Pulmonary Arterial Hypertension

Revatio® (sildenafil), Ventavis® (iloprost), Tracleer® (bosentan), Letairis® (ambrisentan), Opsumit®(macitentan), Adcirca® (tadalafil), Tyvaso®/Tyvaso DPI™ (treprostinil), Orenitram® (treprostinil), Adempas® (riociguat), Uptravi® (selexipag), Tadliq® (tadalafil), Liqrev® (sildenafil)

(Oral/Inhalation)

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I. Length of Authorization

Coverage will be provided for 6 months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

Drug	Drug strength/formulation	# of Units	Per # of Days	Units/Day
Orenitram	All strengths (0.125 mg, 0.25 mg, 1 mg, 2.5 mg, 5 mg tablets)	90	30	3
	Titration Kit (all strengths)	1	N/A	N/A
Tyvaso	1.74 mg/2.9 mL ampule	28	28	1
Tyvaso DPI	Titration Kit (all strengths)	1	N/A	N/A
	Maintenance Kit (16, 32, 48, & 64 mcg)	1	28	N/A
	Maintenance Kit (32 mcg and 48 mcg)	1	28	N/A
	Institutional Kit (16, 32, 48, & 64 mcg)	1	4	N/A
	Institutional Kit (32 mcg and 48 mcg)	1	4	N/A
Ventavis	10 mcg/mL ampule 20 mcg/mL ampule	270 amps	30	9
Letairis	5 mg tablet 10 mg tablet	30	30	1
Tracleer	62.5 mg tablet 125 mg tablet	60	30	2

Tracleer	32 mg tablet for oral suspension	120	30	4
Opsumit	10 mg tablet	30	30	1
Adempas	All strengths (0.5, 1, 1.5, 2, & 2.5 mg tablets)	90	30	3
Uptravi	All strengths (200, 400, 600, 800, 1000, 1200, 1400, & 1600 mcg tablets)	60	30	2
	Titration Pack	1	N/A	N/A
Revatio	20 mg tablet	360	30	12
	10 mg/mL oral suspension	720 mL	30	24
Sildenafil	20 mg tablet	360	30	12
	10 mg/mL oral suspension	720 mL	30	24
Adcirca	20 mg tablet	60	30	2
Tadalafil	20 mg tablet	60	30	2
Tadliq	20 mg/5 mL oral suspension	300 ml	30	10
Liqrev	10 mg/mL oral suspension	180 ml	30	6

B. Max Units (per dose and over time) [HCPCS Unit]:

Tyvaso inhalation solution

- 1 billable unit per day

Ventavis

- 9 billable units per day

III. Initial Approval Criteria [1-13]

Coverage is provided in the following conditions:

- Patient is at least 18 years or age (unless otherwise specified); **OR**
    - Patient is at least 3 years of age for Tracleer; **OR**
    - Patient is at least 1 year of age for Revatio; **AND**
- Patients of reproductive potential have had a negative pregnancy test prior to start of therapy (**Opsumit, Letairis, Tracleer, and Adempas ONLY**); **AND**

Universal Criteria [1-13]

- Patient is NOT receiving concurrent treatment with organic nitrates (e.g., isosorbide mononitrate, isosorbide dinitrate, nitroglycerin) (**Revatio, Adcirca, Adempas, Liqrev, and Tadliq ONLY**); **AND**
- Patient does not have severe hepatic impairment (Child Pugh Class C) (**Orenitram ONLY**); **AND**
- Patient does not have Idiopathic Pulmonary Fibrosis (IPF), including IPF patients with pulmonary hypertension (WHO Group 3) (**Letairis, ONLY**); **AND**
- Patient does not have pulmonary hypertension associated with idiopathic interstitial pneumonias (PH-IIP) (**Adempas ONLY**); **AND**
- Both patient and prescriber are enrolled in the manufacturer's REMS program (**Opsumit, Letairis, Tracleer, and Adempas ONLY**); **AND**

Pulmonary Arterial Hypertension (PAH) † Φ [1-13,38,40,43]

