Adempas (riociguat)

Override(s)	Approval Duration
Prior Authorization Quantity Limit	1 year

Medications	Quantity Limit
Adempas (riociguat) tablets	May be subject to quantity limit

APPROVAL CRITERIA

Initial requests for Adempas (riociguat) may be approved if the following criteria are met:

I. Individual has pulmonary arterial hypertension (PAH) [World Health Organization (WHO) Group 1][2]; **AND**
II. Individual has the diagnosis of PAH confirmed by a right-heart catheterization showing all of the following (Hoeper, 2013; Ivy, 2013; Abman, 2015):
    A. Mean pulmonary artery pressure (mPAP) greater than or equal to 25 mm Hg at rest;
    B. Pulmonary capillary wedge pressure (PCWP), mean pulmonary artery wedge pressure (PAWP), left atrial pressure, or left ventricular end-diastolic pressure (LVEDP) less than or equal to 15 mm Hg;
    C. Pulmonary vascular resistance (PVR) greater than 3 Wood units; **AND**
III. Individual has WHO functional class II-IV[3] symptoms;

OR

IV. Individual has chronic thromboembolic pulmonary hypertension (CTEPH) (WHO Group 4)[2]; **AND**
V. Individual has the diagnosis of CTEPH confirmed by a right-heart catheterization showing a mPAP greater than 25 mm Hg caused by thromboemboli in the pulmonary arterial system (ACCF/AHA 2009); **AND**
VI. Individual has WHO functional class II-IV[3] symptoms; **AND**
VII. Individual is using for one of the following:
    A. Persistent or recurrent pulmonary hypertension after at least 180 days following surgical treatment with pulmonary endarterectomy; **OR**
    B. Inoperable (via pulmonary endarterectomy) CTEPH.

Continuation requests for Adempas (riociguat) may be approved if the following criterion is met:

I. There is confirmation of clinically significant improvement or stabilization in clinical signs and symptoms of disease (including but not limited to improvement in walk distance, dyspnea and/or functional class).

Adempas (riociguat) may **not** be approved for the following:

I. Individual has a diagnosis of severe hepatic impairment (Child-Pugh Class C); **OR**
II. Individual is on dialysis or has a creatinine clearance less than 15 mL/min; **OR**
III. Individual has a diagnosis of pulmonary veno-occlusive disease (PVOD); **OR**
IV. Individual has a diagnosis of pulmonary hypertension associated with idiopathic interstitial pneumonias (PH-IIP); **OR**
V. Use in combination with phosphodiesterase (PDE) inhibitors [such as, PDE-5 inhibitors (sildenafil, tadalafil, vardenafil) or nonspecific PDE inhibitors (dipyridamole, theophylline)]; **OR**
VI. Use in combination with nitrates (such as but not limited to, nitroglycerin) or nitric oxide donors (such as but not limited to, amyl nitrite) in any form; OR
VII. Use in combination with another soluble guanylate cyclase stimulator [including Verquvo (vericiguat)].

Notes:

1. Adempas (riociguat) has a black box warning for embryo-fetal toxicity. Pregnancy should be excluded prior to start of treatment, monthly during treatment, and 1 month after stopping treatment in females of reproductive potential. Adempas should not be administered to pregnant females due to the potential of causing fetal harm. Pregnancy should be prevented using acceptable means of contraception during treatment and for one month after therapy discontinued. Adempas will be available for all females, regardless of reproductive potential, through a restricted risk evaluation and mitigation strategy (REMS) program.

2. WHO Pulmonary Hypertension (PH) Group Classification (ACCF/AHA 2009):
    A. Group 1: Pulmonary arterial hypertension (PAH)
    B. Group 2: PH due to left heart disease
    C. Group 3: PH due to lung diseases and/or hypoxia
    D. Group 4: Chronic thromboembolic PH (CTEPH)
    E. Group 5: Miscellaneous/PH with unclear multifactorial mechanisms

3. WHO functional classification of PH (CHEST 2019):
    A. Class I: No limitation of physical activity. Ordinary physical activity does not cause undue dyspnea or fatigue, chest pain or near syncope.
    B. Class II: Slight limitation of physical activity. Comfortable at rest but ordinary physical activity causes undue dyspnea or fatigue, chest pain or near syncope.
    C. Class III: Marked limitation of physical activity. Comfortable at rest but less than ordinary activity causes undue dyspnea or fatigue, chest pain or near syncope.
    D. Class IV: Inability to carry out any physical activity without symptoms. Dyspnea and/or fatigue may be present at rest and discomfort is increased by any physical activity.

Key References:

1. Abman SH, Hansmann G, Archer SL, et al. Pediatric pulmonary hypertension: guidelines from the American Heart Association and American Thoracic Society. *Circulation*. 2015; 132(21):2037-2099.
2. Badesch BD, Abman SH, Simonneau G, et al. Medical therapy for pulmonary arterial hypertension: updated ACCP evidence-based clinical practice guidelines. *Chest*. 2007; 131(6):1917-1928.
3. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: January 22, 2022.
4. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
5. Hoeper MM, Bogaard HJ, Condliffe R, et al. Definitions and Diagnosis of Pulmonary Hypertension. *J Am Coll Cardiol*. 2013; 62(suppl 25):D42- D50. Available at: http://www.onlinejacc.org/content/62/25_Supplement/D42. Accessed: January 23, 2022.
6. Ivy DD, Abman SH, Barst RJ, et al. Pediatric Pulmonary Hypertension. *J Am Coll Cardiol*. 2013; 62(suppl 25):D117-D126. Available from: http://www.onlinejacc.org/content/62/25_Supplement/D117. Accessed: January 23, 2022.
7. Klinger JR, Elliott CG, Levine DJ, et. al. Therapy for Pulmonary Arterial Hypertension in Adults: Update of the CHEST Guideline and Expert Panel Report. *CHEST*. 2019; 155(3): 565-586.
8. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2022; Updated periodically.
9. McLaughlin VV, Archer SL, Badesch DB, et al. ACCF/AHA 2009 expert consensus document on pulmonary hypertension. A report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association. *J Am Coll Cardiol*. 2009; 53:1573-1619. Available at: http://circ.ahajournals.org/content/119/16/2250.full.pdf+html. Accessed: January 23, 2022.
10. Simonneau G, Montani D, Celermajer DS, et al. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. *Eur Respir J*. 2019; 53(1).

Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.