

Common medications in Cystic Fibrosis

Proper medication use is extremely important in Cystic Fibrosis. Reliably devoting the time and effort to taking our medications is one of the most important things that can be done to control our symptoms, keep us out of the hospital, and lead to an improved quality of life. The following medication list is not meant to be comprehensive but does include the most commonly prescribed medications used in the treatment of Cystic Fibrosis

Multiple factors can affect our use of medications. Speak with your pharmacist if there are any barriers that may prevent you from properly taking your medications. It is important to report any negative effects of our medications to your healthcare providers. If you ever experience any severe reactions to any medications such as swelling, sudden trouble breathing (especially during nebulizer treatments), development of rashes, or itching then speak with a healthcare provider or seek urgent medical attention immediately.

Bronchodilators: These medications are used to “open up the lungs”. They relax the airway and make it easier to breath. Within Cystic Fibrosis these are normally given at the beginning of respiratory therapy because if we use these first, all of the medications we inhale afterwards can reach the lungs a little bit better.			
Generic name	Brand name(s)	How is this taken	Side effects
Albuterol	Proair®, Proair Respiclick®, Ventolin®, Proventil®	Nebulized or with an inhaler- Proair Respiclick® is a dry powder inhaler, all others are an aerosol	Shakiness, increased heartbeat, anxiety
Levalbuterol	Xopenex®		

Mucolytics: These medications work to break apart the mucus within our lungs. After use the mucus is thinner and less sticky, which makes it much easier to be coughed out. These are used right before or during vest therapy for maximum benefit			
Generic name	Brand name(s)	How is this taken	Side effects
Hypertonic Saline, Sodium chloride for inhalation	HyperSal®, PulmoSal®, NebuSal®	Nebulized	Increased cough, sore throat, chest tightness
Dornase Alfa	Pulmozyme®	Nebulized	Change in voice, runny nose, chest tightness, rash, fever
Acetylcysteine, NAC	Mucomyst®	Nebulized	Runny nose, nausea, sudden airway constriction

Antibiotics: These medications are used to both treat and prevent infections of the airway. Nebulized antibiotics are typically taken after vest therapy, because otherwise the mucus in the lungs may prevent the medication from reaching its target. Timing of oral antibiotics can vary based upon what we are taking. There are a multitude of IV antibiotics that are commonly used in Cystic Fibrosis under direct healthcare supervision, feel free to ask a healthcare professional about these medications if they are ever to be used.

Inhaled antibiotics

Generic name	Brand name(s)	How is this taken	Side effects
Tobramycin for inhalation	TOBI®, Bethkis®, Kitabis®	Nebulized	Cough, wheezing, change in voice, cough, runny nose, chest discomfort, headache
Aztreonam for inhalation	Cayston®	Nebulized	Cough, wheezing, nasal congestion, throat irritation
Colistimethate, Colistin	ColyMycin M®	Nebulized (the IV formulation is mixed by the patient prior to use)	Change in voice, cough, chest discomfort, cough, wheezing

Oral antibiotics

Generic name	Brand name(s)	How is this taken	Side effects
Azithromycin	Zithromax®	Oral- liquid or tablet	Upset stomach, diarrhea, nausea, secondary infections (such as yeast infections)
Amoxicillin (with or without clavulanic acid)	Amoxil®, Augmentin® (if it contains clavulanic acid)	Oral- tablets, chewable tablets, capsules, or liquid	
Cephalexin	Keflex®	Oral- tablets, capsules, or liquid	
Ciprofloxacin	Cipro®	Oral- tablets, liquid, or compounded liquid (for use in feeding tubes)	
Levofloxacin	Levaquin®	Oral- tablets or liquid	
Sulfamethoxazole/Trimethoprim	Bactrim®	Oral- tablets or liquid	
Linezolid	Zyvox®	Oral- liquid or tablets	

Inhaled corticosteroids: These medications prevent swelling in the airway and work to prevent drops in lung function. Always rinse mouth, brush teeth, or have something to drink after use to prevent the infection known as thrush from occurring.

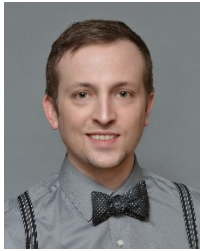
Generic name	Brand name(s)	How is this taken	Side effects
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Budesonide for inhalation	Pulmicort®	Nebulized or dry powder inhaler	Lung and ear infections, runny/stuffy nose
Fluticasone	Flovent HFA®, Flovent Diskus®, Arnuity Ellipta®	Inhaler- Flovent HFA® is an aerosol, other forms are dry powder inhalers	Throat and sinus infections, joint pain, fatigue, congestion
Beclomethasone	QVAR Redihaler®	Dry powder inhaler	Runny nose, nausea, sudden airway constriction
Inhaled corticosteroids with long acting bronchodilator: These medications consist of the corticosteroid with an additional bronchodilator. This helps relax the airway to make it easier to breath throughout the day. Always rinse mouth, brush teeth, or have something to drink after use to prevent the infection known as thrush from occurring.			
Fluticasone/salmeterol	Advair®	Aerosol inhaler	Change in voice, increased risk for the common cold, headache
Mometasone/formoterol	Dulera®		
Budesonide/formoterol	Symbicort®		Increased risk for the common cold, headache

Cystic Fibrosis Transmembrane Conductance Regulators (CFTRs): These medications work to correct the defective protein that causes the symptoms of Cystic Fibrosis. Use of these medications is dependent on the specific genes that the CF patient has which cause the disease. Use of these medications may affect liver function and may cause cataracts, regular monitoring is conducted before and after starting these medications.			
Generic name	Brand name(s)	How is this taken	Side effects
Ivacaftor	Kalydeco®	Oral- tablets or granules (mixed with liquid or soft food)	Diarrhea, upset stomach, headache, rash, shortness of breath, nasal congestion, throat pain, effects on liver function
Lumacaftor/Ivacaftor	Orkambi®	Oral- tablets	Diarrhea, upset stomach, shortness of breath, effects on liver function, chest discomfort
Tezacaftor/Ivacaftor	Symdeko®	Oral- tablets	Headache, nausea, congestion

Pancreatic enzymes: These medications consist of naturally occurring proteins that aid in digestion. They are generally taken before all meals and snacks, and it is recommended that the patient always keep a supply with them. Only use the specific brand that has been prescribed and follow the recommended dosing.

Generic name	Brand name(s)	How is this taken	Side effects
Lipase/amylase/protease	Creon [®] , Pertzye [®] , Zenpep [®] , Pancreaze [®] , Ultresa [®] , Viokace [®]	Oral- Capsules or tablets (Viokace [®] only)	Upset stomach, gas, and headache



Tanner Kowalski, Pharm D.
Skywalk Pharmacy
Phone: 414-266-1893
Fax: 414-266-1894