- Diagnosis confirmed by documented right heart catheterization with ALL of the following:
    - Mean pulmonary arterial pressure (mPAP) > 20 mmHg; **AND**
    - Pulmonary arterial wedge pressure (PAWP) ≤ 15 mmHg; **AND**
    - Pulmonary vascular resistance (PVR) ≥ 3 wood units (240 dynes-sec/cm^5); **AND**
- Baseline assessment of 6-minute walk distance (6MWD), brain natriuretic peptide (BNP) plasma levels, and/or B-type natriuretic peptide plasma levels (NT-proBNP); **AND**
- Diagnosed with pulmonary arterial hypertension and classified as WHO Group 1 (See below for description of WHO classification for pulmonary hypertension); **AND**
    - Pediatric patients are diagnosed with idiopathic or congenital pulmonary arterial hypertension (**Tracleer ONLY**); **AND**
- Designated as New York Heart Association (NYHA) or World Health Organization (WHO) functional class II-IV (See below for description of functional classes); **AND**
- Patient is treatment-naïve to PAH-specific pharmacotherapy **§**; **AND**
    - Patient is Functional Class II or Functional Class III without evidence of rapid disease progression or poor prognosis; **AND**
        - Patient had an inadequate response to calcium channel blocker therapy or is not a candidate for treatment with a calcium channel blocker (i.e., negative results for acute vasoreactivity, right ventricular failure, or contraindication to calcium channel blocker); **AND**
            - Patient will be treated with a combination of Letairis and Adcirca; **OR**
            - Patient is unwilling or unable to tolerate combination therapy and will receive monotherapy with an endothelial-receptor antagonist (ERA) **§**, phosphodiesterase-5 inhibitor (PDE5i) **§**, or Adempas; **OR**
    - Patient is Functional Class IV; **AND**
        - Patient is unwilling or unable to manage intravenous or subcutaneous prostacyclin analog therapy **§**; **AND**
        - Patient will be treated with an inhaled prostacyclin analog in combination with an oral PDE5i and an ERA **§**; **OR**
- Patient is Functional Class III or IV and had an inadequate clinical response **‡** (see criteria below) to monotherapy and will be adding a second class of PAH therapy as one of the following (see PAH pharmacotherapy table below **§**):
    - Adding an inhaled prostacyclin analog to an ERA or a PDE5i
    - Adding Revatio to an intravenous epoprostenol
    - Adding Adempas to Tracleer, Letairis, or an inhaled prostacyclin analog
    - Adding Opsumit to a PDE5i or an inhaled prostacyclin analog; **OR**
- Patient is Functional Class III or IV with an inadequate clinical response **‡** (see criteria below) to two classes of PAH pharmacotherapy and will be adding a third class of PAH therapy (see PAH pharmacotherapy table below **§**); **OR**

- Patient is currently on Letairis with stable or symptomatic disease and will add Adcirca; **OR**
- Patient is transitioning from Remodulin to Orenitram and using Remodulin (treprostinil) and Orenitram (treprostinil) concurrently

Chronic-Thromboembolic Pulmonary Hypertension (Adempas only) † Φ [12,35,43]

- Baseline 6-minute walk test (6MWD) performed; **AND**
- Must not be used in combination with phosphodiesterase-5 inhibitors (PDE5i), prostacyclin analogs, or endothelial-receptor antagonists (ERAs) **§**; **AND**
- Patient does not have left heart disease or lung disease (e.g., COPD, interstitial lung disease, combined pulmonary fibrosis and emphysema [CPFE], etc.); **AND**
- Diagnosis of chronic pulmonary thromboembolic hypertension (CTEPH) confirmed after at least 3 months of effective anticoagulation with ALL of the following:
    - Mean pulmonary arterial pressure (mPAP) ≥ 25mmHg
    - Pulmonary arterial wedge pressure (PAWP) ≤ 15 mmHg
    - Mismatch perfusion defects and/or specific diagnostic signs for CTEPH as seen on at least two of the following imaging methods: ventilation–perfusion (V/Q) scanning, pulmonary angiography, spiral computed tomography, or magnetic resonance angiography; **AND**
- Diagnosed with CTEPH and classified as WHO Group 4 (See below for description of WHO classification for pulmonary hypertension); **AND**
    - Patient is inoperable for surgery (i.e., pulmonary thromboendarterectomy); **AND**
        - Patient's pulmonary vascular resistance (PVR) >300 dyn*sec*cm^{-5} measured at least 90 days after the start of full anticoagulation; **OR**
    - Patient has recurrent or persisting pulmonary hypertension with pulmonary vascular resistance (PVR) >300 dyn*sec*cm^{-5} measured at least 180 days following pulmonary thromboendarterectomy

