Mass General Brigham
Health Plan

Adempas (riociguat)
Effective 09/01/2023

Plan	☐ MassHealth UPPL ☒Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☐ Pharmacy Benefit ☒ Medical Benefit		
Specialty Limitations	N/A		
Contact Information	**Medical and Specialty Medications** All Plans | Phone: 877-519-1908 | Fax: 855-540-3693		
	Non-Specialty Medications All Plans | Phone: 800-711-4555 | Fax: 844-403-1029		
Exceptions	N/A		

Overview

FDA-Approved Indications

1. 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.
2. 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.

Coverage Guidelines

Authorization may be granted for members new to General Brigham Health Plan who are currently receiving treatment with Adempas excluding when the product is obtained as samples or via manufacturer's patient assistance programs.

OR

Pulmonary Arterial Hypertension (PAH)

Authorization 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)
   a. PAH was confirmed by right heart catheterization with all of the following pretreatment results:
   b. mPAP > 20 mmHg
   c. PCWP ≤ 15 mmHg
   d. PVR ≥ 3 Wood units

Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

Authorization 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 ONE of the following:
    a. Recurrent or persistent CTEPH after pulmonary endarterectomy (PEA)
    b. Inoperable CTEPH with diagnosis confirmed by BOTH of the following:
        i. Computed tomography (CT)/magnetic resonance imaging (MRI) angiography or pulmonary angiography
        ii. Pretreatment right heart catheterization with all of the following results:
            A. mPAP > 20 mmHg
            B. PCWP ≤ 15 mmHg
            C. PVR ≥ 3 Wood units

Continuation of Therapy

Authorization of 12 months may be granted for members with a covered indication 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.

Limitations

1. Initial approvals and reauthorizations will be granted for 12 months.
2. The following quantity limits apply:

Adempas	90 tablets per 30 days

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
- 2.1 PH due to heart failure with reduced LVEF
- 2.2 Valvular heart disease
- 2.3 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

References

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6. Barst RJ, Gibbs SR, Ghofrani HA, et al. Updated evidence-based treatment algorithm in pulmonary arterial hypertension. *J Am Coll Cardiol.* 2009;54:S78-S84.
7. Taichman DB, Ornelas J, Chung L, et al. Pharmacologic therapy for pulmonary arterial hypertension in adults. CHEST guideline and expert panel report. *Chest*. 2014;46(2):449-475.
8. Jaff MR, McMurty MS, Archer SL, et al. Management of massive and submassive pulmonary embolism, iliofemoral deep vein thrombosis, and chronic thromboembolic pulmonary hypertension: a scientific statement from the American Heart Association. *Circulation*. 2011;123(16):1788-1830.
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13. Simonneau G, Montani D, Celermajer DS, et al. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. *Eur Respir J* 2019;53:1801913; doi:10.1183/13993003.01913-2018.

Review History

07/12/2023 – Reviewed at July P&T; Switched from CVS Standard to custom criteria; Effective 9/1/23