| Reference number |
|------------------|
| 1771-H |

SPECIALTY QUANTITY LIMIT PROGRAM

Pulmonary Arterial Hypertension

I. PROGRAM DESCRIPTION

The standard limit is designed to allow a quantity sufficient for the most common uses of the medication. If the member's plan allows a quantity limit exception review for the requested medication, coverage of an additional quantity may be provided up to the exception limit with prior authorization. The recommended dosing parameters for the treatment of pulmonary arterial hypertension fall within the standard limits. Coverage of an additional quantity may be reviewed on a case-by-case basis upon request.

II. COVERED QUANTITIES

| Medication** | Standard Limit | Exception Limit* | FDA-recommended dosing |
|-----------------------------------|-----------------------|------------------|---|
| Adcirca 20 mg tablets | 60 per 30 days | Not applicable | 40 mg per day |
| Adempas 0.5 mg tablets | 90 per 30 days | Not applicable | The recommended starting dosage is 1 mg taken 3 times a day. For patients who cannot tolerate the hypotensive effect of Adempas, |
| Adempas 1 mg tablets | 90 per 30 days | Not applicable | consider a starting dose of 0.5 mg taken three times a day. If systolic blood pressure remains greater than 95 mmHg and the patient has no signs or symptoms of hypotension, up-titrate by 0.5 mg taken three times a day. Dose increases should be no sooner than 2 weeks apart. The dose can be increased to the highest tolerated dosage, up to a maximum of 2.5 mg taken 3 times per day. If at any time, the patient has symptoms of hypotension, decrease the dosage by 0.5 mg taken three times per day. |
| Adempas 1.5 mg tablets | 90 per 30 days | Not applicable | |
| Adempas 2 mg tablets | 90 per 30 days | Not applicable | |
| Adempas 2.5 mg tablets | 90 per 30 days | Not applicable | |
| Letairis 5 mg tablets | 30 per 30 days | Not applicable | Initiate treatment at 5 mg once daily with or without tadalafil 20 mg once daily. At 4-week |
| Letairis 10 mg tablets | 30 per 30 days | Not applicable | intervals, either the dose of Letairis of tadalafil can be increased, as needed and tolerated, to Letairis 10 mg or tadalafil 40 mg. |
| Opsumit 10 mg tablets | 30 per 30 days | Not applicable | 10 mg per day |
| Revatio 10 mg/ml suspension | 224 ml per 30 days | Not applicable | 20 mg three times a day |

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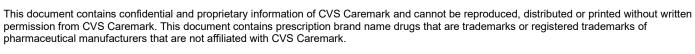
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| Medication** | Standard Limit | Exception Limit* | FDA-recommended dosing |
| Revatio (sildenafil) tablet 20 mg | 90 per 30 days | 360 per 30 days | Up to 20 mg three times a day |
| Tracleer 125 mg tablets | 60 per 30 days | Not applicable | Patients >12 years of age and >40 kg: Initiate treatment at 62.5 mg twice daily for 4 weeks and then increase to maintenance dose of 125 mg twice daily. |
| Tracleer 62.5 mg tablets | 60 per 30 days | Not applicable | Patients >12 years of age and <40 kg - Initial and maintenance dosing: 62.5 mg twice daily Patients <12 years of age – Initial and |
| Tracleer 32 mg tablets | 112 per 28 days | Not applicable | maintenance dosing: • ≥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 |
| Tyvaso 0.6 mg/ml inhalation solution | 81.2 ml (28 ampules) per 28 days | Not applicable | 54 mcg four times a day |
| Uptravi 200 mcg tablets | 60 per 30 days | Not applicable | Starting dose: 200 mcg twice daily. Increase the dose by 200 mcg twice daily at weekly intervals to the highest tolerated dose up to 1600 mcg twice daily. |
| Uptravi 400 mcg tablets | 60 per 30 days | Not applicable | . Toos mag times daily. |
| Uptravi 600 mcg tablets | 60 per 30 days | Not applicable | |
| Uptravi 800 mcg tablets | 60 per 30 days | Not applicable | |
| Uptravi 1000 mcg tablets | 60 per 30 days | Not applicable | |
| Uptravi 1200 mcg tablets | 60 per 30 days | Not applicable | |

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| Medication** | Standard Limit | Exception Limit* | FDA-recommended dosing |
|---|----------------------------|------------------|--|
| Uptravi 1400 mcg tablets | 60 per 30 days | Not applicable | |
| Uptravi 1600 mcg tablets | 60 per 30 days | Not applicable | |
| Ventavis 10 mcg/ml inhalation solution | 270 ampules per 30 days | Not applicable | 5 mcg up to nine times per day. The 20 mcg/ml concentration is intended for patients who are maintained at the 5 mcg dose and who have repeatedly experienced extended |
| Ventavis 20 mcg/ml inhalation solution | 270 ampules per 30 days | Not applicable | treatment times which could result in incomplete dosing. |

^{*}Coverage up to the exception limits may be provided with prior authorization. See Specialty Post Limit Quantity Exception Criteria for the criteria for approval.

III. REFERENCES

- 1. Adcirca [package insert]. Indianapolis, IN: Eli Lilly and Company; August 2017.
- 2. Adempas [package insert]. Whippany, NJ: Bayer HealthCare Pharmaceuticals Inc.; January 2018.
- 3. Letairis [package insert]. Foster City, CA: Gilead Sciences, Inc.; November 2018.
- 4. Opsumit [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US, Inc.; April 2019.
- 5. Revatio [package insert]. New York, NY: Pfizer Labs; January 2019.
- 6. Tracleer [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US, Inc.; October 2018.
- 7. Tyvaso [package insert]. Research Triangle Park, NC: United Therapeutics Corp.; October 2017.
- 8. Ventavis [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US, Inc.; October 2017.
- 9. Uptravi [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US, Inc.; December 2017.



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^{**} The limit may apply to the generic equivalent medications.