Washington State Health Care Authority

Pulmonary Hypertension (PH) Agents (Oral/Inhalation)

Medical policy no. 40.12.00-2 Effective Date: August 1, 2018

Note: *New-to-market drugs included in this class based on the Apple Health Preferred Drug List are non-preferred and subject to this prior authorization (PA) criteria. Non-preferred agents in this class require an inadequate response or documented intolerance due to severe adverse reaction or contraindication to at least TWO preferred agents. If there is only one preferred agent in the class documentation of inadequate response to ONE preferred agent is needed. If a drug within this policy receives a new indication approved by the Food and Drug Administration (FDA), medical necessity for the new indication will be determined on a case-by-case basis following FDA labeling.*

To see the list of the current Apple Health Preferred Drug List (AHPDL), please visit: https://www.hca.wa.gov/assets/billers-and-providers/apple-health-preferred-drug-list.xlsx

Background:

Pulmonary hypertension (PH) is a rare, progressive disorder characterized by high blood pressure (hypertension) in the arteries of the lungs (pulmonary artery). The pulmonary arteries are the blood vessels that carry deoxygenated blood from the right side of the heart to the lungs. The World Health Organization (WHO) classifies pulmonary hypertension into five groups based upon etiology. WHO Group I is classified as pulmonary arterial hypertension (PAH), while the other four groups are referred to as pulmonary hypertension (PH)

PH may develop from many different conditions, but the most common type is idiopathic PAH. Common symptoms of PAH include shortness of breath (dyspnea), chest pain, and fainting.

Medical necessity

Drug	Medical Necessity
Endothelin Receptor Antagonists • ambrisentan (**LETAIRIS®**) • bosentan (**TRACLEER®**) • macitentan (**OPSUMIT®**)	Medications listed in this table may be considered medically necessary when used for the treatment of: • Pulmonary hypertension (PH) • Chronic thromboembolic pulmonary hypertension (CTEPH)
Phosphodiesterase Inhibitors (PDEI) • sildenafil citrate tablets (**REVATIO®**) • tadalafil (**ADCIRCA®, ALYQ™**)	
Prostacyclin Pathway Agonists • Iloprost (**VENTAVIS®**) • selexipag (**UPTRAVI®**) • treprostinil (**ORENITRAM®/TYVASO®**)	
Soluble Guanylate Cyclase (SGC) Stimulator • riociguat (**ADEMPAS®**)	

Clinical policy:

Clinical Criteria	
Pulmonary Hypertension (PH) **Chronic Thromboembolic Pulmonary Hypertension (CTEPH)**	Medications requested for the treatment of PAH may be authorized when **ALL** of the following are met: 1. Patient must have **ONE** (either a or b) of the following diagnoses and criteria: **a.** PH diagnosis WHO Groups 3 or 4 (CTEPH), in which general treatment measures (e.g., anticoagulation) have failed, and PH is thought to be unrelated to underlying lung disease; **OR** b. PAH diagnosis WHO Group 1 i. Documentation of PAH WHO Functional class (II, III, or IV); **AND** ii. History of failure, contraindication, or intolerance to amlodipine, diltiazem, or long-acting nifedipine **EXCEPT** for the following circumstances: (1) Patient had a negative response to acute vasoreactivity testing (AVT); **OR** (2) AVT is not indicated for the patient (PAH due to connective tissue disease, congenital heart disease, HIV, portal hypertension, schistosomiasis, pulmonary veno-occlusive/pulmonary capillary hypertension); **OR** **(3)** AVT is contraindicated (SBP < 90 mmHg; cardiac index < 2 L/min/m^2, or PH functional class IV); **AND** **2.** Requested therapy is not for **ANY** of the following: a. A combination of a phosphodiesterase inhibitor and soluble guanylate cyclase stimulator; **OR** b. A combination of selexipag and parenteral prostanoid; **OR** 3. Patient is currently established on requested therapy; **OR** 4. <u>For Selexipag</u>: history of failure, contraindication or intolerance to an endothelin receptor antagonist; **AND** 5. Prescribed by or in consultation with a specialist in cardiology or pulmonology If all of the above criteria are met, the request will be **approved for 12 months** If all criteria are not met, but there are documented medically necessary circumstances based on the professional judgement of the clinical reviewer, requests may be approved on a case-by-case basis up to the initial authorization duration.
	Criteria (Reauthorization)

