PHARMACY COVERAGE GUIDELINES
SECTION: DRUGS

ORIGINAL EFFECTIVE DATE: 7/16/15
LAST REVIEW DATE: 3/15/18
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ARCHIVE DATE:

ENDOTHELIN RECEPTOR ANTAGONISTS: LETAIRIS® (ambrisentan) oral tablet OPSUMIT® (macitentan) oral tablet TRACLEER® (bosentan) oral tablet

Coverage for services, procedures, medical devices and drugs are dependent upon benefit eligibility as outlined in the member's specific benefit plan. This Pharmacy Coverage Guideline must be read in its entirety to determine coverage eligibility, if any.

This Pharmacy Coverage Guideline provides information related to coverage determinations only and does not imply that a service or treatment is clinically appropriate or inappropriate. The provider and the member are responsible for all decisions regarding the appropriateness of care. Providers should provide BCBSAZ complete medical rationale when requesting any exceptions to these guidelines.

The section identified as "Description" defines or describes a service, procedure, medical device or drug and is in no way intended as a statement of medical necessity and/or coverage.

The section identified as "Criteria" defines criteria to determine whether a service, procedure, medical device or drug is considered medically necessary or experimental or investigational.

State or federal mandates, e.g., FEP program, may dictate that any drug, device or biological product approved by the U.S. Food and Drug Administration (FDA) may not be considered experimental or investigational and thus the drug, device or biological product may be assessed only on the basis of medical necessity.

Pharmacy Coverage Guidelines are subject to change as new information becomes available.

For purposes of this Pharmacy Coverage Guideline, the terms "experimental" and "investigational" are considered to be interchangeable.

This Pharmacy Coverage Guideline does not apply to FEP or other states' Blues Plans.

Information about medications that require precertification is available at www.azblue.com/pharmacy.

Some large (100+) benefit plan groups may customize certain benefits, including adding or deleting precertification requirements.

All applicable benefit plan provisions apply, e.g., waiting periods, limitations, exclusions, waivers and benefit maximums.

Precertification for medication(s) or product(s) indicated in this guideline requires completion of the request form in its entirety with the chart notes as documentation. All requested data must be provided. Once completed the form must be signed by the prescribing provider and faxed back to BCBSAZ Pharmacy Management at (602)

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864-3126 or emailed to Pharmacyprecert@azblue.com. **Incomplete forms or forms without the chart notes will be returned.**

Description:

Letairis (ambrisentan), Tracleer (bosentan), and Opsumit (macitentan) are endothelin receptor antagonists (ERA) indicated for the treatment of pulmonary arterial hypertension (PAH) World Health Organization (WHO) Group I. Goals of treatment are to improve exercise ability, decrease clinical worsening, and to delay disease progression. Disease progression includes: death, initiation of intravenous (IV) or subcutaneous prostanoids, or clinical worsening (measured as decreased 6-minute walk distance, worsened symptoms and need for additional treatment).

Studies establishing effectiveness of Letairis (ambrisentan) included predominantly patients with New York Heart Association (NYHA) Functional Class II-III symptoms whose etiologies included idiopathic or heritable PAH and PAH associated with connective tissue diseases. Studies establishing effectiveness of Tracleer (bosentan) included predominantly patients with NYHA Functional Class II-IV symptoms whose etiologies of PAH were idiopathic or heritable, associated with connective tissue diseases, and associated with congenital heart disease with left-to-right shunts. Effectiveness of Opsumit (macitentan) was established in PAH patients with predominantly NYHA Functional Class II-III symptoms and etiologies of idiopathic and heritable, caused by connective tissue disorders, and caused by congenital heart disease with repaired shunts.

Pulmonary hypertension (PH) may be described by restricted or reduced blood flow through the pulmonary artery, pulmonary vein, or pulmonary capillaries, leading to complaints of shortness of breath, dizziness, fainting, fatigue, chest pain, palpitations, leg swelling and other symptoms. PH is a severe progressive disease with markedly decreased exercise tolerance, heart failure and ultimately death. The rate of progression is highly variable.

