Clinical Policy: Pulmonary Hypertension Agents, Oral and Inhaled

Reference Number: PHW.PDL.171
Effective Date: 01/01/2020
Last Review Date: 11/2024

Policy/Criteria
Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

It is the policy of PA Health and Wellness® that Oral and Inhaled Pulmonary Arterial Hypertension (PAH) Agents are **medically necessary** when the following criteria are met:

I. Requirements for Prior Authorization of Pulmonary Hypertension Agents, Oral and Inhaled

A. Prescriptions That Require Prior Authorization

All prescriptions for Pulmonary Hypertension Agents, Oral and Inhaled must be prior authorized.

B. Review of Documentation for Medical Necessity

In evaluating a request for prior authorization of a prescription for a Pulmonary Hypertension Agent, Oral and Inhaled, the determination of whether the requested prescription is medically necessary will take into account whether the member:

1. **One** of the following:
   a. Is prescribed the Pulmonary Hypertension Agent, Oral and Inhaled for the treatment of a diagnosis that is indicated in the U.S. Food and Drug Administration (FDA)-approved package labeling OR a medically accepted indication, excluding use to treat sexual or erectile dysfunction
   b. For the treatment of pulmonary arterial hypertension (PAH), is prescribed a Pulmonary Hypertension Agent, Oral and Inhaled that is appropriate for the member's level of risk based on current risk calculator assessment (e.g., REVEAL 2.0) and current peer-reviewed medical literature;

   AND

2. Is prescribed a dose that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature; **AND**

3. **One** of the following:

a. If less than 18 years of age, is prescribed the Pulmonary Hypertension Agent, Oral and Inhaled by or in consultation with a pediatric pulmonologist, pediatric cardiologist, or heart and lung transplant specialist skilled in treating pulmonary hypertension

b. If greater than or equal to 18 years of age, **one** of the following:

   i. Is prescribed the Pulmonary Hypertension Agent, Oral and Inhaled by or in consultation with a practitioner at a Pulmonary Hypertension Association-accredited center
   ii. If unable to access a Pulmonary Hypertension Association-accredited center, is prescribed the Pulmonary Hypertension Agent, Oral and Inhaled by or in consultation with an appropriate specialist (i.e., pulmonologist, cardiologist, or rheumatologist) skilled in treating pulmonary hypertension;

AND

4. Does not have a contraindication to the prescribed medication; **AND**

5. For a diagnosis of PAH (WHO Group 1), **all** of the following:

   a. Has chart documentation of right heart catheterization indicating **all** of the following hemodynamic values:

      i. A mean pulmonary arterial pressure greater than 20 mmHg,
      ii. A pulmonary capillary wedge pressure, left atrial pressure, or left ventricular end-diastolic pressure less than or equal to 15 mm Hg,
      iii. A pulmonary vascular resistance greater than or equal to 3 Wood units,

   b. For a member with idiopathic PAH, **both** of the following:

      i. **One** of the following:
         a) Has a H_2FPEF score less than 2
         b) Has a left atrial volume index less than 35 mL/m^2
         c) Has a negative provocative test in a heart catheterization lab (fluid challenge with pulmonary capillary wedge pressure, left atrial pressure, or left ventricular end-diastolic pressure less than or equal to 17 mmHg)

      ii. **One** of the following:
         a) Has chart documentation of acute vasoreactivity testing
         b) Has a contraindication to vasoreactivity testing or is at increased risk of adverse events during acute vasoreactivity testing (e.g., high risk stratification based on current risk calculator assessment (e.g., REVEAL 2.0), low systemic blood pressure, low cardiac index, or pulmonary veno-occusive disease),

c. For a member with idiopathic PAH that demonstrates acute vasoreactivity,[1] has a documented history of therapeutic failure, contraindication, or intolerance of calcium channel blockers (i.e., amlodipine, nifedipine, or diltiazem);

AND

6. For a diagnosis of chronic thromboembolic pulmonary hypertension (CTEPH), has chart documentation of right heart catheterization indicating **both** of the following hemodynamic values:

   a. A mean pulmonary arterial pressure greater than 20 mmHg
   b. A pulmonary vascular resistance greater than or equal to 3 Wood units

AND

7. For a non-preferred Pulmonary Hypertension Agent, Oral and Inhaled, **one** of the following:

   a. Has a history of therapeutic failure, contraindication, or intolerance of the preferred Pulmonary Hypertension Agents, Oral and Inhaled approved or medically accepted for the member's diagnosis or indication
   b. Has a current history (within the past 90 days) of being prescribed the same non-preferred Pulmonary Hypertension Agent, Oral and Inhaled (does not apply to non-preferred brands when the therapeutically equivalent generic is preferred or to non-preferred generics when the therapeutically equivalent brand is preferred)

   See the Preferred Drug List (PDL) for the list of preferred Pulmonary Hypertension Agents, Oral and Inhaled at: https://papdl.com/preferred-drug-list

AND

8. If the prescription for a Pulmonary Hypertension Agent, Oral and Inhaled is for a quantity that exceeds the quantity limit, the determination of whether the prescription is medically necessary will also take into account the guidelines set forth in PA.CP.PMN.59 Quantity Limit Override.

