

# Specialty Quantity Limit Cystic Fibrosis Medications

## Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Alyftrek	vanzacaftor/tezacaftor/deutivacaftor
Bethkis	tobramycin inhalation solution
Bronchitol	mannitol inhalation powder
Cayston	aztreonam
Kalydeco	ivacaftor
Kitabis Pak	tobramycin inhalation solution
Orkambi	lumacaftor/ivacaftor
Pulmozyme	dornase alfa
Symdeko	tezacaftor/ivacaftor
TOBI	tobramycin inhalation solution
TOBI Podhaler	tobramycin inhalation powder
Trikafta	elexacaftor/tezacaftor/ivacaftor

## Program Description

The standard limit is designed to allow a quantity sufficient for the most common uses of the medication. The recommended dosing parameters for all FDA-approved indications fall within the standard limits. Coverage of an additional quantity may be reviewed on a case-by-case basis upon request.

# Covered Quantities

The limit may apply to the generic equivalent medications.

Medication	Standard Limit
Alyftrek (vanzacaftor/tezacaftor/deutivacaftor) 4 mg/20 mg/50 mg tablets	1 carton (84 tablets) per 28 days
Alyftrek (vanzacaftor/tezacaftor/deutivacaftor) 10 mg/50 mg/125 mg tablets	1 carton (56 tablets) per 28 days
Bethkis (tobramycin inhalation solution) 300 mg/4 mL ampules	224 mL per 28 days (56 ampules per 28 days)
Bronchitol (mannitol inhalation powder) 40 mg capsules	560 capsules per 28 days
Bronchitol Tolerance Test Pack (mannitol inhalation powder) 40 mg capsules	2 packs (20 capsules) per 7 days
Cayston (aztreonam for inhalation solution) 75 mg vials	84 vials per 28 days
Kalydeco (ivacaftor) tablets 150 mg blister carton	1 carton (56 tablets) per 28 days
Kalydeco (ivacaftor) tablets 150 mg	60 tablets per 30 days
Kalydeco (ivacaftor) oral granules 5.8 mg packets	56 packets per 28 days
Kalydeco (ivacaftor) oral granules 13.4 mg packets	56 packets per 28 days
Kalydeco (ivacaftor) oral granules 25 mg packets	56 packets per 28 days
Kalydeco (ivacaftor) oral granules 50 mg packets	56 packets per 28 days
Kalydeco (ivacaftor) oral granules 75 mg packets	56 packets per 28 days
Kitabis Pak (tobramycin inhalation solution) 300 mg/5 mL ampules	280 mL per 28 days (56 ampules per 28 days)
Orkambi (lumacaftor/ivacaftor) 100 mg/125 mg tablets	112 tablets per 28 days
Orkambi (lumacaftor/ivacaftor) 200 mg/125 mg tablets	112 tablets per 28 days
Orkambi (lumacaftor/ivacaftor) 75 mg/94 mg oral granule packets	56 packets per 28 days
Orkambi (lumacaftor/ivacaftor) 100 mg/125 mg oral granule packets	56 packets per 28 days
Orkambi (lumacaftor/ivacaftor) 150 mg/188 mg oral granule packets	56 packets per 28 days

Medication	Standard Limit
Pulmozyme (dornase alfa) inhalation solution 2.5 mg/2.5 mL ampules	150 mL per 30 days (60 ampules per 30 days)
Symdeko (tezacaftor/ivacaftor) 50 mg/75 mg tablets	56 tablets per 28 days
Symdeko (tezacaftor/ivacaftor) 100 mg/150 mg tablets	56 tablets per 28 days
TOBI (tobramycin inhalation solution) 300 mg/5 mL ampules	280 mL per 28 days (56 ampules per 28 days)
TOBI Podhaler (tobramycin inhalation powder) 28 mg capsules	224 capsules per 28 days
Trikafta (elexacaftor/tezacaftor/ivacaftor) 50 mg/25 mg/37.5 mg tablets	84 tablets per 28 days
Trikafta (elexacaftor/tezacaftor/ivacaftor) 100 mg/50 mg/75 mg tablets	84 tablets per 28 days
Trikafta (elexacaftor/tezacaftor/ivacaftor) 80 mg/40 mg/60 mg oral granules	56 packets per 28 days
Trikafta (elexacaftor/tezacaftor/ivacaftor) 100 mg/50 mg/75 mg oral granules	56 packets per 28 days

## FDA-recommended Dosing

### Alyftrek

#### Tablets

##### 4 mg/20 mg/50 mg

Age 6 to less than 12 years weighing less than 40 kg: Three tablets of vanzacaftor 4 mg/tezacaftor 20 mg/deutivacaftor 50 mg once daily

##### 10 mg/50 mg/125 mg

Age 6 to less than 12 years weighing 40 kg or more AND 12 years or older: Two tablets of vanzacaftor 10 mg/tezacaftor 50 mg/deutivacaftor 125 mg once daily

### Bethkis

One ampule (300 mg/4 mL) twice daily for 28 days (28 days on/28 days off cycle)

### Bronchitol

#### Capsules

Ten 40 mg capsules twice daily

Reference number(s)
1875-H

## **Tolerance Test Pack**

The Bronchitol Tolerance Test is administered to identify patients who are appropriate for inhaled mannitol use.

