Drug Coverage Policy

Effective Date.....9/1/2024
Coverage Policy Number IP0483
Policy Title.....Pulmozyme

Cystic Fibrosis – Pulmozyme

- Pulmozyme® (dornase alfa inhalation solution – Genentech/Roche)

INSTRUCTIONS FOR USE
The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment and have discretion in making individual coverage determinations. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

Cigna Healthcare Coverage Policy

OVERVIEW
Pulmozyme, a recombinant human deoxyribonuclease I, is indicated in conjunction with standard therapies for the management of patients with **cystic fibrosis** to improve pulmonary function.[1]

Guidelines
According to Patient Registry data compiled by the Cystic Fibrosis Foundation (2022), Pulmozyme is used by the vast majority of patients with cystic fibrosis.[2] Guidelines from the Cystic Fibrosis Foundation (2007, updated in 2013) address the chronic use of medications for management of

lung health in cystic fibrosis patients ≥ 6 years of age.[3,4] These guidelines recommend Pulmozyme use for patients with cystic fibrosis regardless of disease severity to improve lung function and reduce exacerbations. Separate guidelines have addressed Pulmozyme use in younger patients.[5,6] Although efficacy data are lacking in patients < 5 years of age, safety and tolerability have been established in patients as young as 3 months.[1,6] Cystic Fibrosis Foundation guidelines for infants < 2 years of age (2009) and children between 2 and 5 years of age (2016) support Pulmozyme use in these populations based on individual circumstances.[5,6]

Medical Necessity Criteria

Pulmozyme is considered medically necessary when the following criteria are met:

FDA-Approved Indication

1. **Cystic Fibrosis.** Approve for 1 year if the medication is prescribed by or in consultation with a pulmonologist or a physician who specializes in the treatment of cystic fibrosis.

When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Receipt of sample product does not satisfy any criteria requirements for coverage.

Conditions Not Covered

Any other use is considered experimental, investigational, or unproven, including the following (this list may not be all inclusive; criteria will be updated as new published data are available):

1. **Asthma.** Mucus hypersecretion may be mediated by a variety of causes, including inflammation, irritation, stimulation, or mucus-producing tumors.[7] However, efficacy of Pulmozyme is not established for conditions other than cystic fibrosis.[10] In a pilot study of patients with severe acute asthma (n = 50), there was no significant difference in forced expiratory volume in 1 second (FEV_1) with Pulmozyme use vs. placebo.[8,10]

2. **Bronchiectasis, Idiopathic.** A multicenter, double-blind, randomized, placebo-controlled 24-week trial (n = 349) examined the effect of Pulmozyme vs. placebo in patients with idiopathic bronchiectasis (i.e., bronchiectasis not related to cystic fibrosis).[9] Patients in the Pulmozyme arm experienced worsened lung function and more frequent pulmonary exacerbations vs. placebo. The authors concluded that Pulmozyme should not be used in this population. Another small double-blind, placebo-controlled, Phase II trial (n = 14) examined Pulmozyme in adults with non-cystic fibrosis bronchiectasis. Pulmozyme failed to show any significant change in any of the outcome variables or in sputum transportability.[10,11]

References

1. Pulmozyme® inhalation solution [prescribing information]. South San Francisco, CA: Genentech/Roche; February 2024.
2. Cystic Fibrosis Foundation. Patient Registry: 2022 Annual Data Report. Available at: https://www.cff.org/medical-professionals/patient-registry. Accessed on May 2, 2024.
3. Flume PA, O'Sullivan BP, Robinson KA, et al. Cystic Fibrosis Pulmonary Guidelines: Chronic Medications for Maintenance of Lung Health. *Am J Respir Crit Care Med.* 2007;176:957-969.

4. Maysel PJ Jr, Naureckas ET, Robinson KA, et al, Pulmonary Clinical Practice Guidelines Committee. Cystic Fibrosis Pulmonary Guidelines: Chronic Medications for Maintenance of Lung Health. *Am J Respir Crit Care Med.* 2013;187(7):680-689.
5. Borowitz D, Robinson KA, Rosenfeld M, et al, Cystic Fibrosis Foundation. Cystic Fibrosis Foundation evidence-based guidelines for management of infants with cystic fibrosis. *J Pediatr.* 2009;155(6 Suppl):S73-93.
6. Lahiri T, Hempstead SE, Brady C, et al. Clinical practice guidelines from the Cystic Fibrosis Foundation for preschoolers with cystic fibrosis. *Pediatrics.* 2016;137(4): e20151784.
7. Rubin BK. Aerosol medications for treatment of mucus clearance disorders. *Respiratory Care.* 2015;60(6):825-832.
8. Silverman RA, Foley F, Dalipi R, et al. The use of rhDNase in severely ill, non-intubated adult asthmatics refractory to bronchodilators: a pilot study. *Respir Med.* 2012; 106(8):1096-1102.
9. O'Donnell AE, Barker AF, Ilowite JS, Fick RB. Treatment of idiopathic bronchiectasis with aerosolized recombinant human DNase I. rhDNase Study Group. *Chest.* 1998;113(5):1329-1334.
10. Tarrant BJ, Le Maitre C, Romero L, et al. Mucoactive agents for non-cystic fibrosis lung disease: a systematic review and meta-analysis. *Respirology.* 2017;22(6):1084-1092.
11. Wills PJ, Wodehouse T, Corkery K, Mallon K,Wilson R, and Cole PJ. Short-term recombinant human DNase in bronchiectasis. Effect on clinical state and in vitro sputum transportability. *Am J Respir Crit Care Med.* 1996;154:413-417.

Revision Details

Type of Revision	Summary of Changes	Date
Annual Revision	**Updated** coverage policy title from *Dornase Alfa* to *Cystic Fibrosis – Pulmozyme*. **Cystic Fibrosis.** **Removed** criterion, *"used to improve pulmonary function in cystic fibrosis (CF)."* **Treatment of Complicated Pleural Effusions.** **Removed** criteria for coverage of, *Treatment of Complicated Pleural Effusions*.	9/1/2024

The policy effective date is in force until updated or retired.