Drug Coverage Policy

Effective Date.....5/1/2025
Coverage Policy Number IP0126
Policy Title.....Bronchitol

Cystic Fibrosis – Bronchitol

- Bronchitol® (mannitol oral inhalation powder – Pharmaxis/Chiesi)

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 where appropriate and have discretion in making individual coverage determinations. Where coverage for care or services does not depend on specific circumstances, reimbursement will only be provided if a requested service(s) is submitted in accordance with the relevant criteria outlined in the applicable Coverage Policy, including covered diagnosis and/or procedure code(s). Reimbursement is not allowed for services when billed for conditions or diagnoses that are not covered under this Coverage Policy (see "Coding Information" below). When billing, providers must use the most appropriate codes as of the effective date of the submission. Claims submitted for services that are not accompanied by covered code(s) under the applicable Coverage Policy will be denied as not covered. 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.

OVERVIEW
*Bronchitol, a sugar alcohol, is indicated as add-on maintenance therapy to improve pulmonary function in patients ≥ 18 years of age with **cystic fibrosis** (CF).*[1]

Safety

Bronchitol can cause bronchospasm, which can be severe in susceptible patients.[1] Therefore, Bronchitol is contraindicated in individuals who fail to pass the Bronchitol Tolerance Test. Prior to prescribing Bronchitol, the Bronchitol Tolerance Test must be administered and performed under the supervision of a healthcare practitioner who is able to manage acute bronchospasm, to identify patients who are suitable candidates for Bronchitol maintenance therapy. For patients who have passed the Bronchitol Tolerance Test, the recommended dosage of Bronchitol is 400 mg twice a day by oral inhalation (the contents of 10 capsules administered individually) via the inhaler. A short-acting bronchodilator should be administered by oral inhalation, 5 to 15 minutes before every dose of Bronchitol. Bronchitol should be taken once in the morning and once in the evening, with the later dose taken at least 2 to 3 hours before bedtime.

Guidelines
Bronchitol is not addressed in US guidelines for CF. Guidelines from the CF Foundation (2013) in the US strongly recommend chronic use of Pulmozyme® (dornase alfa inhalation solution) in patients ≥ 6 years of age with moderate to severe disease to improve lung function and quality of life and reduce exacerbations. Pulmozyme is also recommended for chronic use in patients ≥ 6 years of age with asymptomatic or mild disease to improve lung function and reduce exacerbations. Chronic use of hypertonic saline is also recommended in individuals with CF who are ≥ 6 years of age to improve lung function and quality of life and reduce exacerbations. Standards for the care of patients with CF from the European CF Society (2024) recognize inhaled dry powder mannitol as an alternative to hypertonic saline in patients with CF; however, both agents are noted to irritate the airways and therefore initial tolerance testing is recommended often with the use of premedication with bronchodilators.[6]

Coverage Policy

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

FDA-Approved Indication

1. **Cystic Fibrosis.** Approve for 1 year if the patient meets ALL of the following (A, B, C, D, and E):
   - **A)** Patient is ≥ 18 years of age; AND
   - **B)** Patient has tried hypertonic saline; AND
   - **C)** Patient has passed the Bronchitol Tolerance Test; AND
   - **D)** Patient will pre-medicate with a short-acting bronchodilator; AND
   - **E)** The medication is prescribed by or in consultation with a pulmonologist or a physician who specializes in the treatment of cystic fibrosis.

Bronchitol for any other use is considered not medically necessary, including the following (this list may not be all inclusive; criteria will be updated as new published data are available):

1. **Concomitant Use with Hypertonic Saline.** Bronchitol has not been studied in combination with hypertonic saline.[3-5]

References

1. Bronchitol® oral inhalation powder [prescribing information]. Frenchs Forest NSW, Australia/Cary, NC: Pharmaxis/Chiesi; October 2020.

2. Mogayzel PJ, 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.
3. Flume P, Amelina E, Daines CL, et al. Efficacy and safety of inhaled dry-powder mannitol in adults with cystic fibrosis: An international, randomized controlled study. *J Cyst Fibros.* 2020;30(6):1003-1009.
4. Bilton D, Robinson P, cooper P, et al; for the CF301 Study Investigators. Inhaled dry powder mannitol in cystic fibrosis: an efficacy and safety study. *Eur Respir J*. 2011; 38:1071-1080.
5. Aitken ML, Bellon G, De Boeck K, et al; for the CF302 Investigators. Long-term inhaled dry powder mannitol in cystic fibrosis. An international randomized study. *Am J Respir Crit Care*. 2012;185(6): 645-652.
6. Burgel PR, Southern KW, Addy C, et al. Standards for the care of patients with cystic fibrosis (CF); recognizing and addressing CF health issues. *J Cyst Fibros.* 2024;23(2):187-202.

Revision Details

Type of Revision	Summary of Changes	Date
Annual Revision	No criteria changes.	06/01/2024
Annual Revision	No criteria changes.	05/01/2025

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