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DMMA

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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	Activin signaling inhibitor
Ambrisentan Letairis (ambrisentan) Bosentan Tracleer (bosentan) Opsumit (macitentan)	Sildenafil citrate 20mg tablet Sildenafil 10mg/ml suspension Adcirca (tadalafil) Tadalafil Revatio (sildenafil citrate) Revatio Suspension (sildenafil) LiQrev (sildenafil)	Adempas (riociguat)	Ventavis (iloprost) Epoprostenol Flolan (epoprostenol) Orenitram ER (treprostinil) Remodulin (treprostinil SC/IM) Tyvaso (treprostinil) Uptravi (selexipag) Veletri (epoprostenol) Yutrepia (treprostinil)	Winrevair (sotatercept -csrK)

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 are 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.

- For non-preferred agents, must have a therapeutic failure, contraindication, or intolerance to the preferred agent(s) approved or medically accepted for the member's diagnosis and within the same class (e.g. for a non-preferred ERA all equivalent preferred ERA's have been tried or documentation of why they cannot be tried)
- If member is new to the plan and requests a continuation of therapy, the following criteria must be met:
 - Chart documentation indicates member is currently on requested therapy
- The member must meet diagnostic and drug criteria outlined in sections A and B

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 or echocardiography if the provider indicates RHC is not recommended. RHC documentation must contain the following hemodynamic values:
 - Mean pulmonary arterial pressure > 20 mmHg at rest
 - Pulmonary capillary wedge pressure \leq 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 (refer below to Appendix II)
 - For Winrevair (sotatercept-csrk) member has functional class II or III symptoms
- At least one of the following:
 - The requested drug will be used as monotherapy (except Winrevair (sotatercept-csrk) see drug specific criteria below)
 - 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 are met:

- Member has a diagnosis of CTEPH WHO Group 4 (refer below to Appendix I) confirmed by chart documentation of right-heart catheterization or echocardiography if the provider indicates RHC is not recommended. RHC documentation must contain the following hemodynamic values:
 - Mean pulmonary arterial pressure > 20 mmHg at rest
 - Pulmonary capillary wedge pressure ≤ 15 mmHg
 - Thromboembolic occlusion of the proximal or distal pulmonary vasculature from computed tomographic pulmonary angiography (CT-PA) or ventilation-perfusion (V/Q) lung scan
- Request must be for Adempas.
- Member has a confirmed diagnosis of persistent/recurrent CTEPH after surgical treatment (i.e. pulmonary endarterectomy) or inoperable CTEPH.

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:

- Member has a diagnosis of PH-ILD WHO Group 3 (refer below to Appendix I) confirmed by 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 > 20 mmHg
 - Pulmonary vascular resistance > 2 Wood units
- Must have a concurrent chronic lung disease diagnosis (COPD, emphysema, pulmonary fibrosis, sarcoidosis, etc.)
- Request must be for Tyvaso (Treprostinil).

B. DRUG CRITERIA

- **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])

- **Activin Signaling Inhibitor:**

- For Winrevair (sotatercept-csrk) 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

- **Soluble Guanylate Cyclase Stimulator:**

- Adempas [riociguat])
 - 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 (iloprost), Tyvaso (treprostinil)
 - 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)
 - 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 (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%
- 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

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- 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.

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) <ul style="list-style-type: none"> • Idiopathic • Heritable • Drug/toxin-induced • Associated with: <ul style="list-style-type: none"> ○ Connective tissue disease ○ HIV infection ○ Portal hypertension ○ Congenital heart disease ○ 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



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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



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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:
<input type="checkbox"/>
<input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> Class III <input type="checkbox"/> 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): ☐

Drug Name	Strength & Frequency	Rationale for additional therapy

Please check any boxes applicable to the member:	<input type="checkbox"/> Repeated periods of syncope, even with little or regular physical activity
<input type="checkbox"/> Clinical signs of right heart failure	<input type="checkbox"/> $\text{VO}_2 < 11 \text{ ml/min/kg}$ ($< 35\%$ predicted) meter
<input type="checkbox"/> Presence of pericardial effusion	<input type="checkbox"/> 6-minute walking distance (6MWD) index < 2.0
<input type="checkbox"/> Ventilatory equivalents of CO_2 (VE/VCO_2) slope > 45	<input type="checkbox"/> < 165 l/min/m ²
<input type="checkbox"/> Mixed venous oxygen saturation (SvO_2) $< 60\%$	<input type="checkbox"/> Cardiac



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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 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

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

Please describe:

SUPPORTING INFORMATION or CLINICAL RATIONALE

Prescribing Provider Signature

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