# PHARMACY POLICY STATEMENT

| Original Effective Date | Next Annual Review | | Last Revision |
|---|---|---|---|
| 10/06/2015 | 10/06/2017 | | 01/16/2017 |
| **Policy Name** | | | **Policy Number** |
| **Cystic Fibrosis** | | | SRx-0025 |
| **Policy Type** | | | |
| Medical | Administrative | PHARMACY | Reimbursement |

Pharmacy Policy Statements prepared by CSMG Co. and its affiliates (including CareSource) are derived from literature based on and supported by clinical guidelines, nationally recognized utilization and technology assessment guidelines, other medical management industry standards, and published MCO clinical policy guidelines. Medically necessary services include, but are not limited to, those health care services or supplies that are proper and necessary for the diagnosis or treatment of disease, illness, or injury and without which the patient can be expected to suffer prolonged, increased or new morbidity, impairment of function, dysfunction of a body organ or part, or significant pain and discomfort. These services meet the standards of good medical practice in the local area, are the lowest cost alternative, and are not provided mainly for the convenience of the member or provider. Medically necessary services also include those services defined in any Evidence of Coverage documents, Pharmacy Policy Statements, Provider Manuals, Member Handbooks, and/or other policies and procedures.

Pharmacy Policy Statements prepared by CSMG Co. and its affiliates (including CareSource) do not ensure an authorization or payment of services. Please refer to the plan contract (often referred to as the Evidence of Coverage) for the service(s) referenced in the Pharmacy Policy Statement. If there is a conflict between the Pharmacy Policy Statement and the plan contract (i.e., Evidence of Coverage), then the plan contract (i.e., Evidence of Coverage) will be the controlling document used to make the determination.

## Contents of Policy

## A. INTRODUCTION

Cystic fibrosis (CS) is an inherited disease of the secretory glands. It is characterized by buildup of sticky and thick mucus in lungs, pancreas, liver and other organs. The mucus blocks air flow in lungs, making breathing difficult and allowing bacteria to grow. Recurrent lung infections leads to lung damage and decline in lung function. Medications used to treat patients with CF may include antibiotics, mucolytics, CFTR (Cystic fibrosis transmembrane conductance regulator) potentiators, bronchodilators, pancreatic enzyme supplements, and multivitamins. The following Pharmacy Policy Statement covers the coverage criteria only for CFTR potentiators, mucolytic agents and antibiotics for CS.

The intent of CareSource Pharmacy Policy Statements is to encourage appropriate selection of patients for therapy according to product labeling, clinical guidelines, and/or clinical studies as well as to encourage use of preferred agents. The CareSource Pharmacy Policy Statement is a guideline for determining health care coverage for our patients with benefit plans covering prescription drugs. Pharmacy Policy Statements are written on selected prescription drugs requiring prior authorization or step therapy. The Pharmacy Policy Statement is used as a tool to be interpreted in conjunction with the member's specific benefit plan.

*NOTE: The Introduction section is for your general knowledge and is not to be construed as policy coverage criteria. The rest of the policy uses specific words and concepts familiar to medical professionals and is intended for providers. A provider can be a person, such as a doctor, nurse, psychologist, or dentist. A provider can also be a place where medical care is given, like a hospital, clinic or lab. This policy informs providers about when a service may be covered.*

## B. DEFINITIONS

1. None applicable.

## C. POLICY COVERAGE CRITERIA

**1. Site of Service**

| Site of Service Administration | Coverage Criteria |
|---|---|
| **Office, Outpatient, Home** | **Preferred place of service is in the home.**<br><br>CareSource supports administering injectable medications in various settings, as long as those services are furnished in the most appropriate and cost effective settings that are supportive of the patient's medical condition(s) and unique needs and condition(s). The decision on the most appropriate setting for administration is based on the member's current medical condition(s) and any required monitoring or additional services that may coincide with the delivery of the specific medication. |

## 2. Coverage Criteria

CareSource will approve the use of Tobi (tobramycin Inhalation solution), Cayston (aztreonam inhalation solution), Kalydeco (ivacaftor), Orkambi (lumacaftor/ivacaftor), Pulmozyme (dornase alfa inhalation solution) and consider its use medically necessary when the criteria have been met for each condition listed below. Prior authorization request should be submitted with chart notes and documentation supporting medical necessity.

