Updated: 09/2025
Approved: 09/2025

Request for Prior Authorization for Pulmonary Arterial Hypertension (PAH) injectable agents
Website Form – www.wv.highmarkhealthoptions.com
Submit request via: Fax - 1-833-547-2030.

All requests for Pulmonary Arterial Hypertension (PAH) injectable agents require a Prior Authorization and will be screened for medical necessity and appropriateness using the criteria listed below.

Pulmonary Arterial Hypertension (PAH) injectable agents Prior Authorization Criteria:

- Treatment is prescribed by, or in consultation with, a cardiologist or pulmonologist
- The requested dose and frequency is in accordance with FDA-approved labeling, nationally recognized compendia, and/or evidence-based practice guidelines. If a requested dose is above these recommendations, medical rationale must be submitted. For infused products, must provide member's weight, dose, frequency and titration schedule.
- Request meets diagnostic and drug criteria outlined in sections A and B
- If member is new to the plan and requests a continuation of therapy, must provide chart documentation indicating member is currently on requested therapy

A. Diagnostic Criteria

Coverage may be provided with a diagnosis of Pulmonary Arterial Hypertension (PAH) WHO Group I and the following criteria is met:

- Member has a diagnosis of PAH WHO Group I (refer below to Appendix I) confirmed by chart documentation of right-heart catheterization (RHC) or echocardiography if the provider indicates RHC is not recommended. RHC documentation must contain the following hemodynamic values:
    - Mean pulmonary arterial pressure > 20 mmHg
    - Pulmonary capillary wedge pressure ≤ 15 mmHg
    - Pulmonary vascular resistance > 2 Wood units.
- Documentation of member's vasoreactivity test and one of the following, unless member has a contraindication to vasoreactivity testing (e.g. low systemic blood pressure, low cardiac index, or the presence of severe (functional class IV) symptoms):
    - Member had a positive response (pulmonary artery pressure decreases at least 10 mmHg and to a value less than or equal to 40 mmHg, with an increased or unchanged cardiac output, and a minimally reduced or unchanged systemic blood pressure) and had inadequate response, contraindication or intolerance to calcium channel blocker therapy with diltiazem or a dihydropyridine
    - Member did not have a positive response to the vasoreactivity test
- Member has functional class II, III or IV symptoms at baseline prior to initiating therapy with the requested drug(s) (refer below to Appendix II)
- One of the following:

- The requested drug will be used as monotherapy (except Winrevair (sotatercept-csrk) see drug specific criteria below.
- The requested drug will be used for add-on therapy to existing monotherapy or dual therapy in addition to **both** of the following:
  - The medications must be from different therapeutic classes
  - The member must have unresponsive or progressive disease despite established PAH-specific therapies

B. Drug Criteria

- **Prostanoids/prostacyclin therapies**
  - Infused agents: epoprostenol, Flolan, Veletri, Remodulin
    - For Flolan, Veletri, or Remodulin requests in members with functional class III or IV symptoms, must provide documentation of inadequate response, contraindication or intolerance to generic epoprostenol or clinical rationale for why generic epoprostenol cannot be used
    - Must meet **one** of the following criteria:
      - Documentation of WHO functional class IV symptoms or functional class III symptoms with **any** of the following:
        - Evidence of progression of their disease
        - Any marker of poor clinical prognosis defined as:
          - Clinical signs of right heart failure
          - Repeated episodes of syncope, even with little or regular physical activity
          - <165 meter 6-minute walking distance (6MWD)
          - Peak oxygen consumption (VO_2) <11ml/min/kg (<35% predicted)
          - Ventilatory equivalents of CO_2 (VE/VCO_2) slope ≥45
          - BNP >300 ng/l
          - NT-proBNP >1400ng/l
          - Right atrium area ≥26 cm^2
          - Presence of pericardial effusion
          - Right atrial pressure >14 mmHg
          - Cardiac index <2.0 l/min/m^2
          - Mixed venous oxygen saturation (SvO_2) <60%
      - Documentation of WHO functional class II symptoms and all of the following:
        - Request is for Remodulin
        - Inadequate response, contraindication or intolerance to combination therapy of generic ambrisentan and tadalafil
        - Inadequate response, contraindication or intolerance to one additional PAH medication therapy (could have been in combination or monotherapy)
- **Activin Signaling Inhibitor:**
  - Winrevair (sotatercept-crsk) for new starts only: the medication will be taken in combination with other PAH therapies and **ONE** of the following:
    - Member is currently receiving at least 2 other PAH therapies from different pharmacologic categories
    - The member is currently receiving at least one other PAH therapy and the prescriber attests the member is unable to tolerate combination therapy with a

phosphodiesterase type 5 inhibitors (PDE5i), endothelin receptor antagonists (ERAs), soluble guanylate cyclase stimulator (sGCs), or prostacyclin

Initial Duration of Approval: 6 months

Reauthorization criteria

- Must provide documentation that demonstrates member is tolerating and receiving clinical benefit from treatment.

Reauthorization Duration of Approval: 12 months

Coverage may be provided for any non-FDA labeled indication if it is determined that the use is a medically accepted indication supported by nationally recognized pharmacy compendia or peer-reviewed medical literature for treatment of the diagnosis(es) for which it is prescribed. These requests will be reviewed on a case by case basis to determine medical necessity.

When criteria are not met, the request will be forwarded to a Medical Director for review. The physician reviewer must override criteria when, in their professional judgment, the requested medication is medically necessary.

HIGHMARK HEALTH OPTIONS

Updated: 09/2025
Approved: 09/2025

Pulmonary Arterial Hypertension (PAH) injectable agents
PRIOR AUTHORIZATION FORM

Please complete and fax all requested information below including any progress notes, laboratory test results, or chart documentation as applicable to Highmark Health Options Pharmacy Services. **FAX: (833)-547-2030.**
If needed, you may call to speak to a Pharmacy Services Representative. **PHONE**: 1-844-325-6251 Mon – Fri 8 am to 7 pm

PROVIDER INFORMATION

Requesting Provider:	NPI:
Provider Specialty:	Office Contact:
Office Address:	Office Phone:
	Office Fax:

MEMBER INFORMATION

Member Name:	DOB:	
Member ID:	Member weight:	Height:

REQUESTED DRUG INFORMATION

Medication:	Strength:	
Directions:	Quantity:	Refills:
Is the member currently receiving requested medication? ☐ Yes ☐ No	Date Medication Initiated:	

Is this medication being used for a chronic or long-term condition for which the medication may be necessary for the life of the patient? ☐ Yes ☐ No

Billing Information

This medication will be billed: ☐ at a pharmacy **OR** ☐ medically, JCODE:_____

Place of Service: ☐ Hospital ☐ Provider's office ☐ Member's home ☐ Other

Place of Service Information

Name:	NPI:
Address:	Phone:

MEDICAL HISTORY (Complete for ALL requests)

Diagnosis:	ICD Code:

Has chart documentation of right-heart catheterization or echocardiography been provided confirming the diagnosis of pulmonary arterial hypertension? ☐ Yes ☐ No
Mean Pulmonary Arterial Pressure: _____ Pulmonary Capillary Wedge Pressure: _____
Pulmonary Vascular Resistance: _____ Date of Exam: _____

Please select the World Health Organization (WHO) Classification of Pulmonary Hypertension:
☐ Group 1 ☐ Group 2 ☐ Group 3 ☐ Group 4 ☐ Group 5

Please indicate WHO functional class symptoms: ☐ Class I ☐ Class II ☐ Class III ☐ Class IV

Is the member currently taking a nitrate product? ☐ Yes ☐ No

Will the requested medication be used as monotherapy or combination therapy? If combination therapy, please list other drug(s):	☐ Monotherapy ☐ Combination

Drug Name	Strength & Frequency	Rationale for additional therapy

Please check any boxes applicable to the member:
☐ Clinical signs of right heart failure ☐ Repeated episodes of syncope, even with little or regular physical activity
☐ Presence of pericardial effusion ☐ Peak oxygen consumption (VO2) <11ml/min/kg (<35% predicted)
☐ Ventilatory equivalents of CO2 (VE/VCO2) slope ≥ 45 ☐ <165 meter 6-minute walking distance (6MWD)
☐ Mixed venous oxygen saturation (SvO2)<60% ☐ Cardiac index <2.01/min/m2
☐ BNP >300 ng/l ☐ 300 ng/l NT-proBNP >1400ng/l ☐ Right atrium area ≥26 cm2 ☐ Right atrial pressure >14 mmHg

If the request is for Adempas (riociguat) for a diagnosis of Chronic Thromboembolic Pulmonary Hypertension (CTEPH) (WHO Group 4), please answer the following questions:
• Has the member previously failed surgical treatment (such as pulmonary endarterectomy)? ☐ Yes ☐ No
• Does the member have inoperable CTEPH? ☐ Yes ☐ No

• Has chart documentation of computed tomographic pulmonary angiography or ventilation-perfusion lung scan been provided confirming thromboembolic occlusion of the proximal or distal pulmonary vasculature? ☐Yes ☐No

PULMONARY ARTERIAL HYPERTENSION (PAH) AGENTS (ORAL AND INHALED) PRIOR AUTHORIZATION FORM (CONTINUED) – PAGE 2 OF 2

Please complete and fax all requested information below including any progress notes, laboratory test results, or chart documentation as applicable to Highmark Health Options Pharmacy Services. **FAX**: (855) 476-4158 If needed, you may call to speak to a Pharmacy Services Representative. **PHONE**: (844) 325-6251 Monday through Friday 8:00am to 7:00pm

MEMBER INFORMATION

Member Name:	DOB:
Member ID:	Member weight: _____pounds or _____kg

MEDICAL HISTORY (Complete for ALL requests)

If the request is for Adempas (riociguat) for a diagnosis of Chronic Thromboembolic Pulmonary Hypertension (CTEPH) (WHO Group 4), please answer the following questions:

• Has the member previously failed surgical treatment (such as pulmonary endarterectomy)? ☐Yes ☐No

• Does the member have inoperable CTEPH? ☐Yes ☐No

• Has chart documentation of computed tomographic pulmonary angiography or ventilation-perfusion lung scan been provided confirming thromboembolic occlusion of the proximal or distal pulmonary vasculature? ☐Yes ☐No

If the request is for Tyvaso (treprostinil) for a diagnosis of Pulmonary Hypertension associated with Interstitial Lung Disease (PH-ILD) (WHO Group 3), please answer the following questions:

• Please list any concurrent chronic lung disease diagnoses the member has:

CURRENT or PREVIOUS THERAPY

Medication Name	Strength/ Frequency	Dates of Therapy	Status (Discontinued & Why/Current)

REAUTHORIZATION

Has the member experienced a significant improvement with treatment? ☐ Yes ☐ No

Please describe:

SUPPORTING INFORMATION or CLINICAL RATIONALE

Prescribing Provider Signature	Date