

UNDERSTANDING CYSTIC FIBROSIS

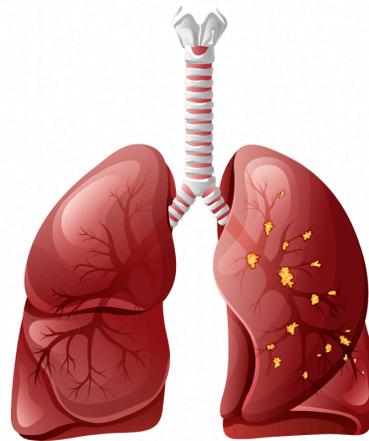


A Patient Guide



UNDERSTANDING CYSTIC FIBROSIS

A Patient Guide



This booklet will help you learn about cystic fibrosis (CF). You will find out what causes it, how it is treated, and what you can do to stay as healthy as possible.

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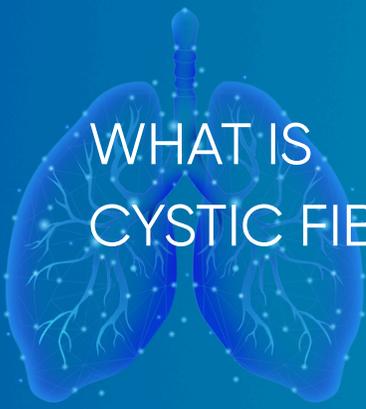
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WHAT IS CYSTIC FIBROSIS?



Cystic fibrosis (CF) is a genetic disease that affects the lungs and digestive system. It causes the body to make thick, sticky mucus that clogs the lungs and blocks the pancreas.

In healthy people, mucus is thin and slippery. It keeps the lungs, airways, and digestive system moist and protected. In people with CF, the mucus is too thick. This makes it hard to breathe and digest food properly.

Think of it this way: Imagine trying to breathe through a straw filled with honey. That's what it can feel like for someone with CF. The thick mucus traps germs and makes lung infections more likely.

Key Terms to Know

Cystic Fibrosis (CF): A genetic disease that causes thick, sticky mucus to build up in the lungs and digestive system.

Mucus: A slippery substance that normally protects and moistens the airways and organs.

CFTR Gene: The gene that is changed (mutated) in people with CF. It controls the flow of salt and water in cells.

References:

1. Cystic Fibrosis Foundation. "About Cystic Fibrosis." CFF.org. <https://www.cff.org/What-is-CF/About-Cystic-Fibrosis/>
2. Mayo Clinic. "Cystic Fibrosis." MayoClinic.org. <https://www.mayoclinic.org/diseases-conditions/cystic-fibrosis/symptoms-causes/syc-20353700>
3. National Heart, Lung, and Blood Institute. "Cystic Fibrosis." NHLBI, NIH. <https://www.nhlbi.nih.gov/health/cystic-fibrosis>



HOW CF AFFECTS THE BODY



CF affects many parts of the body. The thick mucus can cause problems in several organs:

LUNGS AND AIRWAYS

Thick mucus clogs the airways, making it hard to breathe. Mucus traps bacteria, leading to frequent lung infections. Over time, lung damage can occur.

PANCREAS

Mucus blocks the pancreas from releasing enzymes needed to digest food. This makes it hard for the body to absorb nutrients from food.

LIVER

Thick mucus can block bile ducts in the liver, which may cause liver problems over time.

SWEAT GLANDS

People with CF have very salty sweat. This is why the sweat test is used to diagnose CF.

REPRODUCTIVE SYSTEM

CF can affect fertility in both men and women, though many people with CF can still have children.

Important: CF affects each person differently. Some people have mild symptoms while others have more severe disease. Working closely with your CF care team is key to staying healthy.

References:

1. Cystic Fibrosis Foundation. "How CF Affects the Body." CFF.org. <https://www.cff.org/What-is-CF/How-Cystic-Fibrosis-Affects-the-Body/>
2. Mayo Clinic. "Cystic Fibrosis - Symptoms and Causes." MayoClinic.org.



SIGNS & SYMPTOMS



Symptoms can vary from person to person.	They may be mild or severe:
Respiratory Symptoms	<ul style="list-style-type: none">• Persistent cough with thick mucus• Wheezing or shortness of breath• Frequent lung infections (pneumonia, bronchitis)• Stuffy nose or sinus infections
Digestive Symptoms	<ul style="list-style-type: none">• Greasy, bulky, foul-smelling stools• Difficulty gaining weight or growing• Intestinal blockage (especially in newborns)• Constipation or stomach pain
Other Symptoms	<ul style="list-style-type: none">• Very salty-tasting skin• Clubbing (rounding) of fingers and toes• Fatigue• Nasal polyps
In Newborns	<ul style="list-style-type: none">• Meconium ileus (blocked intestine at birth)• Failure to thrive• Prolonged jaundice

Important: Most states now screen newborns for CF. Early diagnosis leads to better outcomes.

