Effective Date: 9/2017
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SPECIALTY GUIDELINE MANAGEMENT
Pulmonary Arterial Hypertension (PAH)

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

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. Pretreatment right heart catheterization with all of the following results:
      i. mPAP ≥ 20mmHg
      ii. PCWP ≤ 15 mmHg
      iii. 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 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

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. Opsumit 10mg tablet – 1 tablet per day
E. Tracleer 32mg tablet – 4 tablets per day; 62.5mg tablet and 125mg tablet – 2 tablets per day
F. Ventavis 10mcg/ml and 20mcg/ml – 9 ampules per day
G. Uptravi – 2 tablets per day for all strengths except for titration pack (1 pack of 200 tablets per 30 days)

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