Updated: 05/2019
PARP Approved: 06/2019

Gateway Health
Prior Authorization Criteria
Pulmonary Arterial Hypertension (PAH) agents

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**
Letairis (ambrisentan) Tracleer (bosentan) Opsumit (macitentan)	**Sildenafil citrate** **Adcirca (tadalafil)** Revatio (sildenafil citrate)	Adempas (riociguat)	**Epoprostenol** Flolan (epoprostenol) Orenitram ER (treprostinil) Remodulin (treprostinil SC/IM) Tyvaso (treprostinil) Uptravi (selexipag) Ventavis (iloprost) Veletri (epoprostenol)

*__Bolded__ medications indicate preferred formulary medications

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 or CTEPH (Adempas 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 25 mmHg
            - Pulmonary capillary wedge pressure OR left atrial pressure OR left ventricular end-diastolic pressure ≤ to 15 mmHg
            - Pulmonary vascular resistance greater than 3 Wood units.
        - If CTEPH, must provide chart documentation of the following:
            - Mean pulmonary arterial pressure ≥ to 25 mmHg

- Pulmonary capillary wedge pressure ≤ to 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

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 ≥ to 25 mmHg
  - Pulmonary capillary wedge pressure **OR** left atrial pressure **OR** left ventricular end-diastolic pressure ≤ to 15 mmHg
  - Pulmonary vascular resistance greater than 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)
- One of the following:
  - The requested drug will be used as monotherapy
  - Letairis and Adcirca are requested as initial combination therapy
  - 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
- 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 (e.g. pulmonary endarterectomy) or inoperable CTEPH. Documentation must include the following:
    - Mean pulmonary arterial pressure ≥ to 25 mmHg
    - Pulmonary capillary wedge pressure ≤ to 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
- Member is not concurrently using nitrates (regularly or intermittently) or phosphodiesterase inhibitors

B. Drug Criteria

- **Endothelin receptor-antagonists (ERA): Letairis**, Tracleer, Opsumit
    - If the request is for Tracleer or Opsumit, must have documentation of inadequate response, contraindication or intolerance to the combination of Letairis and Adcirca
- **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 tablets, must have documentation of inadequate response, contraindication or intolerance to the combination of Letairis and Adcirca
    - 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)
- **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 the combination of Letairis and Adcirca
- **Prostanoids/prostacyclin therapies**
    - Inhaled agents: Tyvaso, Ventavis
        - Must have documentation of inadequate response, contraindication or intolerance to an infused prostacyclin therapy (e.g. epoprostenol, Flolan, Veletri, Remodulin)
        - 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%

- Infused agents: **epoprostenol**, Flolan, Veletri, Remodulin
  - 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%
  - For Flolan, Veletri, or Remodulin must provide documentation of inadequate response, contraindication or intolerance to generic epoprostenol or clinical rationale for why generic epoprostenol cannot be used
- Oral agents: Uptravi, Orenitram ER
  - Must have documentation of inadequate response, contraindication or intolerance to the combination of Letairis and Adcirca
  - 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.

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.

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

Group 1	Pulmonary Arterial Hypertension (PAH)
	• Idiopathic
	• Heritable
	• Drug/toxin-induced
	• Associated with:
	  ○ 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

Appendix II: 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

Gateway Health

Updated: 05/2019
PARP Approved: 06/2019

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 Gateway Health℠ Pharmacy Services. **FAX:** (888) 245-2049
If needed, you may call to speak to a Pharmacy Services Representative.
PHONE: (800) 392-1147 Monday through Friday 8:30am to 5:00pm

PROVIDER INFORMATION

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

MEMBER INFORMATION

Member Name:	DOB:
Gateway 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:

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: **ICD10 Code(s):**

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: _____

Left Atrial Pressure: _____ Left Ventricular End-diastolic 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

Gateway Health

Updated: 05/2019
PARP Approved: 06/2019

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 Gateway Health℠ Pharmacy Services. **FAX:** (888) 245-2049
If needed, you may call to speak to a Pharmacy Services Representative.
PHONE: (800) 392-1147 Monday through Friday 8:30am to 5:00pm

MEMBER INFORMATION

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

MEDICAL HISTORY (Complete for ALL requests)

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

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):

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