Pulmonary Hypertension Associated with Interstitial Lung Disease (PH-ILD) (Tyvaso/Tyvaso DPI only) † [8,9,35,38,43]

- Patient diagnosed with pulmonary hypertension and classified as WHO Group 3 (See below for description of WHO classification for pulmonary hypertension); **AND**
- Diagnosis confirmed by documented right heart catheterization with ALL of the following:
    - Mean pulmonary arterial pressure (mPAP) > 20 mmHg; **AND**
    - Pulmonary arterial wedge pressure (PAWP) ≤ 15 mmHg; **AND**
    - Pulmonary vascular resistance (PVR) ≥ 3 wood units (240 dynes-sec/cm^5); **AND**
- Baseline assessment of 6-minute walk distance (6MWD) and/or B-type natriuretic peptide plasma levels (NT-proBNP)

Pulmonary Hypertension Pharmacotherapy § [1,3,11,12,40]

Class	Drug	Route of Administration
Phosphodiesterase-5 inhibitors (PDE5i)	Revatio (sildenafil) Adcirca (tadalafil) Tadliq (tadalafil) Liqrev (sildenafil)	IV, Oral Oral Oral Oral
Prostacyclin analogs	Flolan (epoprostenol) Veletri (epoprostenol) Orenitram (treprostinil) Remodulin (treprostinil) Tyvaso/Tyvaso DPI (treprostinil) Ventavis (iloprost)	IV IV Oral IV/SC Inhaled Inhaled
Endothelial-receptor antagonists (ERA)	Tracleer (bosentan) Letairis (ambrisentan) Opsumit (macitentan)	Oral Oral Oral
Soluble guanylate cyclase stimulators	Adempas (riociguat) ▪ Must NOT be used in combination with PDE5i (e.g., Revatio, Adcirca, Tadliq, Liqrev) or intravenous prostacyclin analogs (e.g., Flolan, Veletri, Remodulin) ▪ *Subcutaneous administration of Remodulin is allowable with Adempas*	Oral
Prostacyclin receptor agonists	Uptravi (selexipag) ▪ May be used in combination with BOTH a PDE5i AND an ERA	Oral, IV

Inadequate Clinical Response Criteria ‡ [22]

- Inadequate clinical response for patients who were initially in WHO Functional Class II or III:
  - Resulting clinical status defined as stable and not satisfactory; **OR**
  - Resulting clinical status defined as unstable and deteriorating
- Inadequate clinical response for patients who were initially in WHO Functional Class IV:
  - No rapid improvement to WHO Functional Class III or better; **OR**
  - Resulting clinical status defined as stable and not satisfactory

Reference charts [40,41]

WHO Classification of Pulmonary Hypertension (PH):

- Group 1 PAH: Pulmonary arterial hypertension (PAH)
- Group 2 PH: Pulmonary hypertension owing to left heart disease
- Group 3 PH: Pulmonary hypertension owing to lung diseases and/or hypoxia
- Group 4 PH: Chronic thromboembolic pulmonary hypertension (CTEPH)
- Group 5 PH: Pulmonary hypertension with unclear multifactorial mechanisms

New York Heart Association (NYHA) Functional Classification

- Class I: No symptoms with ordinary physical activity. No limitation of physical activity. Comfortable at rest.
- Class II: Symptoms with ordinary physical activity. Slight limitation of physical activity. Comfortable at rest.
- Class III: Symptoms with less than ordinary physical activity. Marked limitation of physical activity.
- Class IV: Symptoms with any physical activity or even at rest. Unable to perform any physical activity.

World Health Organization (WHO) Functional Assessment Classification

– Class I: Patients with PH but 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-heart failure. Dyspnea and/or fatigue may even be present at rest. Discomfort is increased by any physical activity.

