

Pharmacy Medical Necessity Guidelines: Pulmonary Hypertension Medications

Effective: February 15, 2021

Prior Authorization Required	✓	Type of Review – Care Management	
Not Covered		Type of Review – Clinical Review	✓
Pharmacy (RX) or Medical (MED) Benefit	RX/ MED	Department to Review	RxUM/ PRECERT /MM
<p>These pharmacy medical necessity guidelines apply to the following:</p> <p>Commercial Products</p> <ul style="list-style-type: none"> <input checked="" type="checkbox"/> Tufts Health Plan Commercial products – large group plans <input checked="" type="checkbox"/> Tufts Health Plan Commercial products – small group and individual plans <input checked="" type="checkbox"/> Tufts Health Freedom Plan products – large group plans <input checked="" type="checkbox"/> Tufts Health Freedom Plan products – small group plans • CareLinkSM – Refer to CareLink Procedures, Services and Items Requiring Prior Authorization <p>Tufts Health Public Plans Products</p> <ul style="list-style-type: none"> <input checked="" type="checkbox"/> Tufts Health Direct – A Massachusetts Qualified Health Plan (QHP) (a commercial product) <input checked="" type="checkbox"/> Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans <input checked="" type="checkbox"/> Tufts Health RITogether – A Rhode Island Medicaid Plan 		<p>Fax Numbers:</p> <p><i>Adempas, ambrisentan, bosentan, Opsumit, Orenitram sildenafil, tadalafil, Tracleer, Uptravi</i> RXUM: 617.673.0988</p> <p><i>epoprostenol, Flolan, Remodulin, Tyvaso, Veletri, Ventavis</i> Commercial Products PRECERT: 617.972.9409</p> <p>Tufts Health Public Plans Products MM: 888.415.9055</p>	

Note: This guideline does not apply to Medicare Members (includes dual eligible Members).

OVERVIEW

FOOD AND DRUG ADMINISTRATION-APPROVED INDICATIONS

Adcirca (tadalafil) is a phosphodiesterase 5 (PDE5) inhibitor indicated for the treatment of pulmonary arterial hypertension (PAH) (World Health Organization [WHO] Group 1) to improve exercise ability. Studies establishing effectiveness included predominately patients with New York Heart Association (NYHA) Functional Class II – III symptoms and etiologies of idiopathic or heritable PAH (61%) or PAH associated with connective tissue diseases (23%).

Adempas (riociguat) is a soluble guanylate cyclase stimulator indicated:

- For the treatment of adults with PAH, (WHO Group 1), to improve exercise capacity, WHO functional class and to delay clinical worsening. Efficacy was shown in patients on Adempas (riociguat) monotherapy or in combination with endothelin receptor antagonists or prostanoids. Studies establishing effectiveness included predominately patients with WHO functional class II–III and etiologies of idiopathic or heritable PAH (61%) or PAH associated with connective tissue diseases (25%).
- For the treatment of adults with persistent/recurrent chronic thromboembolic pulmonary hypertension (CTEPH), (WHO Group 4) after surgical treatment, or inoperable CTEPH, to improve exercise capacity and WHO functional class.

Flolan (epoprostenol sodium) is a prostacyclin vasodilator indicated for the treatment of PAH (WHO Group 1) to improve exercise capacity. Studies establishing effectiveness included predominantly patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH or PAH associated with connective tissue diseases.

Letairis (ambrisentan) is an endothelin receptor antagonist (ERA) indicated for the treatment of PAH (WHO Group 1) to improve exercise ability and delay clinical worsening and in combination with tadalafil to reduce the risks of disease progression and hospitalization for worsening PAH, and to improve exercise ability. Studies establishing effectiveness included predominantly patients with WHO Functional Class II–III symptoms and etiologies of idiopathic or heritable PAH (60%) or PAH associated with connective tissue diseases (34%).

Opsumit (macitentan) is an ERA indicated for the treatment of PAH (WHO Group I) to delay disease progression. Disease progression included: death, initiation of intravenous or subcutaneous prostanoids, or clinical worsening of PAH (decreased 6-minute walk distance, worsened PAH symptoms and need for additional PAH treatment). Opsumit (macitentan) also reduced hospitalization for PAH. Effectiveness was established in a long-term study in PAH patients with predominantly WHO Functional Class II-III symptoms treated for an average of 2 years. Patients were treated with Opsumit (macitentan) monotherapy or in combination with phosphodiesterase-5 inhibitors or inhaled prostanoids. Patients had idiopathic and heritable PAH (57%), PAH caused by connective tissue disorders (31%), and PAH caused by congenital heart disease with repaired shunts (8%).