PH may be categorized, using the WHO scheme, into five classes or groups based on etiology and may be further characterized using the NYHA Functional Class system modified for PH that is based on activity level and symptoms in an attempt to classify severity of disease clinically. It should be noted that while together all groups are called pulmonary hypertension, WHO Group 1 is called PAH and WHO Groups 2 through 5 are called PH. Other factors are also used to determine an individual's risk category and assessment of prognosis. NYHA Functional Class I are those individuals least affected by their disease while those in WHO Functional Class IV are most affected.

PAH is placed in WHO Group 1 and includes a large number of etiologies. It is important to distinguish PAH from other types of PH as PH from other causes is thought to differ pathophysiologically from PAH and may be managed differently.

The pathogenesis of PAH (WHO Group 1) is complex and incompletely understood; it is thought to involve an imbalance between vasoconstriction, vasodilation, and abnormal cellular proliferation. It includes genetic, inflammatory, and environmental factors that alter vascular structure and function in smooth muscle, endothelial cells, and adventitia. Included in this complexity are endothelial dysfunction (favoring vasoconstriction,

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thrombosis, and mitogenesis); increased levels of thromboxane A2, endothelin-1 (ET-1), and serotonin (5HT) which stimulate vasoconstriction, cell proliferation, and thrombosis; decreased levels of prostacyclin, nitric oxide, and vasoactive intestinal peptide (VIP) which favor vasoconstriction, cell proliferation, and thrombosis; and low levels of other mediators such as vascular endothelial growth factor (VGEF). VEGF is a signal protein that stimulates creation of new blood vessels which restore oxygen supply to tissues when blood flow is inadequate.

The pathogenesis of pulmonary hypertension from left heart disease (WHO Group 2) is completely different. There is no obstruction to blood flow in the lungs. Instead, the left heart fails to pump blood efficiently, leading to pooling of blood, causing pulmonary edema and pleural effusions. In hypoxic pulmonary hypertension (WHO Group 3), low levels of oxygen are thought to cause vasoconstriction of pulmonary arteries. In chronic thromboembolic pulmonary hypertension (CTEPH or WHO Group 4), the blood vessels are blocked or narrowed with blood clots. These last two groups also share some similar pathophysiology as seen in PAH (WHO Group 1).

A baseline assessment to determine PAH severity is performed before initiating therapy. Therapy should not be administered unless a diagnostic right heart catheterization (RHC) and extensive investigations for the etiology of PH have been performed. This assessment includes the following three key measures:

1. Functional impairment: This is determined by measuring exercising capacity and determining WHO or NYHA Functional Class.

2. Hemodynamic derangement: The diagnosis of PH can be suspected based on echocardiography. However, a RHC is performed to accurately measure hemodynamic parameters and confirm PAH. Individuals with PAH typically undergo an invasive hemodynamic assessment and an acute vasoreactivity test before the initiation of advanced therapy. The hemodynamic definition of PAH is a mean pulmonary artery pressure greater than 25 mm Hg at rest. A pulmonary arterial wedge pressure or left ventricular end-diastolic pressure of less than 15 mm Hg is needed to exclude WHO Group 2 PH (due to left heart disease). PAH is also supported by increased pulmonary vascular resistance and transpulmonary gradient.

3. Acute vasoreactivity test: The test involves administration of a short-acting vasodilator, then measuring hemodynamic response with a right heart catheter. An acute vasoreactivity test is considered positive if mean pulmonary artery pressure decreases by at least 10 mm Hg and to a value less than 40 mm Hg, with an increased or no change in cardiac output and a minimally reduced or no change in systemic blood pressure.

ET-1 is a potent autocrine and paracrine peptide. There are two receptor subtypes for ET-1 binding, ETA and ETB. Binding of ET-1 to its receptors is thought to play a role in vasoconstriction, fibrosis, proliferation, hypertrophy, and inflammation. The primary actions of stimulating ETA receptor are vasoconstriction and cell proliferation, while the predominant actions of stimulating ETB receptor are vasodilation, antiproliferation, and clearance of ET-1. In PAH (WHO Group 1), the local endothelin system is up-regulated and is involved in vascular hypertrophy and in organ damage; it is important to note that other factors besides the ET system are involved in the overall pathogenesis of PAH (WHO Group 1).

