# 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 not medically necessary.

#### II. CRITERIA FOR INITIAL APPROVAL

## A. Pulmonary Arterial Hypertension (PAH)

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 right heart catheterization with all of the following pretreatment results:
  - i. mPAP > 20 mmHg
  - ii. PCWP ≤ 15 mmHg
  - iii. PVR ≥ 3 Wood units

# B. Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

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

- 1. Member has CTEPH defined as WHO Group 4 class of pulmonary hypertension (Refer to Appendix)
- 2. Member meets either criterion (i) or criterion (ii) below:
  - i. Recurrent or persistent CTEPH after pulmonary endarterectomy (PEA)
  - 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 ≥ 3 Wood units

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#### **III. CONTINUATION OF THERAPY**

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

## 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

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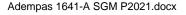
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- 5.1 Hematologic disorders: Chronic hemolytic anemia, myeloproliferative disorders
- 5.2 Systemic and metabolic disorders: Pulmonary Langerhans cell histiocytosis, Gaucher disease, glycogen storage disease, neurofibromatosis, sarcoidosis
- 5.3 Others: chronic renal failure with or without hemodialysis, fibrosing mediastinitis
- 5.4 Complex congenital heart disease

#### V. REFERENCES

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