Orenitram (treprostinil) is a prostacyclin vasodilator indicated for the treatment of PAH (WHO Group 1) to improve exercise capacity. The study that established effectiveness included predominately patients with WHO functional class II-III symptoms and etiologies of idiopathic or heritable PAH (75%) or PAH associated with connective tissue disease (19%). When used as the sole vasodilator, the effect of Orenitram on exercise is about 10% of the deficit, and the effect, if any, on a background of another vasodilator is probably less than this. Orenitram (treprostinil) is probably most useful to replace subcutaneous, intravenous, or inhaled treprostinil, but this use has not been studied.

Remodulin (treprostinil sodium) is a prostacyclin vasodilator indicated:

- For the treatment of PAH (WHO Group 1) to diminish symptoms associated with exercise. Studies establishing effectiveness included patients with NYHA Functional Class II-IV symptoms and etiologies of idiopathic or heritable PAH (58%), PAH associated with congenital systemic-to-pulmonary shunts (23%), or PAH associated with connective tissue diseases (19%).
- In patients with PAH requiring transition from Flolan (epoprostenol sodium) to diminish the rate of clinical deterioration. The risks and benefits of each drug should be carefully considered prior to transition.

Revatio (sildenafil) is a PDE5 inhibitor indicated for the treatment of PAH (WHO Group I) in adults to improve exercise ability and delay clinical worsening. The delay in clinical worsening was demonstrated when Revatio (sildenafil) was added to background epoprostenol therapy. Studies establishing effectiveness were short-term (12 to 16 weeks), and included predominately patients with NYHA Functional Class II-III symptoms and idiopathic etiology (71%) or associated with connective tissue disease (CTD) (25%). Adding sildenafil to bosentan therapy does not result in any beneficial effect on exercise capacity.

Tracleer (bosentan) is an ERA indicated for the treatment of PAH (WHO Group 1)

- In adults to improve exercise ability and to decrease clinical worsening. Studies establishing effectiveness included predominantly patients with WHO Functional Class II-IV symptoms and etiologies of idiopathic or heritable PAH (60%), PAH associated with connective tissue diseases (21%), and PAH associated with congenital heart disease with left-to-right shunts (18%).
- In pediatric patients aged 3 years and older with idiopathic or congenital PAH to improve pulmonary vascular resistance, which is expected to result in an improvement in exercise ability.

Tyvaso (treprostinil) is a prostacyclin vasodilator, available as an inhalation solution and indicated for the treatment of PAH (WHO Group 1) to improve exercise ability. Studies establishing effectiveness included predominately patients with NYHA Functional Class III symptoms and etiologies of idiopathic or heritable PAH (56%) or PAH associated with connective tissue diseases (33%). The effects diminish over the minimum recommended dosing interval of 4 hours; treatment timing can be adjusted for planned activities. While there are long-term data on use of treprostinil by other routes of administration, nearly all controlled clinical experience with inhaled treprostinil has been on a background of bosentan (an endothelin receptor antagonist) or sildenafil (a phosphodiesterase type 5 inhibitor). The controlled clinical experience was limited to 12 weeks in duration.

Uptravi (selexipag) is a prostacyclin receptor antagonist indicated for the treatment of PAH (WHO Group I) to delay disease progression and reduce the risk of hospitalization for PAH. Effectiveness was established in a long-term study in PAH patients with WHO Functional Class II-III symptoms. Patients had idiopathic and heritable PAH (58%), PAH associated with connective tissue disease (29%), PAH associated with congenital heart disease with repaired shunts (10%).

Veletri (epoprostenol sodium) is indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to improve exercise capacity. Studies establishing effectiveness included predominantly patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH or PAH associated with connective tissue diseases.

Ventavis (iloprost) is a synthetic analog of prostacyclin indicated for the treatment of PAH (WHO Group 1) to improve a composite endpoint consisting of exercise tolerance, symptoms (NYHA Class), and lack of deterioration. Studies establishing effectiveness included predominately patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH (65%) or PAH associated with connective tissue diseases (23%).

COVERAGE GUIDELINES

Chronic-Thromboembolic Pulmonary Hypertension

The plan may authorize coverage of Adempas (riociguat) for Members when the following criteria are met:

1. The Member has a definitive diagnosis of pulmonary arterial hypertension (World Health Organization Group 4) and confirmed by right heart catheterization
- AND**
2. The prescriber is a cardiologist or pulmonologist
- AND**
3. Documentation of **one (1) of the following:**
 - a. The chronic thromboembolic pulmonary hypertension persists or recurs after surgical treatment
 - b. Member is diagnosed as inoperable by a center specializing in chronic thromboembolic pulmonary hypertension or pulmonary thromboendarterectomy

Pulmonary Arterial Hypertension (PAH)