	Medications used for the treatment of PH may be reauthorized when documentation of response (e.g. disease stability or mild progression indicated by a slowing of decline using WHO Functional Class scale) is provided. If all of the above criteria are met, the request will be **approved for 12 months** If all criteria are not met, but there are circumstances supported by clinical judgement and documentation, requests may be approved by a clinical reviewer on a case-by-case basis up to the reauthorization duration

Coding:

HCPCS Code	Description
J7686	Treprostinil, inhalation solution, FDA-approved final product, non-compounded, administered through DME, unit dose form, 1.74 mg
Q4074	Iloprost, inhalation solution, FDA-approved final product, non-compounded, administered through DME, unit dose form, up to 20 mcg

Dosage and Quantity Limits:

Drug Name	Dose and Quantity Limits
Ambristentan (Letairis)	10 mg per day; #60 tablets per 30 day supply (5 mg strength) **OR** #30 tablets for 30 day supply (10 mg strength)
Bosentan (Tracleer)	250 mg per day; #60 tablets per 30 day supply
Macitentan (Opsumit)	10 mg per day; #30 tablets per 30 day supply
Sildenafil citrate (Revatio)	60 mg per day; #90 tablets per 30 day supply
Tadalafil (Adcirca)	40 mg per day; #60 tablets per 30 day supply
Selexipag (Uptravi)	3200 mcg per day; can increase to the highest tolerated dose in 200 mcg twice daily increments at weekly intervals
Iloprost (Ventavis)	45 mcg per day;
Treprostinil (Tyvaso)	216 mcg per day;
Treprostinil diolamine (Orenitram)	0.5 mg per day; titrate by 0.25 mg or 0.5 mg twice daily **OR** 0.125 mg 3 times daily not more than every 3-4 days to the highest tolerated dose
Riociguat (Adempas)	7.5 mg per day;

Appendix:

Table 1. WHO Clinical Classification of Pulmonary Hypertension (PH)

WHO Clinical Classification	Description
Group 1	Pulmonary Arterial Hypertension (PAH) - Idiopathic - Heritable - Drug/toxin induced

	- Associated with connective tissue disease, HIV infection, portal hypertension, congenital heart disease
Group 2	PH due to left heart disease
Group 3	PH due to chronic lung disease or hypoxemia
Group 4	Chronic thromboembolic pulmonary hypertension (CTEPH)
Group 5	PH due to unclear multifactorial mechanisms

Table 2. WHO Functional Classification of Patients with PH

WHO Functional Classification	Description
Class I	Patients with PH without resulting limitation of physical activity. Ordinary physical activity does not cause undue dyspnea or fatigue, chest pain, or near syncope.
Class II	Patients with PH resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity causes undue dyspnea or fatigue, chest pain, or near syncope
Class III	Patients with PH resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity causes undue dyspnea or fatigue, chest pain, or near syncope.
Class IV	Patients with PH with inability to carry out any physical activity without symptoms. These patients manifest signs of right-sided heart failure. Dyspnea and/or fatigue may even be present at rest. Discomfort is increased by any physical activity

References

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History

Date	Action and Summary of Changes
02/17/2021	Approved by DUR Board
10/29/2020	Annual policy update - Updated preferred/non-preferred status - Updated PAH clinical criteria - Updated CTEPH clinical criteria
10/02/2019	Edit Note
11/07/2018	HCPCS update
07/31/2018	Update
08/16/2017	New Policy