Overview

Cystic fibrosis

Cystic fibrosis causes sticky mucus to build up in the lungs and digestive system. This causes lung infections and problems digesting food.

It's a condition some people are born with. It runs in families. The heel prick test babies have when they're born picks up most cases of cystic fibrosis.

Symptoms usually start in early childhood. They vary from child to child.

The condition gets worse over time. The lungs and digestive system become more and more damaged.

Treatments are available to help reduce the problems caused by the condition. These make it easier to live with cystic fibrosis. But life expectancy is short.

Symptoms of cystic fibrosis

The build-up of sticky mucus in the lungs can cause breathing problems. It also increases the risk of lung infections. Over time, the lungs may stop working the way they should.

Mucus also clogs the pancreas (the organ that helps with digestion). This stops enzymes reaching food in the gut and affects digestion.

This means you can't absorb all the nutrients from food. You need to eat more calories to avoid malnutrition.

Symptoms of cystic fibrosis include:

- chest infections that come and go
- wheezing, coughing, shortness of breath and damage to the airways (bronchiectasis)
- difficulty putting on weight and growing
- jaundice
- diarrhoea, constipation, or large, smelly poo
- a bowel blockage in newborn babies (meconium ileus) – they may need surgery

You can also develop a number of related conditions.

These include:

- diabetes
- thin and weakened bones (osteoporosis)
- infertility in males
- liver problems

Diagnosing cystic fibrosis

All newborn babies are screened for cystic fibrosis as part of the heel prick screening test. This is done shortly after birth.

If the test shows a child may have cystic fibrosis, they'll have more tests to check for sure.

The tests include:

- a sweat test – this measures the amount of salt in sweat which is very high in someone with cystic fibrosis
- a genetic test – a blood or saliva sample to check for the faulty gene that causes cystic fibrosis

These tests can also diagnose cystic fibrosis in older children and adults. You might have them if you never had the heel prick test.

The genetic test can also be used to see if someone is a "carrier" of cystic fibrosis. This can be important for someone who thinks they may have the faulty gene and would like to have children.

Treatments for cystic fibrosis

There's no cure for cystic fibrosis.

But some treatments can help to:

- control symptoms
- prevent or reduce complications

This can make the condition easier to live with.

You may need to take different medicines to treat and prevent lung problems.

Staying active and clearing your airways may help clear mucus from the lungs.

Read more about treatments for cystic fibrosis

Complications of cystic fibrosis

If you have cystic fibrosis, you have a higher risk of developing other conditions.

These include:

- weak and brittle bones (osteoporosis) – medicines called bisphosphonates can sometimes help
- diabetes – you may need insulin and a special diet to control blood sugar levels
- nasal polyps and sinus infections – steroids, antihistamines, antibiotics or sinus flushes can help
- liver problems

You are also more likely to pick up infections and have complications from to an infection. This includes COVID-19 (coronavirus).

> **Important**
>
> If you have cystic fibrosis, do not meet other people with cystic fibrosis face-to-face. You may pass on or pick up an infection.

Fertility problems

It's possible for women with cystic fibrosis to have children.

97% of men with cystic fibrosis are born without a vas deferens (part of the male reproductive system). This means they will need fertility treatment. But 3% have a normal vas deferens and will be fertile without needing help.

Cause of cystic fibrosis

Cystic fibrosis is caused by a faulty gene passed through families.

It affects the movement of salt and water in and out of cells in the body. This can cause a build-up of thick, sticky mucus in the body's tubes and passageways. This usually happens in the lungs and digestive system.

A person with cystic fibrosis is born with the condition. It's not possible to catch cystic fibrosis from someone else who has it.

How cystic fibrosis is inherited

To be born with cystic fibrosis, you have to inherit a copy of the faulty gene from both your parents.

This can happen if your parents are "carriers" of the faulty gene. This means your parents do not have cystic fibrosis themselves.

It's estimated around 1 in 19 people in Ireland are carriers of cystic fibrosis.

If both parents are carriers, there's a:

- 1 in 4 chance their child won't inherit any faulty genes and won't have cystic fibrosis or be able to pass it on
- 1 in 2 chance their child will inherit a faulty gene from one parent and be a carrier
- 1 in 4 chance their child will inherit the faulty gene from both parents and have cystic fibrosis

If one parent has cystic fibrosis and the other is a carrier, there's a:

- 1 in 2 chance their child will be a carrier
- 1 in 2 chance their child will have cystic fibrosis

Living with cystic fibrosis

Cystic fibrosis tends to get worse over time. It can be fatal if it leads to a serious infection or the lungs stop working properly.

People with cystic fibrosis are now living longer because of advancements in treatment.

About half of people who now have cystic fibrosis will live past the age of 40. Children born with the condition nowadays are likely to live longer than this.

Support

Support is available to help people with cystic fibrosis live as independently as they can.

It can be helpful to speak to others who have the same condition, and to connect with a charity.

Cystic Fibrosis Ireland provides information and resources for people living with cystic fibrosis.

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