The plan may authorize coverage of Adempas (riociguat), ambrisentan, bosentan tablets, epoprostenol sodium, Flolan (epoprostenol sodium), Opsumit (macitentan), Orenitram (treprostinil), Remodulin (treprostinil sodium), sildenafil, tadalafil, Tracleer (bosentan) tablet for oral suspension, Tyvaso (treprostinil), Uptravi (selexipag), Veletri (epoprostenol sodium), or Ventavis (Ioprost) for Members when the following criteria are met:

Note: See additional coverage criteria for brand Flolan (epoprostenol sodium) and Veletri (epoprostenol sodium) for Pulmonary Arterial Hypertension below

1. The Member must have a definitive diagnosis of Pulmonary Arterial Hypertension (World Health Organization [WHO] Group I; see **APPENDIX**) and confirmed by right heart catheterization
- AND**
2. The prescriber is a cardiologist or pulmonologist

Note: Please refer to References Section for a description of NYHA and WHO Functional Class descriptions.

Additional Coverage Criteria for Flolan (epoprostenol sodium) and Veletri (epoprostenol sodium) for Pulmonary Arterial Hypertension

In addition to the coverage criteria above for Pulmonary Arterial Hypertension, the plan may authorize coverage of Flolan (epoprostenol sodium) or Veletri (epoprostenol sodium) when all of the following criteria are met:

1. Documented failure of an adequate trial of or inability to tolerate generic epoprostenol

APPENDIX

World Health Organization (WHO) Group 1 Classification of Pulmonary Arterial Hypertension

- Idiopathic pulmonary arterial hypertension (primary pulmonary hypertension)
- Heritable pulmonary arterial hypertension
- Drug- and toxin-induced pulmonary arterial hypertension
- Pulmonary arterial hypertension associated with:
 - Connective tissue diseases
 - Human immunodeficiency virus infection
 - Portal hypertension
 - Congenital heart disease
 - Schistosomiasis
- Pulmonary arterial hypertension long-term responders to calcium channel blockers
- Pulmonary arterial hypertension with overt features of venous/capillaries involvement
- Persistent pulmonary hypertension of the newborn

LIMITATIONS

- The plan will not authorize coverage of Adempas (riociguat), ambrisentan, bosentan tablets, epoprostenol sodium, Flolan (epoprostenol sodium), Opsumit (macitentan), Orenitram (treprostinil), Remodulin (treprostinil sodium), sildenafil, tadalafil, Tracleer (bosentan) tablet for oral suspension, Tyvaso (treprostinil), Uptravi (selexipag), Veletri (epoprostenol sodium), or Ventavis (loprost) for pulmonary hypertension secondary to the following conditions:
 - Diseases of the left atrium and ventricle such as congestive heart failure or cardiomyopathy
 - Diseases of the mitral and aortic valves
 - Chronic lung diseases such as chronic obstructive pulmonary disease, restrictive pulmonary disease or interstitial pulmonary disease
 - Obstructive sleep apnea or other sleep disorders involving breathing or alveolar hyperventilation disorders
- Adempas (riociguat) will not be approved if administered concomitantly with phosphodiesterase inhibitors or nitrates.
- The plan does not cover the following brand medications: Adcirca, Letairis, Revatio, and Tracleer tablets. Refer to the Pharmacy Medical Necessity Guidelines for non-covered medications.

CODES

The following HCPCS/CPT code(s) are:

Code	Description
J1325	Injection, epoprostenol, 0.5mg
J3285	Injection, treprostinil
J7686	Treprostinil, inhalation solution, FDA-approved final product, noncompounded, administered through DME, unit dose form, 1.74 mg
Q4074	Iloprost, inhalation solution, administered through DME, up to 20mcg

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APPROVAL HISTORY

May 2003: Reviewed by Pharmacy & Therapeutics Committee.

Subsequent endorsement date(s) and changes made:

