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Cystic Fibrosis

What is Cystic Fibrosis?

Cystic fibrosis¹ (CF) is a disease that causes breathing and digestive problems. This disease is **progressive**², which means it gets worse over time. A person **inherits**³ this disease, meaning they get it from their parents. To get CF, you must get two copies of a bad gene, one from each parent. The parents probably don't have the disease themselves, but are **carriers**⁴ for it.

Did You Know?

Over 70,000 people worldwide have cystic fibrosis.

What Causes Cystic Fibrosis (CF)?

A bad protein called **Cystic Fibrosis Transmembrane Regulator**⁵ (CFTR) causes CF. Because this

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What are Some Symptoms of Cystic Fibrosis?

Cystic fibrosis makes sweat and skin taste very salty. It can also cause you to feel tired and weak. It leads to low blood pressure, high heart rate, and dehydration (not having enough water).



CF makes you feel tired.

¹ **Cystic fibrosis**: an inherited disease that causes breathing and digestive problems and gets worse over time.

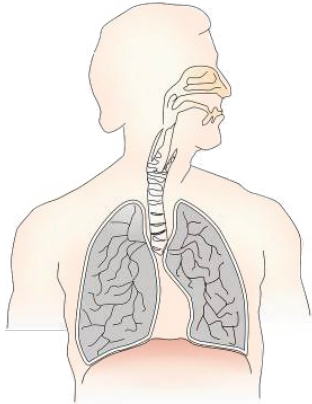
² **Progressive disease**: a disease that gets worse over time.

³ **Inherit**: to get something from you parents.

⁴ **Carriers**: people who do not have a disease, but carry the genes for a disease and can pass the disease on to their kids.

⁵ **Cystic Fibrosis Transmembrane Regulator**: a bad protein that causes cystic fibrosis.

In the respiratory (breathing) system, blocked airways can get swollen and painful. CF also causes a bad cough that won't go away and sometimes people cough up **phlegm**⁶. People also wheeze and get out of breath. Bacteria in the lungs can cause serious infections (e.g. bronchitis, pneumonia). Infections return over and over, causing permanent lung damage and **respiratory failure**⁷. In fact, respiratory failure is the leading cause of death in people with CF.



Mucus buildup in the lungs causes difficulty breathing and infections.

Reduced insulin can cause diabetes.

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How is Cystic Fibrosis Diagnosed?

Doctors test people's sweat to see if they have CF. In the lab, they measure the levels of chloride ions in sweat.

How is Cystic Fibrosis Treated?

There is no cure for CF yet, but there are several therapies.

⁶ **Phlegm**: a thick gel-like liquid that can be yellow, green, brown, or gray.

⁷ **Respiratory failure**: when you can't get enough oxygen.

People with CF must have therapy every day to help with some of their symptoms. They have to clear their airways and take medicines that help them get nutrients from their food. They must take this medicine every time they eat.



Even though there is no cure yet for cystic fibrosis, daily therapy helps people live longer.

These daily therapies have allowed people with CF to live much longer than they used to. In the 1950s, a child with CF would be lucky to live long enough to attend elementary school. With daily therapy, people with CF are living into their 40s or even 50s.

Some companies are working on medications to treat CF. They aren't cures, but they make life much easier for people with CF and will help them live longer, fuller lives. It will be a while before they are ready for people to use, though.



Drug companies are developing CF medicines.

Reading Comprehension Questions:

1. What
2. What causes cystic fibrosis?
3. How
4. How does cystic fibrosis affect the respiratory system?
5. How does
6. How do the drugs Kalydeco and Orkambi treat cystic fibrosis?

Extension Questions:

7. How might parents obtain a clue to whether their child has CF?
8. What

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