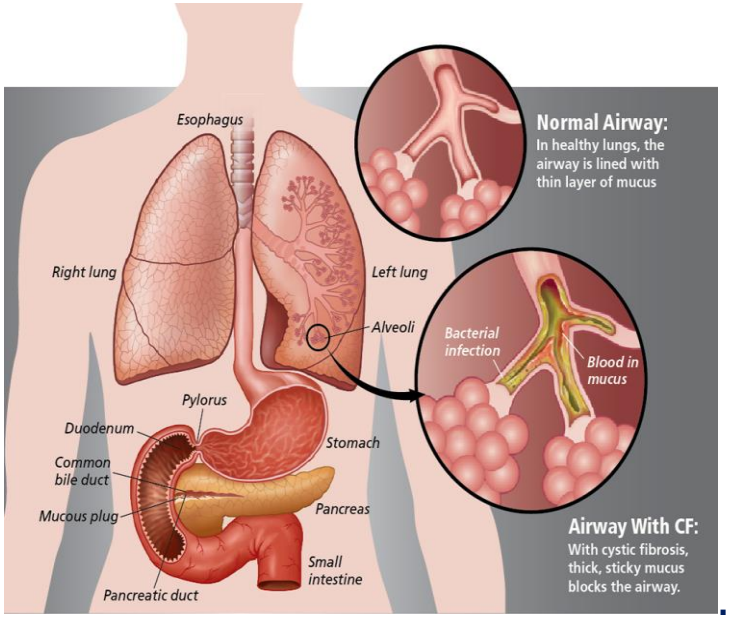
**Cystic Fibrosis: A heredity disorder affecting the exocrine glands. It causes the production of abnormally thick mucus, leading to the blockage of the pancreatic ducts, intestines, bronchi and often resulting in respiratory infection**

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# Cystic Fibrosis Self-Care

**Tampa General Hospital**
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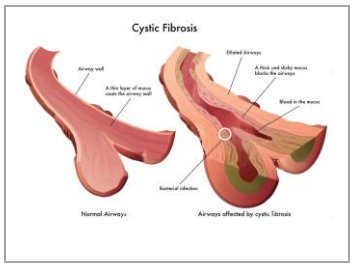
813-

# Table of Contents



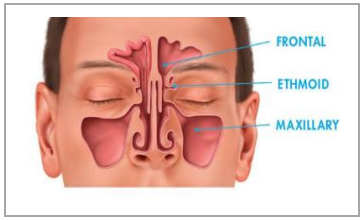
# Symptoms of Cystic Fibrosis

## Respiratory



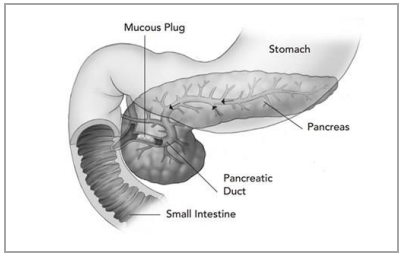

- Cough that produces thick sputum and mucous
- Wheezing and breathlessness
- Repeated lung infections

## Sinus




- Sinusitis
- Inflammation
- Nasal Polyps

## Digestive




- Intestinal blockages
- Poor weight gain and growth
- Stool changes
- Stomach ache

# Exacerbation of Symptoms

*Recognizing Pulmonary Exacerbation with Cystic Fibrosis*

- An increase in sputum amount or change in sputum color
- An increase in cough or a wheeze
- New or increased shortness of breath
- New or increased blood in sputum
- New or increased sinus pain, head congestion and headache
- Recent decrease in energy, feeling bad
- Fever, chills or sweats
- Pain in chest and/or back with or without coughing

**Call your CF doctor or care provider when symptoms present.**



# Maintenance of Lung Health

## Medications

**Bronchodilators**

**Hypertonic saline**

**Dornase Alfa**

**Aerosolized antibiotic**

**Inhaled anti-inflammatory**

## Airway Clearance Techniques


**Huff cough techniques**

**Airway clearance devices**

**Mechanical airway clearance**

**Percussion and postural drainage**

# Sequence of Therapies

**Step 1: Bronchodilator**

**Step 2: Airway clearance/ Hypertonic saline can be combined together**

**Step 3: Dornase Alfa (Pulmozyme)**

**Step 4: Aerosolized Antibiotics**

**Step 5: Inhaled Steroids**

## Step 1: Bronchodilator

- Albuterol - It works by opening breathing passages and relaxing the smooth muscles in the airway.
- It is given via Nebulizer or MDI.
- MDI- Always use a space for proper deposition of the medication.