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All female patients using Letairis, Opsumit, or Tracleer are subject to a Risk Evaluation and Mitigation Strategies (REMS) program that requires provider, patient, and dispensing pharmacy be enrolled into the program. Only providers and pharmacies enrolled into the REMS may prescribe and dispense the drug, respectively, to individuals who are also in the program. A REMS program attempts to manage known or potentially serious risks associated with a drug product and is required by the Food and Drug Administration (FDA) for some drugs to ensure that the benefits of a drug outweigh its risks.

Definitions:

Pulmonary Hypertension Association (PHA)
The largest and oldest pulmonary hypertension (PH) association in the world. PHA is a community-based nonprofit support, education, advocacy and awareness association for PH.

Pulmonary Hypertension Care Centers (PHCC)
Center of Comprehensive Care (CCC):
A PHA accredited highly organized, full-time PH center that proficiently evaluates individuals with PH based on published evidence-based guidelines and provides expert treatment of individuals with PAH with all of the FDA-approved therapies. CCC also make important contributions to PH research and education.

Regional Clinical Program (RCP):
A PHA accredited center that proficiently evaluates individuals with PH based on published evidence-based guidelines and provides expert treatment of individuals with PAH with all non-parenteral therapies. A RCP must collaborate with its regional CCC by referring individuals that may benefit from opportunities unavailable at the RCP, including the initiation of advanced parenteral therapies and participation in clinical research protocols.

WHO Group, classification of Pulmonary Hypertension (PH)
- WHO Group 1 - Pulmonary arterial hypertension (PAH)
  - Idiopathic (IPAH)
  - Heritable / Familial
    - Activin receptor-like kinase (ALK1), endoglin (with or without hereditary hemorrhagic telangiectasia)
    - Bone Morphogenic Protein Receptor type II (BMPR)
    - Unknown
  - Drug- and toxin-induced
  - Associated with (APAH):
    - Congenital heart diseases – systemic to pulmonary shunts
    - Connective tissue disease
    - HIV infection
    - Portal hypertension
    - Schistosomiasis

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- Persistent pulmonary hypertension of the newborn
- Associated with significant venous or capillary involvement
  - Pulmonary capillary hemangiomatosis (PCH)
  - Pulmonary veno-occlusive disease (PVOD)

- WHO Group 2 - Pulmonary hypertension owing to left heart disease
  - Left-sided arterial or ventricular heart disease
  - Left-sided valvular heart disease
  - Diastolic dysfunction
  - Systolic dysfunction

- WHO Group 3 - Pulmonary hypertension owing to lung disease and/or hypoxia
  - Alveolar hypoventilation disorders
  - Chronic exposure to high altitude
  - Chronic obstructive pulmonary disease
  - Developmental abnormalities
  - Interstitial lung disease
  - Other pulmonary diseases with mixed restrictive and obstructive pattern
  - Sleep-disordered breathing

- WHO Group 4 - Pulmonary hypertension due to Chronic thromboembolic pulmonary hypertension (CTEPH)
  - Non-thrombotic pulmonary embolism (tumor, parasites, foreign material)
  - Thromboembolic obstruction of distal pulmonary arteries
  - Thromboembolic obstruction of proximal pulmonary arteries

- WHO Group 5 - Pulmonary hypertension with unclear multifactorial mechanisms
  - Hematologic diseases: chronic hemolytic anemia (including sickle cell disease)myeloproliferative disease, splenectomy
  - Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid diseases
  - Systemic diseases: sarcoidosis, pulmonary Langerhans cell histiocytosis: lymphangioleiomyomatosis, neurofibromatosis, vasculitis
  - Others: tumor obstruction, fibrosing mediastinitis, chronic renal failure on dialysis, compression of pulmonary vessels, Hemoglobinopathies, Hereditary hemorrhagic telangiectasia

WHO Functional Class (modified New York Heart Association (NYHA) for PH)

Functional Class I

No limitation in physical activity; ordinary physical activity does not cause dyspnea or fatigue, chest pain or near syncope

Functional Class II

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Slight limitations in physical activity; ordinary physical activity produces dyspnea, fatigue, chest pain, or near-syncope; comfortable with no symptoms at rest