1. May 10, 2005: No changes.
2. September 13, 2005: Add Ventavis (iloprost) to the TOPIC, CLINICAL COVERAGE CRITERIA and LIMITATIONS. Change criteria #1 from "The Member must have the definitive diagnosis of either Primary or Secondary symptomatic Pulmonary Hypertension (PH) from a cardiologist or pulmonologist confirmed by the two diagnostic tests, Right-heart cardiac catheterization and Echocardiogram showing right atrial and ventricular enlargement" to "The Member must have a definitive diagnosis of pulmonary artery hypertension (WHO group I) from a cardiologist or pulmonologist and confirmed by right heart catheterization". Add criteria #3, "The Member's symptoms are New York Heart Association (NYHA) class III to IV" (see table above).
3. November 8, 2005: Change the brand name "Viagra" in the Topic, Coverage Criteria and Coverage Limitations to sildenafil. Add the coverage limitations: "Tufts Health Plan may authorize coverage of Viagra for pulmonary hypertension when the dosage for sildenafil exceeds 20mg three times a day."
4. October 10, 2006: No changes.
5. November 13, 2007: Add Letairis (ambrisentan) to the Overview, Pharmacy Coverage Guidelines, and Limitations. Add description of WHO Pulmonary Arterial Hypertension Functional Classes
6. November 11, 2008: No changes.
7. September 8, 2009: For effective date December 1, 2009: Added guidelines for "*Coverage for Flolan" Added WHO class II to FDA approved functional class for Tracleer.
8. November 10, 2009: Added Adcirca to Pharmacy Coverage Guidelines. Added dispensing limitation for Adcirca and Revatio to Pharmacy Medical Necessity Guidelines. Updated FDA Approved Functional Class of Symptoms for sildenafil. Revised WHO Classification of Pulmonary Hypertension Group I
9. January 1, 2010: Removal of Tufts Health Plan Medicare Preferred language (separate criteria have been created specifically for Tufts Health Plan Medicare Preferred)
10. January 12, 2010: Added Tyvaso (treprostinil) inhalation solution to Medical Necessity Guidelines
11. May 11, 2010: Clarified dosage form of sildenafil and Revatio as tablets
12. January 1, 2011: Administrative Update: Added reimbursement code J7686. Removed discontinued code Q4080
13. January 11, 2011: Added Veletri (epoprostenol for injection) to Medical Necessity Guidelines
14. January 10, 2012: No changes.
15. January 15, 2013: Added quantity limitation for sildenafil 20mg tablets.
16. October 15, 2013: Removed quantity limitation from Adcirca, Revatio and sildenafil. Updated table of FDA Approved Functional Class of Symptoms to reflect current FDA-approved prescribing information.
17. February 11, 2014: Added Adempas (riociguat) and Opsumit (macitentan) to Medical Necessity Guidelines.
18. June 10, 2014: Added Orenitram (treprostinil) to Medical Necessity Guidelines, and Updated WHO Group I classifications under criteria #1: Removed: Chronic hemolytic anemias and Added: Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis and Persistent pulmonary hypertension of the newborn (PPHN). Updated criterion #4 under Chronic-Thromboembolic Pulmonary Hypertension to "Member is diagnosed as inoperable by a center specializing in chronic thromboembolic pulmonary hypertension or pulmonary thromboendarterectomy."
19. December 9, 2014: Added sildenafil oral suspension to Medical Necessity Guidelines. Removed authorization of coverage of Viagra tablets when the dosage for sildenafil exceeds 20mg three times a day.

20. November 10, 2015: No changes.
21. January 1, 2016: Administrative change to rebranded template.
22. July 12, 2016: Added Uptravi (selexipag) to Medical Necessity Guidelines.
23. April 11, 2017: Administrative update, Adding Tufts Health RITogether to the template.
24. July 11, 2017: No changes.
25. January 9, 2018: Added Tracleer (bosentan) tablets for oral suspension to the Medical Necessity Guideline.
26. January 8, 2019: No changes.
27. January 14, 2020: No changes.
28. February 9, 2020: For Pulmonary Arterial Hypertension, removed "The pulmonary hypertension has progressed despite surgical treatment and/or maximal medical treatment of the underlying condition" and "The medication used for treatment is consistent with its Food and Drug Administration (FDA) approved functional class." Administrative updates to format of Medical Necessity Guideline and based on generic availability of Pulmonary Hypertension Medications, added the following Limitation to be line with current coverage of multi-source brands "The plan does not cover the following brand medications: Adcirca, Letairis, Revatio, and Tracleer tablets. Refer to the Pharmacy Medical Necessity Guidelines for non-covered medications." Effective February 15, 2021, Medical Necessity Guideline applies to Tufts Health Together and Tufts Health RITogether. Effective July 1, 2021, added the following Limitation "Coverage of sildenafil and tadalafil formulations that do not have an FDA-approved indication for pulmonary hypertension will not be authorized for the treatment of pulmonary hypertension."

BACKGROUND, PRODUCT AND DISCLAIMER INFORMATION

Pharmacy Medical Necessity Guidelines have been developed for determining coverage for plan benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. The plan makes coverage decisions on a case-by-case basis considering the individual member's health care needs. Pharmacy Medical Necessity Guidelines are developed for selected therapeutic classes or drugs found to be safe, but proven to be effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in the service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. The plan revises and updates Pharmacy Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Pharmacy Medical Necessity Guideline and a self-insured Member's benefit document, the provisions of the benefit document will govern.

Treating providers are solely responsible for the medical advice and treatment of members. The use of this policy is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to member eligibility and benefits on the date of service, coordination of benefits, referral/authorization and utilization management guidelines when applicable, and adherence to plan policies and procedures and claims editing logic.

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