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

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

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** Revatio (sildenafil citrate) Adcirca (tadalafil)	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

PAH Agents Initial Prior Authorization Criteria

- Coverage will be provided when the diagnosis is **Pulmonary Arterial Hypertension (PAH) WHO Group I** and all of the corresponding criteria are met in **sections A, B and C:**

A. Diagnosis Criteria
- Treatment is prescribed by, or in consultation with, a cardiologist or pulmonologist; **AND**
- 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; **AND**
- Member has functional class II, III or IV symptoms (refer below to Appendix II); **AND**
- Proceed below to Section B. Treatment Criteria

B. Treatment 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 Letairis; **AND**
  - Proceed below to Section C. Planned PAH Treatment Regimen

- **Phosphodiesterase type 5 inhibitors (PDE-5 inhibitors): sildenafil citrate tablets**, Revatio, Adcirca
  - Member is not concurrently using nitrates (regularly or intermittently) or a guanylate cyclase stimulator (e.g. Adempas [riociguat]); **AND**
  - If the request is for Revatio tablets or Adcirca**, must have documentation of inadequate response, contraindication or intolerance to generic sildenafil citrate tablets

    ******If Adcirca is requested with Letairis as initial combination therapy, this requirement does not apply.

    OR
  - 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)

AND

- Proceed below to Section C. Planned PAH Treatment Regimen

- **Soluble Guanylate Cyclase Stimulator:** Adempas
  - Member is not concurrently using nitrates (regularly or intermittently) or phosphodiesterase inhibitors; **AND**
  - Must have documentation of inadequate response, contraindication or intolerance to a PDE-5 inhibitor (i.e. sildenafil citrate, Revatio, Adcirca); **AND**
  - Proceed below to Section C. Planned PAH Treatment Regimen

- **Prostanoids/prostacyclin therapies**
  - Inhaled agents: Tyvaso, Ventavis
    - 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 citrate, Adcirca or Revatio), an ERA (e.g. Letairis, Opsumit, Tracleer), or a guanylate cyclase stimulator (e.g. Adempas) **OR** documentation the member has previously received an infused prostacyclin therapy (e.g. epoprostenol, Flolan, Veletri, Remodulin); **AND**
    - Proceed below to Section C. Planned PAH Treatment Regimen

  - Infused agents: **epoprostenol**, Flolan, Veletri, Remodulin
    - 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
      AND
    - Proceed below to Section C. Planned PAH Treatment Regimen

  - Oral agents: Uptravi, Orenitram ER
    - For Uptravi:
      - 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 citrate, Adcirca or Revatio), an ERA (e.g. Letairis, Opsumit, Tracleer), or a guanylate cyclase stimulator (e.g. Adempas); **AND**
      - Member is not taking Uptravi in combination with a prostanoid/prostacyclin therapy (i.e. epoprostenol, iloprost, treprostinil); **AND**
      - Proceed below to Section C. Planned PAH Treatment Regimen

    - For 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 citrate, Adcirca or Revatio), an ERA (e.g. Letairis, Opsumit, Tracleer), or a guanylate cyclase stimulator (e.g. Adempas); **AND**
      - Member is not taking Orenitram ER in combination with a prostanoid/prostacyclin therapy (i.e. epoprostenol, iloprost, treprostinil); **AND**
      - Proceed below to Section C. Planned PAH Treatment Regimen

C. Planned PAH Treatment Regimen

- The requested drug will be used as monotherapy; **OR**
- Letairis and Adcirca are requested as initial combination therapy; **OR**
- 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:
  i. Medications are from different therapeutic classes
  AND
  ii. Member has unresponsive or progressive disease despite established PAH-specific therapies

- Coverage will be provided when the diagnosis is **Chronic Thromboembolic Pulmonary Hypertension (CTEPH) (WHO Group 4)** and all of the following criteria are met:
  - For Adempas:
    - Treatment is prescribed by, or in consultation with, a cardiologist or pulmonologist; **AND**
    - Member has a confirmed diagnosis of persistent/recurrent CTEPH after surgical treatment (i.e. pulmonary endarterectomy) or inoperable CTEPH; **AND**
    - Member is not concurrently using nitrates (regularly or intermittently) or phosphodiesterase inhibitors

- 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; **AND**
  - Documentation from prescriber substantiates a diagnosis of PAH WHO Group I or CTEPH (depending on agent requested); **AND**
  - Treatment is prescribed by, or in consultation with, a cardiologist or pulmonologist

- The requested dose is within FDA-approved dosing recommendations. 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.

- Initial approval duration will be for 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 will be for 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 prior authorization 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: 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 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

References

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2.) Galie N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS). *Eur Heart J*. 2016:37(1):67-119. Available at: http://http://eurheartj.oxfordjournals.org/content/37/1/67.long. Accessed March 29, 2016.
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10.) Ventavis [package insert]. Actelion Pharmaceuticals US, Inc.; Nov 2013.
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14.) Remodulin [package insert]. Research Triangle Park, NC: United Therapeutics Corp.; Sept 2013.
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