Market Applicability													
Market	**DC**	**FL & FHK**	**FL MMA**	**FL LTC**	**GA**	**KY**	**MD**	**NJ**	**NV**	**NY**	**TN**	**TX**	**WA**
Applicable	X	X	NA	NA	X	X	X	X	X	X	NA	NA	NA

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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

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.

Market Applicability													
Market	**DC**	**FL & FHK**	**FL MMA**	**FL LTC**	**GA**	**KY**	**MD**	**NJ**	**NV**	**NY**	**TN**	**TX**	**WA**
Applicable	X	X	NA	NA	X	X	X	X	X	X	NA	NA	NA

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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.

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. As a component of the Adempas REMS, prescribers, individuals, and pharmacies must enroll in the program.
2. WHO Pulmonary Hypertension (PH) Group Classification (ACCF/AHA 2009, Simonneau et al. 2013):
    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 2014):
    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.

Market Applicability													
Market	**DC**	**FL & FHK**	**FL MMA**	**FL LTC**	**GA**	**KY**	**MD**	**NJ**	**NV**	**NY**	**TN**	**TX**	**WA**
Applicable	X	X	NA	NA	X	X	X	X	X	X	NA	NA	NA

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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.

State Specific Mandates		
State name N/A	Date effective N/A	Mandate details (including specific bill if applicable) N/A

Key References:

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3. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2019. URL: http://www.clinicalpharmacology.com. Updated periodically.
4. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: January 11, 2019.
5. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
6. 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 11, 2019.
7. 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 11, 2019.
8. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2019; Updated periodically.
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Market Applicability													
Market	**DC**	**FL & FHK**	**FL MMA**	**FL LTC**	**GA**	**KY**	**MD**	**NJ**	**NV**	**NY**	**TN**	**TX**	**WA**
Applicable	X	X	NA	NA	X	X	X	X	X	X	NA	NA	NA

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This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

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