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Φ Orphan Drug (applies to all medications included in the policy, except for Revatio)

Note: Clinical review for use in pediatric patients, unless specified above, will occur on a case by case basis

IV. Renewal Criteria [1-13,40,41,43]

Coverage may be renewed based upon the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section III; **AND**
- Disease response as determined by one or more of the following:
  - Progress towards an improvement in WHO functional class status
  - Improvement in right ventricular function (based on echocardiogram or cardiac MRI)
  - Improvement (from baseline) on the 6-minute walk distance (6MWD)
  - Improvement (from baseline) in B-type natriuretic peptide plasma levels (NT-proBNP)
  - Improvement (from baseline) in brain natriuretic peptide (BNP) plasma levels
  - Increase in time to first clinical worsening event (e.g., hospitalization due to worsening of disease, etc.) (**Orenitram and Tyvaso/Tyvaso DPI ONLY**); **AND**

Revatio®, Adcirca®, Liqrev® and Tadliq®

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: pulmonary edema/pulmonary veno-occlusive disease (PVOD), hearing or visual impairment, symptomatic hypotension, epistaxis, and prolonged erection.

Orenitram®

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: worsening PAH symptoms after abrupt discontinuation or large dose reductions and potential for tablets getting lodged in the diverticulum in patients with diverticulosis.

Tyvaso®/Tyvaso DPI™

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: symptomatic hypotension, anticoagulation abnormalities (bleeding), and bronchospasm.

Ventavis®

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: hypotension (systolic BP < 85 mm Hg), pulmonary edema, and bronchospasm.

Tracleer®, Letairis®, and Opsumit®

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: hepatic impairment, fluid retention, pulmonary edema/pulmonary veno-occlusive disease (PVOD), and decreased hemoglobin and hematocrit.

Adempas®

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: symptomatic hypotension, bleeding, and pulmonary edema/pulmonary veno-occlusive disease (PVOD).

Uptravi®

- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: pulmonary edema/pulmonary veno-occlusive disease (PVOD).

V. Dosage/Administration [1-13]

Indication	Dose
Revatio® (oral tablet or suspension)	Adult patients: • Administer 20 mg orally three times a day. Dose may be titrated to a maximum of 80 mg three times a day, if required, based on symptoms and tolerability. Pediatric patients 1 to 17 years of age: • ≤ 20 kg: Administer 10 mg orally 3 times a day • 20 to 45 kg: Administer 20 mg orally 3 times a day • > 45 kg: Administer 20 mg orally three times a day. Dose may be titrated to a maximum of 40 mg three times a day, if required, based on symptoms and tolerability.
Adcirca® (oral tablet)	Administer 40 mg (two 20 mg tablets) orally once daily. (Dividing the dose (40 mg) over the course of the day is not recommended). May be given in combination with Letairis. *Refer to prescribing information for dose adjustments in patients with renal impairment (CrCl ≤ 80 mL/min), hepatic impairment, or in combination with ritonavir.*
Orenitram® (oral tablet)	Starting dose: Administer 0.25 mg by mouth twice daily or 0.125 mg three times daily. Titrate by 0.25 mg or 0.5 mg twice daily or 0.125 mg three times daily, not more than every 3 to 4 days as tolerated. Maximum dose is determined by tolerability. Transitioning from SC/IV routes of treprostinil (Remodulin): • Orenitram total daily dose (mg) = 0.0072 X Remodulin dose (ng/kg/min) X weight (kg)