Functional Class III
Marked limitation of physical activity; less than ordinary physical activity produces dyspnea, fatigue, chest pain, or near-syncope; comfortable with no symptoms at rest

Functional Class IV
Unable to perform any physical activity without symptoms; dyspnea and/or fatigue present at rest; discomfort increased by any physical activity

Therapeutic classes of drugs used to treat pulmonary hypertension:

Calcium Channel Blockers – used only in a very select group of individuals
- Dihydropyridine class preferred

Endothelin receptor antagonists – bind to receptors in endothelium and vascular smooth muscle
- Ambrisentan (Letairis) – oral
- Bosentan (Tracleer) – oral
- Macitentan (Opsumit) – oral

Phosphodiesterase type 5 inhibitors – inhibit Phospodiesterase type-5 to increased cAMP
- Sildenafil (Revatio, and generics) – oral (generics available) and IV (available as brand Revatio)
- Tadalafil (Adcirca) – oral

Prostanoids – direct vasodilation of pulmonary & systemic arterial vascular beds, inhibit platelet aggregation
- Epoprostenol (Flolan, Veletri, generics) – continuous IV
- Iloprost (Ventavis) – inhaled delivery system
- Treprostinil
  - Orenitram ER – oral
  - Remodulin – can be SQ or IV
  - Tyvaso – inhaled delivery system

Soluble Guanylate Cyclase Stimulators – stimulate Nitric Oxide cGMP pathway to increase cGMP
- Riociguat (Adempas) – oral

Selective prostacyclin receptor (IP receptor) agonist
- Selexipag (Uptravi) – oral

Letairis (ambrisentan)
Opsumit (macitentan)
Tracleer (bosentan)

Medication class:
Endothelin Receptor Antagonist; Vasodilator

FDA-approved indication(s):
- Letairis (ambrisentan)
  - Treatment of pulmonary arterial hypertension (PAH) (World Health Organization [WHO] group 1) to improve exercise ability and delay clinical worsening
  - Treatment of PAH (WHO group 1) 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 World Health Organization functional class (WHO-FC) II-III symptoms
- Opsumit (macitentan)
  - Treatment of PAH (WHO group 1) to delay disease progression
  - Effectiveness was established in a long-term study in PAH patients with predominantly WHO-FC II-III symptoms
- Tracleer (bosentan)
  - Treatment of PAH (WHO group 1), in adults with WHO-FC II-IV symptoms to improve exercise ability and to decrease clinical deterioration;
  - Treatment of PAH (WHO Group 1) in pediatric patients ≥ 3 years of age with idiopathic or congenital PAH to improve pulmonary vascular resistance (PVR), resulting in an improvement in exercise ability

Recommended Dose:
- Letairis: Initial dose is 5 mg once daily, titrated at 4 week intervals, with or without tadalafil
- Opsumit: 10 mg once daily
- Tracleer:
  - Patients > 12 years of age: initiate at 62.5 mg twice daily
    - Patients weighing > 40 kg the dose can be increased to 125 mg twice daily after 4 weeks
    - Patients weighing < 40 kg the dose is maintained at 62.5 mg twice daily
  - Patients ≤ 12 years of age:
    - ≥ 4-8 kg: 16 mg twice daily initial and maintenance
    - > 8-16 kg: 32 mg twice daily initial and maintenance
    - > 16-24 kg: 48 mg twice daily initial and maintenance
    - > 24-40 kg: 64 mg twice daily initial and maintenance

Maximum dosage
- Letairis: not stated
- Opsumit: 10 mg once daily

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- Tracleer: 125 mg twice daily

Available Dosage Forms:
- Letairis: 5 mg and 10 mg tabs
- Opsumit: 10 mg tabs
- Tracleer: 32 mg tab for oral suspension, 62.5 mg, 125 mg tabs
    - The tabs for suspension contain 1.87 mg phenylalanine per tab

