RX.PA.029.MPC Pulmonary Arterial Hypertension (PAH) Products

The purpose of this policy is to define the prior authorization process for pulmonary arterial hypertension products.

DEFINITIONS

Chronic thromboembolic pulmonary hypertension (CTEPH) – a mean pulmonary artery pressure greater than 25 mm Hg that persists 6 months after pulmonary embolism is diagnosed.

Pulmonary arterial hypertension (PAH) – PAH is a syndrome resulting from restricted flow through the pulmonary arterial circulation resulting in increased pulmonary vascular resistance and subsequent right heart failure. The diagnosis of PAH requires confirmation with a complete right heart catheterization. The hemodynamic definition of PAH includes a mean pulmonary artery pressure (mPAP) greater than 25 mm Hg in the setting of a normal pulmonary capillary wedge pressure (PCWP) of 15 mm Hg or less with a pulmonary vascular resistance (PVR) greater than 3 Wood units.

Pulmonary endarterectomy (PEA) – surgical procedure to remove obstructive hardened, thromboembolic materials from the pulmonary arteries to improve hemodynamics in patients CTEPH.

Pulmonary Hypertension Specialist – a cardiologist or pulmonologist who is a member of the Pulmonary Hypertension Association

Pulmonary Vascular Resistance (PVR) – PVR (wood units) = (mPAP – PCWP)/ Cardiac Output. PVR (wood units) = PVR (dynes-sec-cm-5)/80

WHO Etiologic Classification of Pulmonary Hypertension

Group 1	Pulmonary arterial hypertension
Group 2	Pulmonary hypertension with left heart disease
Group 3	Pulmonary hypertension associated with lung disease and/or hypoxemia
Group 4	Pulmonary hypertension due to chronic thrombotic and/or embolic disease
Group 5	Miscellaneous

WHO Functional Classification of Pulmonary Hypertension

Class I	Members with no symptoms and for whom ordinary physical activity does not cause dyspnea or fatigue, chest pain or near syncope.

Class II	Members who are comfortable at rest but who have symptoms** with ordinary physical activity.
Class III	Members who are comfortable at rest but have symptoms** with less-than-ordinary effort.
Class IV	Members who have symptoms** at rest

**Key symptoms of PH include dyspnea or fatigue, chest pain, or near syncope (fainting)

PREFERRED – PA REQUIRED	NON-PREFERRED – PA REQUIRED
Phosphodiesterase-5 (PDE-5) Inhibitors	
Adcirca (tadalafil) tablets [Functional class II, III, or IV]	Revatio (sildenafil)- tablets, oral susp [Functional class II, III, or IV]
Sildenafil (generic for Revatio) tablets [Functional class II, III, or IV]	
Prostacyclin Analogs	
Epoprostenol (generic for Flolan) [Functional class III or IV]	Orenitram (treprostinil) [Functional class II or III]
Remodulin (treprostinil) [Functional class II, III, or IV]	Uptravi (selexipag) [Functional class II or III]
Tyvaso (treprostinil) [Functional class III]	Flolan (epoprostenol) [Functional class III or IV]
Epoprostenol (generic for Veletri) [Functional class III or IV]	Veletri (epoprostenol) [Functional class III or IV]
Ventavis (iloprost) inh soln [Functional class III or IV]	
Endothelial-Receptor Antagonists	
Letairis (ambrisentan) tablets [Functional class II, III, or IV]	
Opsumit (macitentan) tablets [Functional class II or III]	
Tracleer (bosentan) tablets [Functional class II, III, or IV]	
Soluble Guanylate Cyclase Stimulators	
Adempas (riociguat) tablets [Functional class II or III]	

PROCEDURE

A. Initial Authorization Criteria:

Must meet all of the criteria listed under the respective diagnosis:

1. Pulmonary Arterial Hypertension (PAH):

- Patient must be age 18 years or older
- Must have a diagnosis of PAH classified as WHO Group 1
- Must be prescribed by or in consultation with a cardiologist of pulmonologist experienced in the diagnosis and treatment of PAH
- Documentation of baseline 6-minute walk test
- Documentation of right catheterization with mean pulmonary artery pressure (mPAP) > 25 mmHg

- Must not be used concomitantly with organic nitrates (e.g., isosorbide mononitrate, isosorbide dinitrate, nitroglycerin)
- Must have appropriate New York Heart Association (NYHA) or WHO functional class symptoms for the requested drug listed in the table above
- For combination therapy (2+ PAH agents)
    - Remodulin (treprostinil) and Flolan (epoprostenol) combination is allowed in patients who require transition from Flolan. Combination therapy for the down-titration of Flolan can be up to 14 days.
    - Adempas must NOT be used in combination with PDE-5 inhibitors [e.g., Adcirca (tadalafil), Revatio (sildenafil), sildenafil (generic for Revatio)]
    - Must have documentation of an inadequate clinical response to monotherapy, defined as:
        - For WHO Functional Assessment Classification II or III: Resulting clinical status defined as stable and not satisfactory, or unstable and deteriorating
        - For WHO Functional Assessment Classification IV: No rapid improvement to WHO Functional Assessment Classification III or better
        - Clinical status defined as stable and not satisfactory
- For Tracleer in members less than 18 years of:
    - Must be age 3 years or older
    - Must have idiopathic or congenital PAH WHO Group 1
    - Must provide baseline pulmonary vascular resistance (PVR)
- For non-preferred agents:
    - Must have a documented trial and failure, contraindication, or intolerance to at least TWO of the preferred agents within the same class
- For Letairis, Opsumit, Adempas, and Tracleer verification prescriber and patient are enrolled in associated REMS Program
- If patient is a woman of childbearing potential, documented contraceptive plan and negative pregnancy test

2. Chronic Thromboembolic Pulmonary Hypertension (CTEPH): Adempas only

- Requested drug must be a soluble guanylate cyclase stimulator (Adempas)
- Must be age 18 years or older
- Must be prescribed by or in consultation with a cardiologist of pulmonologist experienced in the diagnosis and treatment of PAH
- Must have a diagnosis of CTEPH classified as WHO Group 4
- Must have New York Heart Association (NYHA) or WHO functional class II or III symptoms

- Documentation of baseline 6-minute walk test
- Documentation of Pulmonary vascular resistance (PVR) > 3 Wood units following pulmonary thromboendarterectomy
- Documentation of ALL of the following:
  - Patient inoperable for pulmonary endarterectomy
  - PVR >3 Wood units
  - Mean pulmonary artery pressure (mPAP) >25 mmHg
- Must not be used concomitantly with organic nitrates or phosphodiesterase-5 inhibitors [Revatio (sildenafil) or Adcirca (tadalafil)]
- Verification prescriber and patient are enrolled in Adempas REMS Program
- If patient is a woman of childbearing potential, documented contraceptive plan and negative pregnancy test

Note: Documentation MUST include either paid claims OR specific dates of use for medication trials AND/OR chart documentation from the provider noting a contraindication, intolerance, or failure to all pre-requisite medications

B. Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc) listed in the FDA approved labeling.

C. Medications in this policy used for the treatment of pulmonary arterial hypertension will be considered investigational or experimental for any other use and will not be covered.

D. <u>Reauthorization Criteria:</u>

PREFERRED – PA REQUIRED	NON-PREFERRED – PA REQUIRED
Phosphodiesterase-5 (PDE-5) Inhibitors	
Adcirca (tadalafil) tablets [Functional class II, III, or IV]	Revatio (sildenafil)- tablets, oral susp [Functional class II, III, or IV]
Sildenafil (generic for Revatio) tablets [Functional class II, III, or IV]	
Prostacyclin Analogs	
Epoprostenol (generic for Flolan) [Functional class III or IV]	Orenitram (treprostinil) [Functional class II or III]
Remodulin (treprostinil) [Functional class II, III, or IV]	Uptravi (selexipag) [Functional class II or III]
Tyvaso (treprostinil) [Functional class III]	Flolan (epoprostenol) [Functional class III or IV]
Epoprostenol (generic for Veletri) [Functional class III or IV]	Veletri (epoprostenol) [Functional class III or IV]
Ventavis (iloprost) inh soln [Functional class III or IV]	
Endothelial-Receptor Antagonists	
Letairis (ambrisentan) tablets [Functional class II, III, or IV]	

PREFERRED – PA REQUIRED	NON-PREFERRED – PA REQUIRED
Opsumit (macitentan) tablets [Functional class II or III]	
Tracleer (bosentan) tablets [Functional class II, III, or IV]	
Soluble Guanylate Cyclase Stimulators	
Adempas (riociguat) tablets [Functional class II or III]	

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of treatment. Authorization may be extended at 1-year intervals based upon:
MPC Renewal:

- Chart documentation from the prescriber that the member's condition has improved based upon the prescriber's assessment while on therapy.

- Must have chart documentation of clinical response, such as one of the following:
  - Reduction in mPAP of at least 10 mm-Hg from baseline
  - mPAP <40 mmHg without a decrease in cardiac output
  - Improvement on 6-minute walk test from baseline
- For Tracleer less than 18 years old:
  - Must provide documentation of improvement in PVR and/or exercise ability
- Must not have unacceptable toxicity from the drug which requires discontinuation of therapy, such as:
  - Revatio, Adcirca: pulmonary edema; hearing or visual impairment; symptomatic hypotension; epistaxis
  - Flolan, Veletri: anticoagulation abnormalities (bleeding); pulmonary edema
  - Remodulin: blood stream infections (BSIs); sepsis; infusion site reactions
  - Tyvaso: symptomatic hypotension; anticoagulation abnormalities (bleeding)
  - Ventavis: hypotension (systolic BP < 85 mm Hg); pulmonary edema
  - Tracleer, Letairis, Opsumit: hepatic impairment; fluid retention; pulmonary edema; decreased hemoglobin and hematocrit
  - Adempas: symptomatic hypotension, bleeding, and pulmonary edema.
  - Uptravi: decreased hemoglobin
- For Tracleer, Letairis, Opsumit, and Adempas, verification prescriber and patient are enrolled in associated REMS Program
- If patient is a woman of childbearing potential, documented contraceptive plan and negative pregnancy test

Renewal from Previous Insurer:

- Members who have received prior approval (from insurer other than MPC),

or have been receiving medication samples, should be considered under criterion A (Initial Authorization Criteria)

- Provider has documented clinical response of the member's condition which has stabilized or improved based upon the prescriber's assessment

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	Up to 1 year
Reauthorization	Same as initial

If the established criteria are not met, the request is referred to a Medical Director for review.

Code	Description
J1325	Injection, epoprostenol, 0.5 mg

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REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Update to include preferred vs non-preferred product list under the continuation of therapy pathway	*09/2026*
Annual Review	*02/2026*
Annual Review	*02/2025*
Annual Review *Change in Non-MPC renewal to renewal from previous insurer*	*02/2024*
Annual review	*02/2023*
Selected Revision *Addition of MPC vs Non-MPC Renewal*	*10/2022*
Annual review	*02/2022*
Addition of dosing requirements and off-label restrictions	*12/2021*
P&T Review	*11/2020*