Indication	Dose
	• Decrease the dose of Remodulin while simultaneously increasing the dose of Orenitram. The Remodulin dose can be reduced up to 30 ng/kg/min per day and dose of Orenitram simultaneously increased up to 6 mg per day (2 mg TID) if tolerated. *Refer to prescribing information for dose adjustments in patients with hepatic impairment or in combination with strong CYP2C8 inhibitors (e.g., gemfibrozil).*
Tyvaso® (inhalation solution)	Inhale 3 breaths (18 mcg) orally via the Tyvaso Inhalation System, 4 times daily. (If 3 breaths aren't tolerated, reduce to 1 or 2 breaths and subsequently increase to 3 breaths as tolerated). Dosage should be increased by an additional 3 breaths per treatment session, 4 times daily at approximately 1- to 2-week intervals to eventually reach 9 (54 mcg) to 12 (72 mcg) breaths per treatment session, administered 4 times daily if tolerated. (1 ampule contains sufficient medication for all 4 treatment sessions in a single day). Max dose of 12 (72 mcg) breaths, 4 times daily.
Tyvaso DPI™ (oral inhalation)	Initial dose: Administer one 16 mcg cartridge via oral inhalation four times daily. Maintenance dose: Increase dosage by an additional 16 mcg per treatment session at approximately 1- to 2-week intervals. The target maintenance dosage is usually 48 mcg to 64 mcg per session. *Refer to prescribing information for dosage transition from Tyvaso® Inhalation Solution to Tyvaso DPI™.*
Ventavis® (oral inhalation)	Initially, inhale 2.5 mcg delivered at the mouthpiece via I-neb AAD system. If well tolerated, the dose may be increased and maintained at 5 mcg administered 6 to 9 times daily, but no more than every 2 hours, during waking hours based on individual need and tolerability. Max daily dose is 45 mcg (5 mcg administered 9 times per day). (1 ampule should be used for each dose; the 20 mcg/mL strength is intended to decrease treatment times in patients who experience extended treatment times)
Letairis® (oral tablet)	Initiate at 5 mg orally once daily, with or without tadalafil 20 mg daily. At 4-week intervals, either the dose of Letairis or tadalafil can be increased, as needed and tolerated, to Letairis 10 mg or tadalafil 40mg.
Tracleer® (oral tablet & tablet for oral suspension)	Patients > 12 years and > 40 kg: Initiate at 62.5 mg orally twice daily for 4 weeks and then increase to 125 mg twice daily. Patients > 12 years and <40 kg: Administer 62.5 mg orally twice daily Patients ≤ 12 years of age*: • ≥ 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 *Disperse tablets (or dispersible tablet half) for oral suspension in a minimal amount of water immediately before administration. *Reduce dose in patients with aminotransferase elevations > 3 x ULN.*

Indication	Dose
	Stop treatment permanently in patients with aminotransferase elevations > 8 x ULN, if liver aminotransferase elevations are accompanied by clinical symptoms of hepatotoxicity (i.e. nausea, vomiting, jaundice, etc.), or bilirubin ≥ 2 x ULN.
Opsumit® (oral tablet)	Administer 10 mg orally once daily
Adempas® (oral tablet)	Initiate treatment at 1 mg orally three times a day. For patients who may not tolerate the hypotensive effect, consider starting dose of 0.5 mg three times a day. Dosage may be increased by 0.5 mg at intervals of no sooner than 2-weeks as tolerated to a maximum of 2.5 mg three times a day. *Refer to prescribing information for dose adjustments in patients who smoke or when co-administered with strong CYP450 or P-gp/BCRP inhibitors.*
Uptravi® (oral tablet)	Recommended starting dose is 200 mcg orally twice daily. Increase the dose in increments of 200 mcg twice daily, usually at weekly intervals, to the highest tolerated dose up to 1600 mcg twice daily. *Tolerability may be improved when taken with food *Refer to prescribing information for dose adjustments in patients with moderate hepatic impairment (Child-Pugh class B) or when co-administered with moderate CYP2C8 inhibitors (use with strong CYP2C8 inhibitors is contraindicated).*
Tadliq® (oral suspension)	Administer 40 mg (10 mL) orally once daily *Refer to prescribing information for dose adjustments in patients with hepatic impairment (Child-Pugh Class A, B ,or C), renal impairment (CrCl ≤ 80 mL/min), or in combination with ritonavir.*
Liqrev® (oral suspension)	Administer 20 mg (2 mL) orally three times daily
***NOTE:** Refer to the respective prescribing information for each individual medication addressed in this policy for all drug to drug interactions.*	

VI. Billing Code/Availability Information

Drug	HCPCS Code	Billable Units (BU)	Drug strength/formulation	NDC
Orenitram (United Therapeutics)	J8499	n/a	0.125 mg tablet	66302-0300-xx
			0.25 mg tablet	66302-0302-xx
			1 mg tablet	66302-0310-xx
			2.5 mg tablet	66302-0325-xx
			5 mg tablet	66305-0350-xx
			Month 1 Titration Kit containing 4 weekly cartons (0.125 mg, 0.25 mg)	66302-0361-xx
			Month 2 Titration Kit containing 4 weekly cartons (0.125 mg, 0.25 mg)	66302-0362-xx
			Month 3 Titration Kit containing 4 weekly cartons (0.125 mg, 0.25 mg, 1 mg)	66302-0363-xx

