VENTURA COUNTY
HEALTH CARE PLAN

PRIOR AUTHORIZATION POLICY

POLICY: Pulmonary Arterial Hypertension – Adempas Prior Authorization Policy
- Adempas® (riociguat tablets – Bayer)

REVIEW DATE: 09/23/2020

OVERVIEW

Adempas, a soluble guanylate cyclase (sGC) stimulator, is indicated for the treatment of adults with:[1]

- **Chronic thromboembolic pulmonary hypertension** (CTEPH) [World Health Organization {WHO} Group 4], persistent/recurrent, after surgical treatment, or inoperable CTEPH, to improve exercise capacity and WHO functional class.
- **Pulmonary Arterial Hypertension** (PAH) [WHO Group 1), to improve exercise capacity, WHO functional class, and to delay clinical worsening.

Disease Overview

PAH is a serious but rare condition impacting approximately fewer than 20,000 patients in the US. It is classified within Group 1 pulmonary hypertension among the five different groups that are recognized. In this progressive disorder the small arteries in the lungs become narrowed, restricted, or blocked causing the heart to work harder to pump blood, leading to activity impairment.[2,3] In time, right-sided heart failure and/or death may occur. Common PAH symptoms include shortness of breath, fatigue, chest pain, dizziness and fainting, along with impairment in activity tolerance. It is more prevalent in women. Patients of all ages may develop the disease; however, the mean age of diagnosis typically happens between 36 to 50 years. Children may also have PAH. The condition may occur due to various underlying medical conditions or as a disease that uniquely impacts the pulmonary circulation; both genetic and environmental factors may be involved. PAH is defined as a mean pulmonary artery pressure (mPAP) $\geq$ 25 mmHg with a pulmonary capillary wedge pressure (PCWP) $\leq$ 15 mmHg measured by cardiac catheterization. The prognosis in PAH has been described as poor, with the median survival being approximately 3 years. However, primarily due to advances in pharmacological therapies, the long-term prognosis has improved. Lung transplantation may be recommended if pharmacological or medical therapies fail, based upon patient status. The WHO categorizes PAH into stages, which is also referred to as the functional class (Class I to IV) and is an adaptation of the New York Heart Association (NYHA) system to evaluate activity tolerance.

CTEPH is a persistent obstruction of pulmonary arteries and is often a complication of pulmonary embolism.[4,5] It is classified within Group 4 pulmonary hypertension. Symptoms include progressive dyspnea on exertion, as well as fatigue, syncope, hemoptysis, and signs of right heart failure. Pulmonary endarterectomy is the treatment of choice for most patients with CTEPH. However, around 40% of patients are deemed inoperable for various reasons. Medication therapy, including Adempas, may also be recommended. Anticoagulant therapy is also given.

Guidelines

In 2019, an updated CHEST guideline and Expert Panel Report regarding therapy for pulmonary arterial hypertension in adults was released.[5] Evidence for use of the many medications available is also detailed. Adempas is cited as a vital therapy in the management of PAH with several benefits in a variety of clinical scenarios.

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Adempas. All approvals are provided for the duration noted below. Because of the specialized skills required for evaluation and diagnosis of patients treated with Adempas as well as the monitoring required for adverse events and long-term efficacy, approval requires these agents to be prescribed by or in consultation with a physician who specializes in the condition being treated.

Documentation: In the *Pulmonary Arterial Hypertension – Adempas Prior Authorization Policy*, documentation is required for initiation of therapy where noted in the criteria as **[documentation required]**. Documentation may include, but is not limited to, chart notes and catheterization laboratory reports. For a patient case in which the documentation requirement of the right heart catheterization upon prior authorization coverage review for a different medication indicated for WHO Group 1 PAH has been previously provided, the documentation requirement in this *Pulmonary Arterial Hypertension – Adempas Prior Authorization Policy* is considered to be met.

Automation: None.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Adempas is recommended in those who meet the following criteria:

FDA-Approved Indication

1. **Chronic Thromboembolic Pulmonary Hypertension (CTEPH).** Approve for 3 years if prescribed by, or in consultation with, a pulmonologist or a cardiologist.

2. **Pulmonary Arterial Hypertension (PAH) [World Health Organization {WHO} Group 1].** Approve for the duration noted if the patient meets ONE of the following (A or B):
   - **A)** Initial Therapy. Approve for 3 years if the patient meets all of the following criteria (i, ii, and iii):
     - **i.** Patient has a diagnosis of World Health Organization (WHO) Group 1 pulmonary arterial hypertension (PAH); AND
     - **ii.** Patient meets the following criteria (a and b):
       - **a)** Patient has had a right heart catheterization **[documentation required]** (see documentation section above); AND
       - **b)** Results of the right heart catheterization confirm the diagnosis of WHO Group 1 PAH; AND
     - **iii.** Medication is prescribed by, or in consultation with, a cardiologist or a pulmonologist.
   - **B)** Patient is Currently Receiving Adempas. Approve for 3 years if the patient meets all of the following criteria (i, ii, and iii):
     - **i.** Patient has a diagnosis of WHO Group 1 PAH; AND
     - **ii.** Patient meets the following criteria (a and b):
       - **a)** Patient has had a right heart catheterization; AND
       - **b)** Results of the right heart catheterization confirm the diagnosis of WHO Group 1 PAH; AND
     - **iii.** Medication is prescribed by, or in consultation with, a cardiologist or a pulmonologist.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Coverage of Adempas is not recommended in the following situations:

1. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

REFERENCES

1. Adempas® tablets [prescribing information]. Whippany, NJ: Bayer; January 2018.
2. McLaughlin VV, Archer SL, Badesch DB, et al; Writing committee members. 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: Developed in collaboration with the American College of Chest Physicians, American Thoracic Society, Inc., and the Pulmonary Hypertension Association. *Circulation*. 2009;119:2250-2294.
3. 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.
4. Kim NH, Delcroix M, Jais X, et al. Chronic thromboembolic pulmonary hypertension. *Eur Respir J*. 2019;53(1):1801915.
5. Hoeper MM, Madani MM, Nakanishi N, et al. Chronic thromboembolic pulmonary hypertension. *Lancet Respir Med*. 2014;2(7):573-582.