References:

1. Cystic Fibrosis Foundation. "Symptoms of CF." CFF.org.
2. Mayo Clinic. "Cystic Fibrosis - Symptoms." MayoClinic.org.



CAUSES AND RISK FACTORS



What Causes CF?

CF is caused by changes (mutations) in the CFTR gene. This gene tells the body how to make a protein that controls the movement of salt and water in and out of cells.

When the CFTR gene doesn't work properly, mucus becomes thick and sticky instead of thin and slippery.

How is CF Inherited?

CF is inherited in an autosomal recessive pattern. This means:

- A child must inherit TWO copies of the faulty gene (one from each parent)
- If a child inherits only ONE copy, they are a "carrier" but don't have CF
- When both parents are carriers, each pregnancy has a 25% chance of CF

Risk Factors

- Family history of CF
- Both parents are CF carriers
- Ethnicity: CF is most common in people of Northern European descent, but it occurs in all races

Genetic Testing

Carrier testing is available for people planning to have children. More than 10 million Americans are CF carriers and most don't know it.

References:

1. Cystic Fibrosis Foundation. "CF Genetics: The Basics." CFF.org. <https://www.cff.org/What-is-CF/Genetics/CF-Genetics-The-Basics/>
2. National Human Genome Research Institute. "Cystic Fibrosis." Genome.gov.



DIAGNOSIS



How is CF Diagnosed?

Several tests are used to diagnose CF:

Newborn Screening

What It Is: A blood test done on all newborns in the U.S.

What It Checks: Levels of a chemical called IRT (immunoreactive trypsinogen)

Why It Matters: High IRT may indicate CF and leads to more testing

Sweat Test (Gold Standard)

What It Is: Measures the amount of salt in sweat

How It Works: A small amount of sweat is collected from the arm

Why It Matters: People with CF have very salty sweat (high chloride levels)

Genetic Testing

What It Is: A blood or saliva test

What It Checks: Looks for mutations in the CFTR gene

Why It Matters: Identifies the specific gene mutations, which helps guide treatment

Other Tests

- Lung function tests (spirometry)
- Chest X-ray or CT scan
- Sputum culture (to check for bacteria)

References:

1. Cystic Fibrosis Foundation. "Diagnosing Cystic Fibrosis." CFF.org.
2. Mayo Clinic. "Cystic Fibrosis - Diagnosis." MayoClinic.org.



WHAT TO EXPECT



Disease Course

CF is a lifelong condition, but treatments have improved greatly. Many people with CF now live into their 40s, 50s, and beyond.

CF is a Progressive Disease

This means it can get worse over time. However, following your treatment plan closely can slow progression and help you stay healthier longer.

Lung Health is Key

- Lung function may decline over time
- Preventing infections is critical
- Daily treatments help keep lungs clear

Treatment is Daily and Time-Consuming

- Many people spend 2-4 hours per day on treatments
- Consistency is important for best results
- Your CF care team will help you manage

Advances in Treatment

New medications called CFTR modulators have changed the outlook for many people with CF. These drugs treat the underlying cause of CF, not just the symptoms.

Co-Morbid Conditions

People with CF may also develop:

- CF-related diabetes
- Osteoporosis (weak bones)
- Liver disease
- Mental health challenges (anxiety, depression)

References:

1. Cystic Fibrosis Foundation. "Life with CF." CFF.org.



TREATMENT OPTIONS



Treatment focuses on keeping the lungs clear, preventing infections, and maintaining good nutrition.

CFTR MODULATORS	These newer drugs fix the faulty CFTR protein. They treat the root cause of CF and have greatly improved outcomes for many patients.
INHALED MEDICATIONS	Antibiotics: Fight lung infections (like inhaled tobramycin) Mucolytics: Thin the mucus so it's easier to cough up Bronchodilators: Open the airways to help breathing.
AIRWAY CLEARANCE	Techniques and devices that help loosen and remove mucus from the lungs. Usually done 1-4 times daily.
PANCREATIC ENZYMES	Capsules taken with meals and snacks to help digest food and absorb nutrients.
NUTRITIONAL SUPPORT	High-calorie diet, vitamins, and sometimes feeding tubes to maintain healthy weight.
LUNG TRANSPLANT	For severe lung disease, a lung transplant may be an option.

References:

1. Cystic Fibrosis Foundation. "CF Treatments." CFF.org. <https://www.cff.org/Life-With-CF/Treatments-and-Therapies/>
2. Mayo Clinic. "Cystic Fibrosis - Treatment." MayoClinic.org.