Drug	HCPCS Code	Billable Units (BU)	Drug strength/formulation	NDC
Tyvaso (United Therapeutics)	J7686	1.74 mg = 1 BU	1.74 mg/2.9 mL ampule	66302-0206-xx
Tyvaso DPI (United Therapeutics)	J8499	n/a	Titration Kit (16 mcg, 32 mcg)	66302-0600-xx
			Titration Kit (16 mcg, 32 mcg, 48 mcg)	66302-0610-xx
			Maintenance Kit (16 mcg)	66302-0616-xx
			Maintenance Kit (32 mcg)	66302-0632-xx
			Maintenance Kit (48 mcg)	66302-0648-xx
			Maintenance Kit (64 mcg)	66302-0664-xx
			Maintenance Kit (32 mcg, 48 mcg)	66302-0620-xx
			Institutional Kit (16 mcg)	66302-0716-xx
			Institutional Kit (32 mcg)	66302-0732-xx
			Institutional Kit (48 mcg)	66302-0748-xx
			Institutional Kit (64 mcg)	66302-0764-xx
			Institutional Kit (32 mcg, 48 mcg)	66302-0720-xx
Ventavis (Actelion Pharm)	Q4074	20 mcg = 1 BU	10 mcg/mL ampule	66215-0302-xx
			20 mcg/mL ampule	66215-0303-xx
Letairis (Gilead Sciences)	J8499	n/a	5 mg tablet	61958-0801-xx
			10 mg tablet	61958-0802-xx
Tracleer (Actelion Pharm)	J8499	n/a	62.5 mg tablet	66215-0101-xx
			125 mg tablet	66215-0102- xx
			32 mg tablet for oral suspension	66215-0103- xx
Opsumit (Actelion Pharm)	J8499	n/a	10 mg tablet	66215-0501-xx
Adempas (Bayer)	J8499	n/a	0.5 mg tablet	50419–0250-xx
			1 mg tablet	50419–0251-xx
			1.5 mg tablet	50419–0252-xx
			2 mg tablet	50419–0253-xx
			2.5 mg tablet	50419–0254-xx
Uptravi (Actelion Pharm)	J8499	n/a	200 mcg tablet	66215-0602-xx
			400 mcg tablet	66215-0604-xx
			600 mcg tablet	66215-0606-xx
			800 mcg tablet	66215-0608-xx
			1000 mcg tablet	66215-0610-xx
			1200 mcg tablet	66215-0612-xx
			1400 mcg tablet	66215-0614-xx
			1600 mcg tablet	66215-0616-xx
			Titration Pack	66215-0628-xx
Revatio* (Pfizer)	J8499	n/a	20 mg tablet	00069-4190-xx
			10 mg/mL oral suspension	00069-0336-xx
Adcirca* (Eli Lilly)	J8499	n/a	20 mg tablet	66302-0467-xx
Tadliq (CMP Pharma)	J8499	n/a	20 mg/5 mL oral suspension	46287-0045-xx
Liqrev (CMP Pharma)	J8499	n/a	10 mg/mL oral suspension	46287-0055-xx

* *Generic available from multiple manufacturers*

VII. References

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Appendix 1 – Covered Diagnosis Codes

ICD-10	Description
I27.0	Primary pulmonary hypertension
I27.1	Kyphoscoliotic heart disease
I27.20	Pulmonary hypertension, unspecified
I27.21	Secondary pulmonary arterial hypertension
I27.24*	Chronic thromboembolic pulmonary hypertension
I27.83	Eisenmenger's syndrome
I27.89	Other specified pulmonary heart diseases
I27.9	Pulmonary heart disease, unspecified
M34.0	Progressive systemic sclerosis
M34.1	CR(E)ST syndrome

ICD-10	Description
M34.9	Systemic sclerosis, unspecified

**Applicable to Adempas Only*

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Determinations (LCDs), and Local Coverage Articles (LCAs) may exist and compliance with these policies is required where applicable. They can be found at https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC