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Request for Prior Authorization for Pulmonary Arterial Hypertension (PAH) agents
Website Form – www.highmarkhealthoptions.com
Submit request via: Fax - 1-855-476-4158

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

Pulmonary Arterial Hypertension (PAH) agents Prior Authorization Criteria:

Medications addressed in this policy			
Endothelin-Receptor Antagonists (ERAs)	**Phosphodiesterase type 5 inhibitors (PDE-5 inhibitors)**	**Soluble Guanylate Cyclase Stimulator**	**Prostanoids/prostacyclin therapies**
Ambrisentan Letairis (ambrisentan) **Bosentan** Tracleer (bosentan) Opsumit (macitentan)	**Sildenafil citrate** Adcirca (tadalafil) **Tadalafil** Revatio (sildenafil citrate) **Revatio Suspension (sildenafil)**	Adempas (riociguat)	**Ventavis (iloprost)** Epoprostenol Flolan (epoprostenol) Orenitram ER (treprostinil) Remodulin (treprostinil SC/IM) Tyvaso (treprostinil) Uptravi (selexipag) Veletri (epoprostenol)

For all requests for Pulmonary Arterial Hypertension (PAH) agents all of the following criteria must be met:

- 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, **all** of the following criteria must be met:
    - Chart documentation indicates member is currently on requested therapy
    - Documentation from prescriber substantiates a diagnosis of PAH WHO Group I, CTEPH (Adempas only), or PH-ILD (Tyvaso only)
        - If PAH WHO Group I, must provide 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 ≥ to 20 mmHg
            - Pulmonary capillary wedge pressure ≤ to 15 mmHg

- Pulmonary vascular resistance ≥ 3 Wood units.

- If CTEPH, must provide chart documentation of the following:
  - Mean pulmonary arterial pressure ≥ to 20 mmHg
  - Pulmonary capillary wedge pressure ≤ to 15 mmHg
  - Pulmonary vascular resistance ≥ 3 Wood units.
  - Thromboembolic occlusion of the proximal or distal pulmonary vasculature from computed tomographic pulmonary angiography (CT-PA) or ventilation-perfusion (V/Q) lung scan
- If PH-ILD, must have a concurrent chronic lung disease diagnosis (COPD, emphysema, pulmonary fibrosis, sarcoidosis, etc.) and provide chart documentation of right-heart catheterization (RHC) or echocardiography if the provider indicates RHC is not recommended. RHC documentation must meet one of the following hemodynamic value combinations:
  - Mean pulmonary arterial pressure ≥ to 25 mmHg
  - Mean pulmonary arterial pressure 21-24 mmHg and Pulmonary vascular resistance ≥ 3 Wood units.

A. Diagnosis 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 ≥ to 20 mmHg
  - Pulmonary capillary wedge pressure ≤ to 15 mmHg
  - Pulmonary vascular resistance ≥ 3 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 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 (refer below to Appendix II)
  At least one of the following:
  - The requested drug will be used as monotherapy
  - Ambrisentan and Tadalafil are requested as initial combination therapy
  - If the requested drug will be used for add-on therapy to existing monotherapy or dual therapy, then **both** of the following requirements must be met:
    - Medications are from different therapeutic classes

    - Member has unresponsive or progressive disease despite established PAH-specific therapies
- If the requested medication is oral and not being used with prostanoid/prostacyclin therapy for patients with WHO functional class IV symptoms, must have documentation of inadequate response, contraindication or intolerance to prostanoid/prostacyclin therapy (e.g. epoprostenol, iloprost, treprostinil)

Coverage may be provided with a diagnosis of Chronic Thromboembolic Pulmonary Hypertension (CTEPH) (WHO Group 4) and the following criteria is met:

- Request must be for Adempas.
- Member has a confirmed diagnosis of persistent/recurrent CTEPH after surgical treatment (i.e. pulmonary endarterectomy) or inoperable CTEPH.
- Documentation must meet all of the following:
    - Mean pulmonary arterial pressure ≥ to 20 mmHg
    - Pulmonary capillary wedge pressure ≤ to 15 mmHg
    - Pulmonary vascular resistance ≥ 3 Wood units.
    - Thromboembolic occlusion of the proximal or distal pulmonary vasculature from computed tomographic pulmonary angiography (CT-PA) or ventilation-perfusion (V/Q) lung scan

Coverage may be provided with a diagnosis of Pulmonary hypertension associated with interstitial lung disease (PH-ILD) (WHO Group 3) and the following criteria is met:

- Request must be for Tyvaso.
- Member has a diagnosis of PAH WHO Group 3 (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 meet one of the following hemodynamic value combinations:
    - Mean pulmonary arterial pressure ≥ to 25 mmHg
    - Mean pulmonary arterial pressure 21-24 mmHg and Pulmonary vascular resistance ≥ 3 Wood units.
- Member must have a concurrent chronic lung disease diagnosis (COPD, emphysema, pulmonary fibrosis, sarcoidosis, etc.)

B. Drug Criteria

- **Endothelin receptor-antagonists (ERA):** Letairis, **Tracleer**, Opsumit
    - If the request is for a non-preferred ERA, must have documentation of inadequate response, contraindication or intolerance to a preferred agent within this class
- **Phosphodiesterase type 5 inhibitors (PDE-5 inhibitors): sildenafil citrate tablets**, Adcirca, Revatio

- Member is not concurrently using nitrates (regularly or intermittently) or a guanylate cyclase stimulator (e.g. Adempas [riociguat])
- If the request is for Revatio suspension, must provide clinical rationale for why member is unable to take a solid dosage form (e.g. tablet)
- If the request is for Revatio tablets, Adcirca, or generic Tadalafil**, must have documentation of inadequate response, contraindication or intolerance to a preferred agent within this class

**If generic Tadalafil is requested with Ambrisentan as initial combination therapy, this requirement does not apply.

- **Soluble Guanylate Cyclase Stimulator**: Adempas
  - Member is not concurrently using nitrates (regularly or intermittently) or phosphodiesterase inhibitors
  - Must have documentation of inadequate response, contraindication or intolerance to a PDE-5 inhibitor (e.g. sildenafil, Revatio, Adcirca) unless being used for CTEPH
- **Prostanoids/prostacyclin therapies**
  - Inhaled agents: **Ventavis**, Tyvaso
    - If request is for PAH (WHO Group 1) then member must meet all the following:
      - Must have documentation the member has previously received an infused prostacyclin therapy (e.g. epoprostenol, Flolan, Veletri, Remodulin)
      - If the request is for a non-preferred inhaled agent, must have documentation of inadequate response, contraindication or intolerance to a preferred agent within this class
      - Must have 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 (VO2) <11ml/min/kg (<35% predicted)
          - Ventilatory equivalents of CO2 (VE/VCO2) slope ≥45
          - BNP >300 ng/l
          - NT-proBNP >1400ng/l
          - Right atrium area ≥26 cm2
          - Presence of pericardial effusion
          - Right atrial pressure >14 mmHg
          - Cardiac index <2.0 l/min/m2

        - Mixed venous oxygen saturation (SvO2) <60%
    - Infused agents: epoprostenol, Flolan, Veletri, Remodulin
        - Must have 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%
    - Oral agents: Uptravi, Orenitram ER
        - Must have documentation of inadequate response, contraindication or intolerance to at least one oral therapy from one of the following three therapeutic classes: a PDE-5 inhibitor (e.g. sildenafil, Adcirca, Revatio), an ERA (e.g. Letairis, Opsumit, Tracleer), or a guanylate cyclase stimulator (e.g. Adempas)
        - Member is not taking Uptravi or Orenitram ER in combination with a prostanoid/prostacyclin therapy (e.g. epoprostenol, iloprost, treprostinil)

Initial Duration of Approval: 3 months

Reauthorization criteria

- Must provide documentation that demonstrates member is tolerating and receiving clinical benefit from treatment.
- For Revatio suspension: must indicate that member remains unable to take a solid dosage form (e.g. tablet)

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.

Drugs are authorized in generic form unless the branded product is on the preferred drug list or the prescriber has indicated in writing that the branded product is medically necessary. If only the branded product is on the preferred drug list, the generic form will be considered non-preferred and shall not require the prescriber to indicate in writing that the branded product is medically necessary.

Appendix 1: World Health Organization (WHO) Clinical Classification of Pulmonary Hypertension (PH)

Group 1	Pulmonary Arterial Hypertension (PAH) • Idiopathic • Heritable • Drug/toxin-induced • Associated with: o Connective tissue disease o HIV infection o Portal hypertension o Congenital heart disease o Schistosomiasis
Group 1'	Pulmonary veno-occlusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH)
Group 1"	Persistent pulmonary hypertension of the newborn (PPHN)
Group 2	PH due to left heart disease
Group 3	PH due to lung diseases and/or hypoxia
Group 4	Chronic thromboembolic PH (CTEPH)and other pulmonary artery obstructions
Group 5	PH with unclear and/or multifactorial mechanisms

Appendix 2: World Health Organization (WHO) Functional Classification of Members with PH

Classification	**Physical Activity**	**Symptoms** (Dyspnea, fatigue, chest pain, syncope)
Class I	No limitation	None upon ordinary physical activity
Class II	Slight limitation	Symptoms appear upon ordinary physical activity
Class III	Marked limitation	Symptoms appear upon less than ordinary activity
Class IV	Severe limitation	Symptoms appear upon any physical activity or may even be present at rest; signs of right heart failure present

HIGHMARK HEALTH OPTIONS

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Pulmonary Arterial Hypertension (PAH) Agents (Oral and Inhaled)
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:** (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

PROVIDER INFORMATION

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

MEMBER INFORMATION

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

REQUESTED DRUG INFORMATION

Medication:	Strength:
Frequency:	Duration:
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 (if medically please provide a 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:

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? ☐ Monotherapy ☐ Combination
If **combination** therapy, please list other drug(s):

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 <2.0 l/min/m2
☐ BNP >300 ng/l ☐ NT-proBNP >1400ng/l ☐ Right atrium area ≥26 cm2 ☐ Right atrial pressure >14 mmHg

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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 Revatio Suspension, please explain why member is unable to take a solid dosage form (e.g. tablet):

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:

PREVIOUS THERAPY

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

REAUTHORIZATION

If the request is for Revatio Suspension, is the member still unable to take solid dosage forms? ☐ Yes ☐ No

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

SUPPORTING INFORMATION or CLINICAL RATIONALE

Prescribing Provider Signature	Date