Pulmonary Arterial Hypertension (PAH) Agents (Oral/Inhalation)

WA.PHAR.55 Pulmonary Arterial Hypertension (PAH) Agents (Oral & Inhalation

Effective Date: July 1, 2018

Background:

Pulmonary arterial hypertension (PAH) is a rare, progressive disorder characterized by high blood pressure (hypertension) in the arteries of the lungs (pulmonary artery) for many different reasons, with the most common being idiopathic PAH. The pulmonary arteries are the blood vessels that carry blood from the right side of the heart through the lungs. Symptoms of PAH include shortness of breath (dyspnea) especially during exercise, chest pain, and fainting episodes.

Medical necessity

Drug	Medical Necessity
Endothelin Receptor Antagonists • ambrisentan (**LETAIRIS®**) • bosentan (**TRACLEER®**) • macitentan (**OPSUMIT®**)	Agents may be considered medically necessary when used for the treatment of pulmonary hypertension • Non-preferred products require a trial of sildenafil (generic REVATIO®) and two (2) preferred products in the same subclass unless contraindicated or only one preferred product is available
Phosphodiesterase Inhibitors (PDEI) • sildenafil citrate (REVATIO®) • tadalafil (ADCIRCA®)	
Prostacyclin Receptor Agonists • selexipag (**UPTRAVI®**)	
Prostaglandin Vasodilators • iloprost (**VENTAVIS®**) • treprostinil (**ORENITRAM®**) • treprostinil (**TYVASO®**)	
Soluble Guanylate Cyclase (SGC) Stimulator • riociguat (**ADEMPAS®**)	

Clinical policy:

Clinical Criteria	
Pulmonary Arterial Hypertension	1. Diagnosis of pulmonary arterial hypertension confirmed by, or contraindication to right heart catheterization a. History of **ONE** of the following:

	i. Currently on any therapy for diagnosis of pulmonary arterial hypertension (PAH) ii. **BOTH** of the following: 1. History of failure, contraindication or intolerance sildenafil (generic Revatio®) 2. History of failure, contraindication or intolerance to a preferred product in the same subclass with the same indication 2. Prescribed by or in consultation with a cardiologist or pulmonologist **Approve for 12 months** **Criteria (Reauthorization)** Documentation of positive clinical benefit **Approve for 12 months**
Chronic Thromboembolic Pulmonary Hypertension (CTEPH) (WHO Group 4)	1. Diagnosis of persistent/recurrent Chronic Thromboembolic Pulmonary Hypertension (CTEPH) (WHO Group 4) and **ALL** of the following: a. Current anticoagulant therapy **OR** history of failure, contraindication, or intolerance to anticoagulant therapy b. History of **ONE** of the following: i. Currently on any therapy for diagnosis of CTEPH ii. Previous surgical treatment for CTEPH iii. Inoperable CTEPH due to **ONE** of the following: 1. Accessibility of the thrombi 2. Hemodynamic and/or ventilator impairment 3. Comorbid conditions 4. Patient acceptance 2. Greater than or equal to (≥) 18 years of age 3. Prescribed by or in consultation with a cardiologist or pulmonologist **Approve for 12 months** **Criteria (Reauthorization)** Documentation of positive clinical benefit **Approve for 12 months**

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