*Airway clearance- To relieve the obstruction of the airways, as well as reduce infection and inflammation, CF patients become dependent upon cough and other techniques to clear their airways of the thick sputum. Airway clearance therapies are considered fundamental in the management of CF airways disease.*

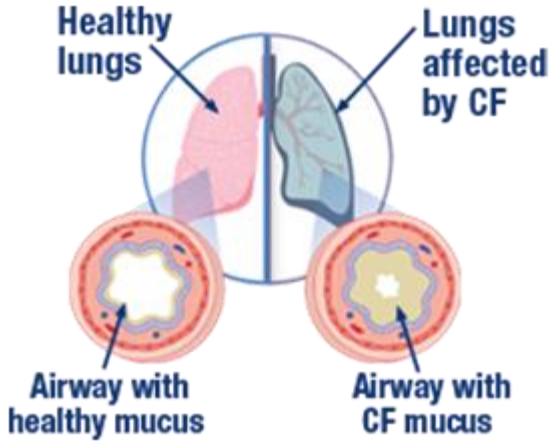
*Hypertonic saline- Inhalation of hypertonic saline increases hydration of the airway surface, thereby improving mucociliary clearance.*

## Step 2: Airway clearance/ Hypertonic saline can be combined together

- Vest 20-30 minutes at 10 Hz or more, 10 minutes or more with Hz > 17-18
- Hypertonic saline: 3% or 7%
  *Delivery device- nebulizer*

## Spilt Vest Therapy in three steps;

1. 10 min vest therapy with Hz > 10, pause therapy, Huff coughing maneuver and hydrate with water
2. 10 min vest therapy with Hz 17-18, pause therapy, Huff coughing maneuver and hydrate with water
3. 10 min vest therapy with Hz > 10, pause therapy, Huff coughing maneuver and hydrate with water




### Step 3: Dornase Alfa (Pulmozyme)

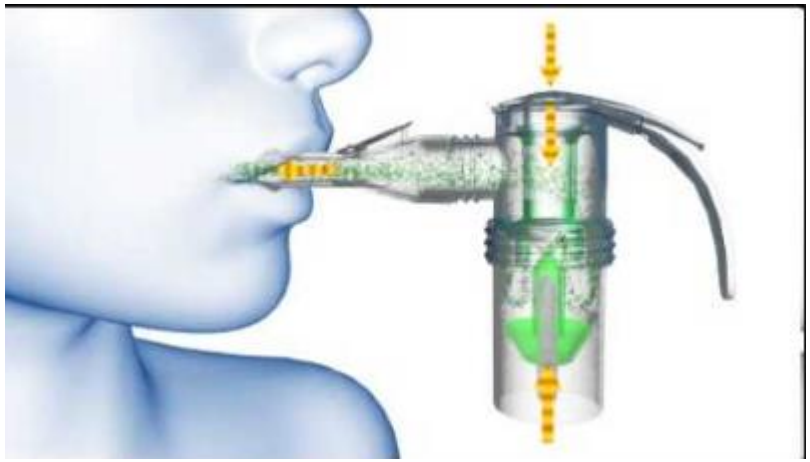
- Do not dilute or mix PULMOZYME with other drugs in the nebulizer. Mixing of PULMOZYME with other drugs could lead to adverse physicochemical and/or functional changes in PULMOZYME or the admixed compound.
- Store Pulmozyme in their protective foil pouch under refrigeration and protected from light. Refrigerate ampules during transport and do not expose to room temperature for a total time of 24 hours.
  *Delivery Device- nebulizer*

*Pulmozyme- Recombinant DNA enzyme. Given as part of a CPT regimen. Selectively cleaves extracellular DNA from pulmonary secretions, improve viscoelastic properties of secretions. Promotes airway clearance of mucous and reduces the risk of respiratory infection.*

## Step 4: Aerosolized Antibiotics

- Inhaled antibiotics should be taken, after bronchodilators (if you take them), mucus thinners, and airway clearance techniques, so your lungs will be clear of as much mucus as possible. This allows the antibiotics to reach deep into your lungs to treat the bacteria that cause infection.
- The following antibiotics are used to improve respiratory symptoms in people with CF who have Pseudomonas aeruginosa. The antibiotics especially made for people to inhale are: Aztreonam for inhalation solution (Cayston®) Tobramycin inhalation solution (such as TOBI®, Bethkis®, Kitabis Pak®).

