  $\geq$  25 mmHg
    - PCWP  $\leq$  15 mmHg
    - PVR > 3 Wood units
  - ii. 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

###### B. **Secondary Raynaud's Phenomenon**

Authorization or 12 months may be granted for treatment Raynaud's phenomenon secondary to systemic sclerosis when the patient has had an inadequate response to one of the following medications:

- Calcium channel blockers
- Angiotensin receptor blockers
- Selective serotonin reuptake inhibitors
- Alpha blockers

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- Angiotensin converting enzyme inhibitors
- Topical nitrates

### III. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for members with PAH or secondary Raynaud's phenomenon who are currently receiving sildenafil/Revatio 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 (BMPR2)
  - 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

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