   NOTE: If the member does not meet the clinical review guidelines above but, in the professional judgement of the physician reviewer, the services are medically necessary to meet the medical needs of the member, the request for prior authorization will be approved.

[1] A positive vasoreactivity test is defined by a decrease in the mean pulmonary artery pressure by at least 10 mmHg to reach an absolute value of 40 mmHg or less without a decrease in cardiac output.

FOR RENEWALS OF PRIOR AUTHORIZATION FOR Pulmonary Hypertension Agents, Oral and Inhaled: The determination of medical necessity of a request for renewal of a prior authorization for a Pulmonary Hypertension Agent, Oral and Inhaled that was previously approved will take into account whether the member:

1. Continues to benefit from the requested Pulmonary Hypertension Agent, Oral and Inhaled based on the prescriber's assessment; **AND**

2. Is prescribed a dose that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature; **AND**

3. **One** of the following:

    a. If less than 18 years of age, is prescribed the Pulmonary Hypertension Agent, Oral and Inhaled by or in consultation with a pediatric pulmonologist, pediatric cardiologist, or heart and lung transplant specialist

    b. If greater than or equal to 18 years of age, **one** of the following:

        i. Is prescribed the Pulmonary Hypertension Agent, Oral and Inhaled by or in consultation with a practitioner at a Pulmonary Hypertension Association-accredited center
        ii. If unable to access a Pulmonary Hypertension Association-accredited center, is prescribed the Pulmonary Hypertension Agent, Oral and Inhaled by or in consultation with an appropriate specialist (i.e., pulmonologist, cardiologist, or rheumatologist);

    AND

4. Does not have a contraindication to the prescribed medication; **AND**

5. For a non-preferred Pulmonary Hypertension Agent, Oral and Inhaled with a therapeutically equivalent brand or generic that is preferred on the PDL, has a history of therapeutic failure of or a contraindication or an intolerance to the preferred therapeutically equivalent brand or generic that would not be expected to occur with the requested medication.
    AND
6. If the prescription for a Pulmonary Hypertension Agent, Oral and Inhaled is for a quantity that exceeds the quantity limit, the determination of whether the prescription is medically necessary will also take into account the guidelines set forth in PA.CP.PMN.59 Quantity Limit Override

NOTE: If the member does not meet the clinical review guidelines above but, in the professional judgement of the physician reviewer, the services are medically

necessary to meet the medical needs of the member, the request for prior authorization will be approved.

C. Clinical Review Process

Prior authorization personnel will review the request for prior authorization and apply the clinical guidelines in Section B. above to assess the medical necessity of a prescription for a Pulmonary Hypertension Agent, Oral and Inhaled. If the guidelines in Section B. are met, the reviewer will prior authorize the prescription. If the guidelines are not met, the prior authorization request will be referred to a physician reviewer for a medical necessity determination. Such a request for prior authorization will be approved when, in the professional judgment of the physician reviewer, the services are medically necessary to meet the medical needs of the member.

D. Approval Duration:
- **New Request: 6 months**
- **Renewal Request: 12 months**

E. References

1. Abman SH. Pediatric Pulmonary Hypertension Network: Implications of the FDA warning against the use of sildenafil for the treatment of pediatric pulmonary hypertension: November 19, 2012.
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3. Adempas Package Insert. Whippany, NJ: Bayer HealthCare Pharmaceuticals Inc.; January 2018.
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13. Letairis Package Insert. Foster City, CA: Gilead Sciences, Inc.; August 2019.
14. Mullen MP, Kulik T. Pulmonary hypertension in children: Classification, evaluation, and diagnosis. Fulton DR, Mallory GB eds. Waltham, MA: UpToDate Inc. Updated March 06, 2019. Accessed July 29, 2019.
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16. Orenitram Package Insert. Research Triangle Park, NC: United Therapeutics Corp.; November 2020.
17. Pulmonary Hypertension Association Consensus Statement; Revatio (sildenafil) for Pediatric Use: September 2012.
18. Revatio Package Insert. New York, NY: Pfizer Labs; February 2018.
19. Rubin LJ, Hopkins W. Clinical features and diagnosis of pulmonary hypertension of unclear etiology in adults. Mandel J ed. Waltham, MA: UpToDate Inc. Updated May 17, 2019. Accessed July 29, 2019.
20. Simonneau G, Montani D, Celermajer DS, et al. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. *Eur Respir J*. 2019;53(1):1801913. Published 2019 Jan 24. [DOI:10.1183/13993003.01913-2018].
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24. Uptravi Package Insert. South San Francisco, CA: Actelion Pharmaceuticals US, Inc.; January 2021.
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Reviews, Revisions, and Approvals	Date
Policy created	01/01/2020
Q3 2020 annual review: no changes.	07/2020
Q1 2021 annual review: no changes.	01/2021
Q1 2022: revised according to DHS revisions 01/03/2022	10/2021
Q1 2023 annual review: no changes.	11/2022
Q1 2024 annual review: no changes.	11/2023
Q1 2025: policy revised according to DHS revisions effective 01/06/2025.	11/2024