What is cystic fibrosis?

- People with cystic fibrosis have thick, sticky mucus which builds up and clogs some of the organs in the body, mainly the lungs and pancreas.
- When mucus builds up in the lungs, it can become difficult to breathe.
  - The thick mucus allows bacteria to get stuck in the airways.
  - This can cause repeated lung infections which can lead to lung damage.
- When mucus clogs the digestive tract and pancreas, digestive enzymes are blocked from getting to the intestines.
  - These enzymes are needed to break down food so the body can absorb nutrients in order to grow and stay healthy.

How do people get cystic fibrosis?

- Cystic fibrosis is a hereditary disease.
- It is passed down from parent to child through genes (or DNA).

Who gets cystic fibrosis?

- In the United States, approximately 30,000 people have cystic fibrosis.
- The disease is more common in people of European and Ashkenazi Jewish descent, but it can affect all races.

What are the symptoms of cystic fibrosis?

Symptoms can vary from person to person. The most common symptoms are:

- Very salty taste to the skin
- Nasal polyps - small growths in the nose
- Poor growth or inability to maintain healthy weight
- Wheezing or shortness of breath
- Persistent coughing sometimes with phlegm
- Frequent lung infections (eg, pneumonia, bronchitis)
- Greasy, foul-smelling, bulky stools or difficult bowel movements
How can I stay healthy?

Cystic fibrosis can be slowed down by staying healthy. The following are some tips:

- **Diet**
  - A high-calorie, high-fat diet is important for normal growth and development of children with cystic fibrosis.
  - The extra calories also provide extra energy needed for breathing.
  - Pancreatic enzyme supplements maybe ordered for you by your doctor. They can help your body absorb the nutrients from the foods you eat.
  - Take a daily vitamin supplement.
  - Talk to your doctor before starting any supplements.

- **Exercise**
  - Exercising can keep your lungs healthy and help you feel better.
  - Drink plenty of liquids to help loosen the mucus and to keep hydrated during and after exercise.

- **Stop the Spread of Germs**
  - Everyone in the house should wash their hands with soap and water or use sanitizing hand gel.
  - Make sure immunizations are up-to-date.
  - Get the yearly flu shot.

- **Smoke-Free Environment**
  - Smoking or being around second-hand smoke can increase cough and infections.
  - Second-hand smoke can also slow down lung growth in children.

- **Keep Up with Your Medical Treatments**
  - Stay in contact and talk to your doctor about any of your concerns or questions.
  - Make sure you make all your appointments with your team of health care providers (eg, doctors, nurses, and respiratory therapists).
  - Take all your medications.
What treatments are available for cystic fibrosis?

There is no cure for cystic fibrosis. But, treatment has improved greatly over the years. A major part of treatment is to clear mucus from the airways using airway clearance techniques (ACT). Airway clearance techniques create vibrations in the lungs to help loosen the mucus so it can be coughed out.

Medications are used to treat lung infections and help people breathe better. These medications include:

- Mucus thinning drugs – These medications thin the mucus making it easier to be cleared from the lungs (eg, Pulmozyme).
- Antibiotics – These medications fight the bacteria that cause lung infections (eg, TOBI, Cayston).
- Bronchodilators – These medications open the airways to make breathing easier (eg, albuterol, Foradil®, Serevent®).
- Anti-inflammatory drugs – These medications reduce inflammation in the lungs (eg, ibuprofen).

What is Pulmozyme?

- Pulmozyme is a medication that is used in cystic fibrosis to help people breathe better.
- Pulmozyme helps to break up any thick mucus that builds up in the lungs.

What are TOBI and Cayston?

- TOBI and Cayston are antibiotics which kill a bacteria called *Pseudomonas aeruginosa*.
- In cystic fibrosis patients, *Pseudomonas aeruginosa* is the main cause of lung infections and lung damage.
- To reduce the development of drug-resistance, TOBI and Cayston should only be used to treat cystic fibrosis patients with known *Pseudomonas aeruginosa* in their lungs.

How is Pulmozyme stored?

- Pulmozyme should be stored in its foil pouch in the refrigerator (36-46°F/2-8°C).
- Do not freeze.
- Protect Pulmozyme from intense light and excessive heat.
- Do not use Pulmozyme if it has been left at room temperature for longer than 24 hours.
- Do not use if it becomes cloudy or discolored.
- Do not use it after the expiration date.
**How are TOBI and Cayston stored?**

- Store TOBI and Cayston in the refrigerator (36-46°F/2-8°C).
- If a refrigerator is not available, TOBI and Cayston can be stored at room temperature (up to 77°F/25°C) for up to 28 days.
  - TOBI will turn slightly yellow and darken with time if unrefrigerated. This doesn’t change the quality of the drug.
- Protect TOBI and Cayston from intense light.
- Do not use TOBI and Cayston if it has been left at room temperature for more than 28 days.
- Do not use TOBI and Cayston if it becomes cloudy or if there are particles in the solution.
- Do not use it after the expiration date.

**What are some things I should know about my medications?**

- Do not mix TOBI and Cayston with any other medications in your nebulizer.
- TOBI is used twice a day (every 12 hours) and Cayston is used 3 times a day. Both medications are used for a 28-day cycle (followed by 28 days off).
- Short-acting bronchodilators (eg, albuterol) can be taken between 15 minutes and 4 hours before TOBI or Cayston.
- Long-acting bronchodilators (eg, Foradil®, Serevent®) can be taken between 30 minutes and 12 hours prior to TOBI or Cayston.
- If you are taking several medications, this is the recommended order:
  1. Bronchodilator (eg, albuterol)
  2. Airway clearance techniques
  3. Other inhaled medications (eg, Pulmozyme)
  4. TOBI or Cayston

**What other resources are available?**

For more information, please contact the following resources.

- **OptumRx**
  Phone: 1-800-562-6223
  Web site: www.optumrx.com

- **TOBI Patient Assistance**
  Phone: 1-866-598-8624
  Web site: www.tobitime.com
- **Cayston Access Program**  
  Phone: 1-877-7-CAYSTON or 1-877-722-9786  
  Web site: www.cayston.com

- **Pulmozyme Access Solutions**  
  Phone: 1-800-690-3023  
  Web site: www.genentechaccesssolutions.com/pulmozyme/patient/index.jsp

- **Cystic Fibrosis Foundation**  
  Phone: 1-800-FIGHT-CF or 1-800-344-4823  
  Web site: www.cff.org

- **The Cystic Fibrosis Center at Stanford**  
  Phone: 1-650-497-8841  
  Web site: www.cfcenter.stanford.edu

- **MedlinePlus**  
  Web site: www.medlineplus.gov

**References:**


The information in this educational handout doesn’t substitute the medical advice, diagnosis, or treatment provided by your doctor. Always seek the help of your doctor or a qualified health provider for any questions about your medical condition.