*Delivery Device- nebulizer*



*Chronic bacterial infection is one of the key challenges in caring for patients with cystic fibrosis (CF). Effective infection control requires appropriate administration of antibiotics. Inhaled antibiotics play a pivotal role in CF management. Pathogens in CF patients target the lumen of the lung, a cavity that is difficult to access via the oral or intravenous route.*

**With a respiratory illness or change in symptoms:**

- Begin or increase airway clearance techniques.
- Use breathing treatments as ordered; you can use bronchodilators every three to four hours, and often additional Vest and/or hypertonic saline treatments are useful.
- Contact your CF doctor or nurse to see if antibiotics or additional intervention is needed.

## Step 5: Inhaled Steroids

- Reduction of lung inflammation is one of the goals of cystic fibrosis therapy. Inhaled corticosteroids are often used to treat children and adults with cystic fibrosis. The rationale for this is their potential to reduce lung damage arising from inflammation, as well as their effect on symptomatic wheezing
- Inhaled steroids are usually best given by a spacer device to reduce the amount of steroid deposited into the mouth; this can be used at all ages and is the safest way to take high doses.
- They may also be given in a dry powder form, which is suitable for those aged five years and above.
- They can also be sprayed directly into the mouth by a meter-dose inhaler, but these are more difficult to use, less efficient at getting the drugs into the lungs, and unsuitable for children.
- **The mouth must be rinsed out after taking inhaled steroids, particularly if they are taken in powder form**.

*Inhaled steroids-The most commonly-used inhaled steroids are beclometasone (Becotide, Clenil, QVAR) and budesonide (Pulmicort), which both come as beige and brown inhalers, and fluticasone (Flixotide), which is an orange inhaler. Sometimes, steroids are combined in the same inhaler with a long-acting medicine that helps relax the muscles of the airways. These are Seretide (fluticasone plus salmeterol), which is a purple inhaler, or Symbicort (budesonide plus formoterol), which is a white turbohaler with a red base.*

# Care of Equipment:

## Step 1: Cleaning
## Step 2: Disinfection
## Step 3: Storage

### Step 1: Cleaning

- Always clean the equipment in a smoke-free and dust-free location, away from open windows.
- Take apart the nebulizer cup and mouthpiece/mask and rinse with warm water and dish soap, then rise it thoroughly. **Do not** use antibacterial soap or white liquid dish soap.
- Shake off excess water and place the parts on a clean, dry towel.
- Allow all parts to air dry thoroughly.
- Alternatively, wash in the dishwasher using a dishwasher basket on the top rack.

## Step 2: Disinfection

- Clean nebulizer as indicated above.
- For low level disinfection every 3rd day mix a solution of one-part vinegar and three parts water in a clean bowl.
- Soak the nebulizer cup and mouthpiece or mask in the solution for 20 minutes.
- Rinse all parts with warm tap water and shake off all excess water, then place them on a clean, dry towel.
- Allow all parts to air dry thoroughly.
- Alternatively, nebulizer parts (except mask and tubing) may be disinfected by boiling in a clean pot of water for 5 minutes.
- Another option for disinfection is a baby bottle sterilizer.

****If using and Altera, eRapid or other eFlow nebulizers – To clean soak for 4 minutes in dish detergent (avoid white and antibacterial soaps). Disinfect by using one of two methods: boil in water for 5 minutes or a baby bottle sterilizer.**


## Step 3: Storage

- Your nebulizer must be kept clean and sterile to ensure that you receive the maximum benefit from treatments.
- Make sure all parts and accessories are clean and completely dry after use.
- Place accessories and parts in a clean plastic zipper bag or Tupperware container that is air tight.
- Designate a safe place to store your nebulizer equipment and supplies.