## **Cayston**

One vial (75 mg) reconstituted with 1 mL of sterile diluent administered 3 times a day for a 28-day course (followed by 28 days off Cayston therapy)

## **Kalydeco**

### **Tablets**

One 150 mg tablet every 12 hours

### **Oral granules**

#### **5.8 mg packets**

Age 1 month to less than 2 months weighing 3 kg or greater: One 5.8 mg packet every 12 hours

#### **13.4 mg packets**

Age 2 months to less than 4 months weighing 3 kg or greater: One 13.4 mg packet every 12 hours

#### **25 mg packets**

Age 4 months to less than 6 months weighing 5 kg or greater: One 25 mg packet every 12 hours

Age 6 months to less than 6 years weighing 5 kg to less than 7 kg: One 25 mg packet every 12 hours

#### **50 mg packets**

Age 6 months to less than 6 years weighing 7 kg to less than 14 kg: One 50 mg packet every 12 hours

#### **75 mg packets**

Age 6 months to less than 6 years weighing 14 kg or greater: One 75 mg packet every 12 hours

## **Kitabis Pak**

One ampule (300 mg/5 mL) twice daily for 28 days (28 days on/28 days off cycle)

## **Orkambi**

### **Tablets**

#### **100 mg/125 mg**

6 through 11 years of age: Two lumacaftor 100 mg/ivacaftor 125 mg tablets every 12 hours

#### **200 mg/125 mg**

12 years of age and older: Two lumacaftor 200 mg/ivacaftor 125 mg tablets every 12 hours

### **Granule packets**

#### **75 mg/94 mg**

Age 1 through 2 years weighing 7 kg to less than 9 kg: One lumacaftor 75 mg/ivacaftor 94 mg packet every 12 hours

Reference number(s)
1875-H

### **100 mg/125 mg**

Age 1 through 2 years weighing 9 kg to less than 14 kg AND ages 2 through 5 years weighing less than 14 kg: One lumacaftor 100 mg/ivacaftor 125 mg packet every 12 hours

### **150mg/188 mg**

Ages 1 through 5 weighing 14 kg or greater: One lumacaftor 150 mg/ivacaftor 188 mg packet every 12 hours

## **Pulmozyme**

One 2.5 mg ampule administered once daily

Some patients may benefit from twice daily administration.

## **Symdeko**

### **Tablets**

#### **50 mg/75 mg**

Age 6 to less than 12 years weighing less than 30 kg: One tablet (tezacaftor 50 mg/ivacaftor 75 mg) in the morning and one 75 mg ivacaftor tablet in the evening

#### **100 mg/150 mg**

Age 6 to less than 12 years weighing 30 kg or more AND 12 years or older: One tablet (tezacaftor 100 mg/ivacaftor 150 mg) in the morning and one 150 mg ivacaftor tablet in the evening

## **TOBI**

One ampule (300 mg/ 5 mL) administered twice daily for 28 days (28 days on/28 days off cycle)

## **TOBI Podhaler**

Four 28 mg capsules twice daily for 28 days (28 days on/28 days off cycle)

## **Trikafta**

### **Tablets**

#### **50 mg/25 mg/37.5 mg**

Age 6 to less than 12 years weighing less than 30 kg: Two tablets (elexacaftor 50 mg/tezacaftor 25 mg/ivacaftor 37.5 mg) in the morning and one 75 mg ivacaftor tablet in the evening

#### **100 mg/50 mg/75 mg**

Age 6 to less than 12 years weighing 30 kg or more AND 12 years or older: Two tablets (elexacaftor 100 mg/tezacaftor 50 mg/ivacaftor 75 mg) in the morning and one 150 mg ivacaftor tablet in the evening

### **Oral granules**

#### **80 mg/40 mg/60 mg**

Age 2 to less than 6 years weighing less than 14 kg: One elexacaftor 80 mg/tezacaftor 40 mg/ivacaftor 60 mg oral granule packet in the morning and one 59.5 mg ivacaftor oral granule packet in the evening

Reference number(s)
1875-H

### **100 mg/50 mg/75 mg**

Age 2 to less than 6 years weighing 14 kg or more: One elexacaftor 100 mg/tezacaftor 50 mg/ivacaftor 75 mg oral granule packet in the morning and one 75 mg ivacaftor oral granule packet in the evening

## **References**

1. Alyftrek [package insert]. Boston, MA: Vertex Pharmaceuticals Incorporated; December 2024.
2. Bethkis [package insert]. Woodstock, IL: Chiesi USA, Inc.; February 2023.
3. Bronchitol [package insert]. Cary, NC: Chiesi USA, Inc.; October 2020.
4. Cayston [package insert]. Foster City, CA: Gilead Sciences, Inc.; November 2019.
5. Kalydeco [package insert]. Boston, MA: Vertex Pharmaceuticals Incorporated; August 2023.
6. Kitabis Pak [package insert]. Midlothian, VA: PARI Respiratory Equipment, Inc.; August 2023.
7. Orkambi [package insert]. Boston, MA: Vertex Pharmaceuticals Inc.; August 2023.
8. Pulmozyme [package insert]. South San Francisco, CA: Genentech, Inc.; February 2024.
9. Symdeko [package insert]. Boston, MA: Vertex Pharmaceuticals Incorporated; August 2023.
10. TOBI [package insert]. Morgantown, WV: Mylan Specialty L.P.; February 2023.
11. TOBI Podhaler [package insert]. Morgantown, WV: Mylan Specialty L.P.; February 2023.
12. Trikafta [package insert]. Boston, MA: Vertex Pharmaceuticals Incorporated; August 2023.