CareSource

PHARMACY POLICY STATEMENT
North Carolina Marketplace

DRUG NAME	Endothelin Receptor Antagonists for Pulmonary Arterial Hypertension: Letairis (ambrisentan), Opsumit (macitentan), Tracleer (bosentan)
BILLING CODE	Must use valid NDC code
BENEFIT TYPE	Pharmacy
SITE OF SERVICE ALLOWED	Home
STATUS	Prior Authorization Required

Pulmonary Arterial Hypertension is a rare but serious condition characterized by elevated pulmonary arterial resistance. Letairis, Opsumit and Tracleer are endothelin receptor antagonists approved for the treatment of pulmonary arterial hypertension (PAH) World Health Organization (WHO) Group 1. Letairis is indicated to improve exercise ability and delay clinical worsening in PAH. It can also be used in combination with tadalafil to reduce the risks of disease progression and hospitalization for worsening PAH, and to improve exercise ability. Opsumit is indicated for the treatment of PAH to reduce the risks of disease progression and hospitalization. Tracleer is indicated in adults to improve exercise ability and to decrease clinical worsening for PAH. It can also be used in pediatric patients with idiopathic or congenital PAH to improve pulmonary vascular resistance.

Endothelin Receptor Antagonists will be considered for coverage when the following criteria are met:

Pulmonary Arterial Hypertension [WHO Group 1]

For **initial** authorization:

1. Tracleer: Member is at least three years of age or older;
   Letairis and Opsumit: Member is at least eighteen years of age or older;
2. Medication must be prescribed by or in consultation with a cardiologist or pulmonologist; AND
3. Member must have a diagnosis of World Health Organization (WHO) Group 1 pulmonary arterial hypertension (PAH) confirmed by right heart catheterization;
4. Member must have documentation pulmonary arterial pressures are not adequately controlled, confirmed by **one** of the following:
   a) Patient had an acute response to vasodilator testing AND has tried a calcium channel blocker (CCB) for at least 3 months; OR
   b) Patient did not have a response to vasodilator testing; OR
   c) Patient cannot undergo vasodilator testing; OR
   d) Patient cannot take CCB therapy
5. **Dosage allowed/Quantity limit:**
   Opsumit: 10 mg once daily
   Letairis: Initiate treatment at 5 mg once daily; Increase to 10mg once daily every 4 weeks as tolerated;

Tracleer: **Patients 12 years and older**: Initially 62.5 mg PO BID for 4 weeks, then increased to 125 mg PO BID (maximum 250 mg/day); **Patients 12 years and younger**: initial and maintenance dosing is weight-based:
≥ 4-8 kg: 16 mg twice daily
> 8-16 kg: 32 mg twice daily
> 16-24 kg: 48 mg twice daily
> 24-40 kg: 64 mg twice daily

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
Endothelin Receptor Antagonists will be reauthorized when chart notes show at least one of the following:
1. Member has documentation of improvement in signs and symptoms of disease as evidenced by at least one of the following:
   a) Stabilization or improvement in WHO functional class (see appendix)
   b) Stabilization or improvement in 6MWD [6-minute walk distance]
   c) Reduction in PAH-related hospitalizations

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Endothelin Receptor Antagonists not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
06/15/2011	Pulmonary Arterial Hypertension policy creation.
05/13/2014	Combined all PAH agents into one policy
07/09/2015	Revised guidelines for therapy aligning with CMS
08/18/2015	Revised guidelines to include diagnosis criteria
10/13/2021	Separated PAH agents by drug class; Updated guidelines; Added provider specialty

References:

1. Letairis [package insert]. Foster City, CA: Gilead Sciences, Inc; August 2019
2. Opsumit [package insert]. San Francisco, CA: Actelion Pharmaceuticals US, Inc.; February 2020
3. Tracleer [package insert]. San Francisco, CA: Actelion Pharmaceuticals US, Inc.; May 2019
4. Coons, J.C., Pogue, K., Kolodziej, A.R. et al. Pulmonary Arterial Hypertension: a Pharmacotherapeutic Update. Curr Cardiol Rep. 2019; 21(141)
5. Klinger JR, Elliott CG et al. Therapy for Pulmonary Arterial Hypertension in Adults; Chest Journal. March 2019; 155(3): 565-586
6. Galie N, Humbert M, Vachiery JL, Gibbs S, Lang I, Torbicki A, 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): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). European heart journal. 2016;37(1):67–119

Effective date: 01/01/2023
Creation date: 10/13/2021

Appendix:

World Health Organization Functional Assessment Classification	
Class I	Patients with PAH but without resulting limitation of physical activity. Ordinary physical activity does not cause undue dyspnea, fatigue, chest pain or near syncope.
Class II	Patients with PAH resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity increases dyspnea, fatigue, chest pain, or near syncope.
Class III	Patients with PAH resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity increases dyspnea, fatigue, chest pain, or near syncope.
Class IV	Patients with PAH unable to carry out any physical activity without symptoms. These patients may have signs of right-heart failure. Dyspnea and/or fatigue may even be present at rest. Discomfort is increased by any physical activity.