***Please note that all members approved for therapy tobramycin Inhalation solution will be required to use generic tobramycin Inhalation solution or Tobi (tobramycin Inhalation solution) first line unless patient has a documented allergy to either products. Other currently available agents are non-formulary and include: Bethkis and Kitabis. Please see the Medical Necessity for Non-Formulary Medications policy for more information.***

| Drug | Coverage criteria: |
|---|---|
| **Tobi Neb solution (tobramycin solution)** | 1) Member is 6 years of age or older<br>2) Diagnosis of Cystic Fibrosis<br>3) Member has a positive culture for Pseudomonas aeruginosa (*Documented in chart notes and submitted with prior authorization request*)<br>4) Prescribed by a pulmonologist or an infectious disease specialist<br>5) Member has documented forced expiratory volume in 1 second (FEV1) > 25% or < 75% predicted (*Chart notes required with prior authorization request*)<br>6) Ineffectiveness, intolerance or contraindication to generic tobramycin inhalation solution (*Documented in chart notes and submitted with prior authorization request)* |
| **Cayston (aztreonam inhalation solution)** | 1) Member is 7 years of age or older<br>2) Diagnosis of Cystic Fibrosis<br>3) Member has a positive culture for Pseudomonas aeruginosa (*Documented in chart notes and submitted with prior authorization request*)<br>4) Prescribed by a pulmonologist or an infectious disease specialist<br>5) Member has documented forced expiratory volume in 1 second (FEV1) > 25% or < 75% predicted (*Documented in chart notes and submitted with prior authorization request*) |
| **Kalydeco (ivacaftor)** | 1) Member is 2 years of age or older<br>2) Diagnosis of Cystic Fibrosis<br>3) Prescribed by a pulmonologist or an infectious disease specialist<br>4) Member has had genetic testing and has one of the following mutations in the CFTR gene: G551D, G1244E, G1349D, G178R, G551S, |

| | |
|---|---|
| | S1251N, S1255P, S549N, S549R or R117H (*Documented in chart notes and submitted with prior authorization request*) |
| **Orkambi (lumacaftor/ivacaftor)** | 1) Member is 6 years of age or older<br>2) Diagnosis of Cystic Fibrosis<br>3) Prescribed by a pulmonologist or an infectious disease specialist<br>4) Member has had genetic testing and has two copies (homozygous) of the F508del mutation (F508del/F508del) in their CFTR gene (*Documented in chart notes and submitted with prior authorization request*) |
| **Pulmozyme (dornase alfa inhalation solution)** | 1) Member is 5 years of age or older<br>2) Diagnosis of Cystic Fibrosis<br>3) Prescribed by a pulmonologist or an infectious disease specialist<br>4) Member has forced vital capacity (FVC) predicted > 40% (*Documented in chart notes and submitted with prior authorization request*) |

**All other uses of Tobi (tobramycin Inhalation solution), Cayston (aztreonam inhalation solution), Kalydeco (ivacaftor), Orkambi (lumacaftor/ivacaftor), Pulmozyme (dornase alfa inhalation solution) are considered experimental/investigational; and therefore, will follow CareSource's off-label policy.**

***Please note that this policy is reviewed on an annual basis. New drugs and indications receiving FDA approval may not be reflected in this policy immediately.***

**Notes:**

- Documented diagnosis must be confirmed by portions of the individual's medical record which need to be supplied with prior authorization request. These medical records may include, but are not limited to test reports, chart notes from provider's office, or hospital admission notes.
- Patient is required to have completed the trial(s) listed in the above criteria unless the patient is unable to tolerate or has a contraindication to trial medications. Documentation such as chart notes or pharmacy claims may be requested to verify trial(s), intolerance, or contraindication(s).
- Refer to the product package insert for dosing, administration and safety guidelines.
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**3. Dosage and Quantity Limits (listed if applicable)**