Warnings, Precautions, and other Clinical Information:
- Letairis (ambrisentan)
    - Females can only receive Letairis through the REMS program that requires a pregnancy test prior, during, and after treatment; woman of child bearing potential must not get pregnant and must use effective contraception; Letairis is contraindicated in females who are pregnant
    - Discontinue if signs of pulmonary edema occur & consider the possibility of pulmonary veno-occlusive disease (PVOD)
    - Use is not recommended in patients with severe anemia, if a significant decrease in hemoglobin occurs and other causes have been excluded, consider stopping Letairis
    - If fluid retention develops, determine if caused by Letairis or heart failure, consider stopping Letairis if it is the cause of the fluid retention
    - There is no information on the use of Letairis in severe renal impairment or individual on dialysis
    - Letairis is not recommended in patients with moderate to severe hepatic impairment
    - Discontinue Letairis if elevations of liver aminotransferases are > 5x ULN or elevations are accompanied by bilirubin > 2x ULN or signs and symptoms of liver dysfunction and other causes are excluded
    - Woman who is breast feeding an infant or child should stop breast feeding
    - When used with cyclosporine, Letairis dose should be limited to 5 mg once daily
    - Letairis should not be used in idiopathic pulmonary fibrosis with or without pulmonary hypertension (WHO Group 3) due to greater risk of disease progression or death
    - Letairis is not indicated for treatment of digital ulcers, systemic sclerosis, Eisenmenger syndrome, or Raynaud phenomenon

- Opsumit (macitentan)
    - Females can only receive Opsumit through the REMS program that requires a pregnancy test prior, during, and after treatment; woman of child bearing potential must not get pregnant and must use effective contraception; Opsumit is contraindicated in females who are pregnant
    - Discontinue if signs of pulmonary edema occur & consider the possibility of pulmonary veno-occlusive disease (PVOD)
    - Hepatotoxicity has been reported, if clinically relevant aminotransferase elevations occur, or if elevations are accompanied by an increase in bilirubin > 2 x ULN, or by clinical symptoms of hepatotoxicity, discontinue Opsumit
    - If fluid retention develops, determine if caused by Opsumit or heart failure, consider stopping Opsumit if it is the cause of the fluid retention
    - Use is not recommended in individual with severe anemia

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- Woman who is breast feeding an infant or child should stop breast feeding
- Avoid use with strong CYP3A4 inducers
- Avoid use with strong CYP3A4 inhibitors
- Hepatic metabolism of Opsumit results in a metabolite that contributes about 40% of the total pharmacologic activity
- Opsumit is not indicated for treatment of idiopathic pulmonary fibrosis, digital ulcers, systemic sclerosis, Eisenmenger syndrome, or Raynaud phenomenon

- Tracleer (bosentan)
  - Females can only receive Tracleer through the REMS program that requires a pregnancy test prior, during, and after treatment; woman of child bearing potential must not get pregnant and must use effective contraception; Tracleer is contraindicated in females who are pregnant
  - Avoid use in patients with elevated aminotransferases > 3x ULN
  - Reduce the dose of Tracleer in patients developing aminotransferase elevations > 3x ULN
  - Discontinue if aminotransferase elevations are accompanied by signs or symptoms of liver dysfunction or injury or increase in bilirubin $\geq$ 2x ULN
  - Permanently discontinue if aminotransferase elevations are > 8x ULN
  - Avoid use in patients with moderate to severe hepatic impairment (Child-Pugh Class B and C)
  - If fluid retention develops, determine if caused by Tracleer or heart failure, consider stopping Tracleer if it is the cause of the fluid retention
  - Discontinue if signs of pulmonary edema occur & consider the possibility of pulmonary veno-occlusive disease (PVOD)
  - Use with both a moderate or strong inhibitor of CYP3A4 (amprenavir, erythromycin, fluconazole, diltiazem, itraconazole, or ketoconazole) and a CYP2C9 inhibitor (fluconazole, amiodarone) is not recommended
  - Estrogen and progestin levels are reduced by Tracleer, hormonal contraceptives may be less effective
  - Woman who is breast feeding an infant or child should stop breast feeding
  - The absolute bioavailability is about 50%
  - Tracleer appears to undergo auto-induction
  - Tracleer is not indicated for treatment of idiopathic pulmonary fibrosis, digital ulcers, systemic sclerosis, Eisenmenger syndrome, or Raynaud phenomenon

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Criteria:

- **Criteria for initial therapy:** Letairis (ambrisentan), Opsumit (macitentan), and Tracleer (bosentan) is considered ***medically necessary*** and will be approved when **ALL** of the following criteria are met:

1. Prescriber is a pulmonologist specializing in treating patients with PAH

2. Individual is 18 years of age or older

3. **Letairis and Opsumit**: A confirmed diagnosis of pulmonary arterial hypertension (PAH, WHO Group 1) with Functional Class II or III symptoms (WHO Group and Functional Class category must be submitted with request)
   Tracleer: A confirmed diagnosis of pulmonary arterial hypertension (PAH, WHO Group 1) with Functional Class II-IV symptoms (WHO Group and Functional Class category must be submitted with request)

4. Right heart catheterization documents **ALL** of the following:
   - Mean pulmonary artery pressure > 25 mm Hg at rest
   - Pulmonary arterial wedge pressure ≤ 15 mm Hg
   - Pulmonary vascular resistance > 3 Wood units

5. Vasoreactivity testing with negative results

6. Chronic lung diseases and other causes of hypoxemia are mild or absent

7. Venous thromboembolic disease is absent

8. Individual does not have other disorders, that would put them into WHO group 5 PH, including:
   - systemic disorders (e.g., sarcoidosis),
   - hematologic disorders (e.g., myeloproliferative diseases), and
   - metabolic disorders (e.g., glycogen storage disease)

9. Individual has failure, intolerance or contraindication to generic oral sildenafil

10. **Opsumit and Tracleer**: Individual has failure, intolerance or contraindication to Letairis (ambrisentan)

11. There are **NO** contraindications.
    - Contraindications include:
      - For Letairis:
        - Idiopathic pulmonary fibrosis, with or without pulmonary hypertension (WHO Group 3)
      - For Tracleer:
        - Use with Cyclosporine
        - Use with Glyburide
        - Hypersensitivity to bosentan or any component of the product

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Initial approval duration: 12 months

- **Criteria for continuation of coverage (renewal request):** Letairis (ambrisentan), Opsumit (macitentan), and Tracleer (bosentan) is considered ***medically necessary*** and will be approved when **ALL** of the following criteria are met:

1. Individual continues to see a pulmonologist specializing in treating patients with PAH

2. Monitor patient for disease status:
   - Reduction in hospitalizations for PAH
   - 6-minute walking distance (6MWD)
   - Functional Class symptoms

3. Individual has been adherent with the medication

4. Individual has not developed any contraindications or other significant level 4 drug effects that may exclude continued use
   - Contraindications as listed in the criteria for initial therapy section
   - Significant adverse effect such as:
     - Pulmonary veno-occlusive disease due to Letairis
     - Severe anemia
     - Fluid retention requiring hospitalization for decompensating heart failure
     - Liver toxicity

5. There are no significant interacting drugs

Renewal duration: 12 months

Resources:

Letairis. Package Insert. Revised by manufacturer October 2015. Accessed 02-28-17, 02-23-18.

Letairis. Package Insert. Revised by manufacturer May 2014. Accessed 06-02-2015. Viewed on 07-01-2015.

Opsumit. Package Insert. Revised by manufacturer March 2017. Accessed 02-23-18.

Opsumit. Package Insert. Revised by manufacturer October 2016. Accessed 02-28-17.

Opsumit. Package Insert. Revised by manufacturer April 2015. Accessed 06-02-2015. Viewed on 07-01-2015.

Tracleer. Package Insert. Revised by manufacturer August 2017. Accessed 02-28-18.

Tracleer. Package Insert. Revised by manufacturer October 2016. Accessed 03-03-17.

Tracleer. Package Insert. Revised by manufacturer October 2012. Accessed 06-02-2015. Viewed on 07-01-2015.

Goldman L, Hashimoto B, Cook EF, Loscalzo A: Comparative reproducibility and validity of systems for assessing cardiovascular functional class: advantages of a new specific activity scale. Circulation 1981; 64 (6):1227-1234.

Badesch DB, Abman SH, Simonneau G, et al.: Medical therapy for pulmonary arterial hypertension: Updating ACCP evidence-based clinical practice guidelines. Chest 2007; 131 (6):1917-1928.

McLaughlin VV, Archer SL, Badesch DB, et al.: ACCF/AHA 2009 Expert Consensus Document on Pulmonary Hypertension: A Report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association. Circulation 2009; 119:2250-2294.

Taichman DB, Ornelas J, Chung L, et al.: Pharmacologic Therapy for Pulmonary Arterial Hypertension in Adults: CHEST Guideline and Expert Panel Report. Chest 2014; 146(2):449-475.

UpToDate: Classification and prognosis of pulmonary hypertension in adults. Current through Jan 2018. https://www-uptodate-com.mwu.idm.oclc.org/contents/classification-and-prognosis-of-pulmonary-hypertension-in-adults?search=pulmonary%20hypertension&source=search_result&selectedTitle=1~150&usage_type=default&display_rank=1

UpToDate: Clinical feature and diagnosis of pulmonary hypertension in adults. Current through Jan 2018. https://www-uptodate-com.mwu.idm.oclc.org/contents/clinical-features-and-diagnosis-of-pulmonary-hypertension-in-adults?search=pulmonary%20hypertension&source=search_result&selectedTitle=2~150&usage_type=default&display_rank=2

UpToDate: Treatment of pulmonary hypertension in adults. Current through Jan 2018. https://www-uptodate-com.mwu.idm.oclc.org/contents/treatment-of-pulmonary-hypertension-in-adults?search=pulmonary%20hypertension&source=search_result&selectedTitle=3~150&usage_type=default&display_rank=3#H5

BlueCross BlueShield of Arizona

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Fax completed prior authorization request form to 602-864-3126 or email to pharmacyprecert@azblue.com.
Call 866-325-1794 to check the status of a request.
All requested data must be provided. **Incomplete forms or forms without the chart notes will be returned.**
Pharmacy Coverage Guidelines are available at www.azblue.com/pharmacy.

Pharmacy Prior Authorization Request Form

Do not copy for future use. Forms are updated frequently.
REQUIRED: Office notes, labs, and medical testing relevant to the request that show medical justification are required.

Member Information

Member Name (first & last):	Date of Birth:	Gender:	BCBSAZ ID#:
Address:	City:	State:	Zip Code:

Prescribing Provider Information

Provider Name (first & last):	Specialty:	NPI#:	DEA#:
Office Address:	City:	State:	Zip Code:
Office Contact:	Office Phone:	Office Fax:	

Dispensing Pharmacy Information

Pharmacy Name:	Pharmacy Phone:	Pharmacy Fax:

Requested Medication Information

Medication Name:	Strength:	Dosage Form:
Directions for Use:	Quantity: / Refills:	Duration of Therapy/Use:

☐ Check if requesting **brand** only ☐ Check if requesting **generic**

☐ Check if requesting continuation of therapy (prior authorization approved by BCBSAZ expired)

Turn-Around Time For Review

☐ Standard ☐ Urgent. Sign here: _____ ☐ Exigent (requires prescriber to include a written statement)

Clinical Information

1. **What is the diagnosis? Please specify below.**

   ICD-10 Code: _____ **Diagnosis Description:** _____

2. ☐ Yes ☐ No **Was this medication started on a recent hospital discharge or emergency room visit?**

3. ☐ Yes ☐ No **There is absence of ALL contraindications.**

4. **What medication(s) has the individual tried and failed for this diagnosis? Please specify below.**
Important note: Samples provided by the provider are not accepted as continuation of therapy or as an adequate trial and failure.

Medication Name, Strength, Frequency	Dates started and stopped or Approximate Duration	Describe response, reason for failure, or allergy

5. **Are there any supporting labs or test results? Please specify below.**

Date	Test	Value

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Pharmacy Prior Authorization Request Form

6. Is there any additional information the prescribing provider feels is important to this review? Please specify below.
For example, explain the negative impact on medical condition, safety issue, reason formulary agent is not suitable to a specific medical condition, expected adverse clinical outcome from use of formulary agent, or reason different dosage form or dose is needed.

Signature affirms that information given on this form is true and accurate and reflects office notes

Prescribing Provider's Signature: Date:

Please note: Some medications may require completion of a drug-specific request form.

Incomplete forms or forms without the chart notes will be returned.

Office notes, labs, and medical testing relevant to the request that show medical justification are required.