AIRWAY CLEARANCE TECHNIQUES



Clearing mucus from the lungs is one of the most important parts of CF care:

Chest Physical Therapy (CPT)	A caregiver claps on the chest and back to loosen mucus. Takes about 20-30 minutes.
High-Frequency Chest Wall Oscillation (The Vest)	An inflatable vest that vibrates to shake mucus loose. Can be done independently.
Positive Expiratory Pressure (PEP) Devices	Handheld devices you breathe into that create pressure to help move mucus.
Active Cycle of Breathing Technique (ACBT)	A series of breathing exercises that help move mucus without equipment.
Autogenic Drainage	A breathing technique that uses different breath depths to move mucus up and out.
Exercise	Physical activity helps loosen mucus and strengthen the lungs. It's an important part of CF care.

Important: Your CF care team will help you find the best airway clearance routine for your lifestyle.

¹References:

1. Cystic Fibrosis Foundation. "Airway Clearance Techniques." CFF.org.



NUTRITION AND CF



Good nutrition is essential for people with CF. The body needs extra calories to fight infections and breathe.

Why Nutrition Matters

- CF makes it harder to absorb nutrients from food
- The body burns more calories fighting infections
- Good nutrition helps maintain lung function
- Healthy weight is linked to better outcomes

High-Calorie, High-Fat Diet

Unlike most people, those with CF often need:

- More calories than average (sometimes 1.5-2x normal)
- High-fat foods
- Extra salt (especially in hot weather)
- Frequent meals and snacks

Pancreatic Enzymes

- Take enzymes with ALL meals and snacks containing fat
- Swallow capsules whole or sprinkle on soft food
- Your doctor will adjust the dose as needed

Vitamins

People with CF often need fat-soluble vitamin supplements (A, D, E, K) because these are harder to absorb.

References:

1. Cystic Fibrosis Foundation. "Nutrition." CFF.org. <https://www.cff.org/Life-With-CF/Daily-Life/Fitness-and-Nutrition/Nutrition/>



TIPS FOR TAKING YOUR MEDICATION



DO:

- Take medications in the correct order (ask your CF team)
- Clean and sterilize nebulizers after each use
- Take enzymes with every meal and snack containing fat
- Store medications properly (some need refrigeration)
- Keep track of when to refill prescriptions
- Complete the full course of antibiotics
- Do airway clearance before inhaled antibiotics

Typical Daily Treatment Order:

1. Bronchodilator (opens airways)
2. Mucolytic (thins mucus)
3. Airway clearance (clears mucus)
4. Inhaled antibiotic (fights infection)

Note: Your CF team will give you a personalized treatment order.



DON'T:

- Skip treatments, even when feeling well
- Share nebulizers or medications with others
- Use tap water to clean nebulizers (use sterile or distilled)
- Stop medications without talking to your doctor
- Crush enzymes or mix with hot food

References:

1. Cystic Fibrosis Foundation. "Therapies for Cystic Fibrosis." CFF.org.



WHEN TO CALL YOUR DOCTOR



! CALL 911 OR GO TO THE EMERGENCY ROOM IF YOU HAVE:

- Severe trouble breathing
- Coughing up large amounts of blood
- Chest pain
- Blue lips or fingernails
- Confusion or extreme drowsiness

! CALL YOUR CF CARE TEAM IF:

- Increased cough or change in mucus color
- Fever over 101°F (38.3°C)
- More shortness of breath than usual
- Weight loss or decreased appetite
- Blood in mucus (even small amounts)
- New or worsening sinus symptoms
- Signs of CF-related diabetes (increased thirst, urination)
- Medication side effects
- Feelings of depression or anxiety



Keep Your Clinic Appointments!

Regular visits (usually every 3 months) help catch problems early.

References:

1. Cystic Fibrosis Foundation. "CF Care Guidelines." CFF.org.



GLOSSARY



AIRWAY CLEARANCE	Techniques to help remove mucus from the lungs
BRONCHODILATOR	Medication that opens the airways
CF	Cystic fibrosis
CFTR	The gene and protein affected in cystic fibrosis
CFTR MODULATOR	Medication that fixes the faulty CFTR protein
EXACERBATION	A flare-up or worsening of symptoms
MECONIUM ILEUS	Blocked intestine in newborns with CF
MUCOLYTIC	Medication that thins mucus
MUCUS	Slippery substance that protects airways and organs
NEBULIZER	Device that turns liquid medicine into a mist to breathe
PANCREATIC ENZYMES	Supplements that help digest food
PULMONARY	Related to the lungs
SPUTUM	Mucus coughed up from the lungs
SWEAT TEST	Test that measures salt in sweat to diagnose CF



RESOURCES



Learn More:

- Cystic Fibrosis Foundation: cff.org
- CF Foundation Care Center Network: cff.org/Care-Centers
- National Heart, Lung, and Blood Institute: nhlbi.nih.gov/health/cystic-fibrosis
- Mayo Clinic - Cystic Fibrosis: mayoclinic.org/diseases-conditions/cystic-fibrosis
- MedlinePlus - Cystic Fibrosis: medlineplus.gov/cysticfibrosis.html
- American Lung Association: lung.org



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