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SPECIALTY GUIDELINE MANAGEMENT

Adempas (riociguat)

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 Indications

A. Pulmonary Arterial Hypertension (PAH)
Adempas is indicated for the treatment of adults with pulmonary arterial hypertension (PAH), (World Health Organization [WHO] Group 1), to improve exercise capacity, WHO functional class and to delay clinical worsening.

B. Chronic Thromboembolic Pulmonary Hypertension (CTEPH)
Adempas is indicated for the treatment of adults with persistent/recurrent chronic thromboembolic pulmonary hypertension (CTEPH), (WHO Group 4) after surgical treatment, or inoperable CTEPH, to improve exercise capacity and WHO functional class.

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

II. CRITERIA FOR INITIAL APPROVAL

Authorization may be granted for treatment of PAH or CTEPH based on the criteria below.

A. Pulmonary Arterial Hypertension (PAH)

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

1. 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 cardiologist, pulmonologist or physician specializing in PAH for pediatric members
2. Documentation that the member has PAH defined as WHO Group 1 class of pulmonary hypertension (Refer to Appendix)
3. PAH was confirmed by right heart catheterization with documentation of all of the following pretreatment results:
    a. Mean pulmonary arterial pressure (mPAP) > 20 mmHg
    b. Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
    c. Pulmonary vascular resistance (PVR) > 2 Wood units
4. 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

5. Dose and dosing regimen prescribed as well as medication therapy regimen is within FDA-approved guidelines and clinical guidelines

B. Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

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

1. Documentation that the member has CTEPH defined as WHO Group 4 class of pulmonary hypertension (Refer to Appendix)
2. Documentation that the member meets either criterion (i) or criterion (ii) below:
   i. Recurrent or persistent CTEPH after pulmonary endarterectomy (PEA)
   ii. Inoperable CTEPH with diagnosis confirmed by BOTH of the following (a. and b.):
      a. Computed tomography (CT)/magnetic resonance imaging (MRI) angiography or pulmonary angiography
      b. Pretreatment right heart catheterization with all of the following results:
         1. mPAP > 20 mmHg
         2. PCWP ≤ 15 mmHg
         3. PVR > 2 Wood units
3. Dose and dosing regimen prescribed as well as medication therapy regimen is within FDA-approved guidelines and clinical guidelines

III. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for members with documentation of an indication listed in Section II who are currently receiving the requested medication through a paid pharmacy benefit, and who are experiencing documented benefit from therapy as evidenced by disease stability or disease improvement.

IV. QUANTITY LIMIT

Adempas 0.5mg, 1mg, 1.5mg, 2mg, & 2.5mg: 3 tablets per day.

V. 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
- 4.2.7 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

VI. REFERENCES

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