*Information for patients with renal or hepatic impairment is not included. See package insert for individual agents.*

| Drug | Dosage and Quantity Limit |
|---|---|
| **Tobi Neb solution (tobramycin solution)** | 300 mg every 12 hours; administer in repeated cycles of 28 days on drug followed by 28 days off drug. |
| **Cayston (aztreonam inhalation solution)** | 75 mg 3 times daily; for 28 days; do not repeat for 28 days after completion.<br><br>Administer only with the Altera® Nebulizer System. Do not administer with any other type of nebulizer. |
| **Kalydeco (ivacaftor)** | One 150 mg tablet every 12 hours. |
| **Orkambi (lumacaftor/ivacaftor)** | Adults and pediatric patients age 12 years and older: two tablets (each containing lumacaftor 200 mg/ivacaftor 125 mg) taken orally every 12 hours.<br><br>Pediatric patients age 6 through 11 years: two tablets (each containing lumacaftor 100 mg/ivacaftor 125 mg) taken orally every 12 hours. |
| **Pulmozyme (dornase alfa inhalation solution)** | 2.5 mg daily through selected jet nebulizers in conjunction with a Pulmo-Aide, Pari-Proneb, Mobilaire, or Porta-Neb compressor system or eRapid Nebulizer System. Some patients may benefit from twice daily administration. |

**4. Authorization Period**

| Drug | Approval Period |
|---|---|
| **Tobi Neb solution**<br>**Cayston**<br>**Pulmozyme** | The initial authorization for Tobi Neb solution, Cayston, and Pulmozyme is valid for 1 year.<br><br>Continued treatment for Tobi Neb solution, Cayston, and Pulmozyme may be considered when patient meets current policy criteria, and has evidence of a beneficial response to treatment. A reauthorization after successful initiation period will be placed for 1 year.<br><br>**ALL** authorizations are subject to continued eligibility. |
| **Kalydeco**<br>**Orkambi** | The initial authorization Kalydeco and Orkambi is valid for 3 months.<br><br>Continued treatment may be considered when adherence is confirmed by claims history, patient meets current policy criteria, and has evidence of a beneficial response to treatment. A reauthorization after successful initiation period will be placed for 1 year.<br><br>**ALL** authorizations are subject to continued eligibility. |

**5. Coding**

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|---|---|
| **CPT** | |
| **81220** | CFTR (cystic fibrosis transmembrane conductance regulator) (eg, cystic fibrosis) gene analysis; common variants (eg, ACMG/ACOG guidelines) |
| **81221** | CFTR (cystic fibrosis transmembrane conductance regulator) (eg, cystic fibrosis) gene analysis; common variants (eg, ACMG/ACOG guidelines) |
| **81222** | CFTR (cystic fibrosis transmembrane conductance regulator) (eg, cystic fibrosis) gene analysis; common variants (eg, ACMG/ACOG guidelines) |
| **81223** | CFTR (cystic fibrosis transmembrane conductance regulator) (eg, cystic fibrosis) gene analysis; common variants (eg, ACMG/ACOG guidelines) |
| **HCPCS** | |
| **Not applicable** | Covered on the pharmacy benefit |

## D. RELATED POLICIES

**AD-0004:** Medical Necessity - Off-Label, Approved Orphan and Compassionate Use Drugs
**AD-0008:** Medical Necessity for Non-Formulary Medications

## E. REVIEW/REVISION HISTORY

| DATE | ACTION/DESCRIPTION |
|---|---|
| **10/06/2015** | Last review/revision date |
| **01/16/2017** | Non-formulary agents Bethkis and Kitabis were removed; their use is covered by policy AD-0008. J –codes for Kalydeco and Orkambi removed as these are billed under pharmacy benefits. |

## F. REFERENCES

1. National Guideline Clearinghouse (NGC). Guideline summary: Cystic fibrosis pulmonary guidelines. Chronic medications for maintenance of lung health. In: National Guideline Clearinghouse (NGC) [Web site]. Rockville (MD): Agency for Healthcare Research and Quality (AHRQ); 2013 Apr 01. [cited 2016 Dec 19]. Available: https://www.guideline.gov
2. Bethkis [package insert]. Foster City, CA: Gilead Sciences Inc; 2014.
3. Tobi [package insert]. East Hanover, New Jersey: Novartis Pharmaceuticals Corporation; 2015.
4. Kitabis [package insert]. Midlothian, VA: Catalent Pharma Solutions LLC; 2014.
5. Cayston [package insert]. Foster City, CA: Gilead Sciences Inc; 2014.
6. Kalydeco [package insert]. Boston, MA: Vertex Pharmaceuticals Inc; 2015.

7. Orkambi [package insert]. Boston, MA: Vertex Pharmaceuticals Inc; 2016.
8. Pulmozyme [package insert]. South San Francisco, CA: Genentech Inc; 2014.

**The Pharmacy Policy detailed above has received due consideration and is approved.**