Patient information from BMJ

Last published: Jul 08, 2020

Cystic fibrosis: what treatments work?

Cystic fibrosis is a genetic (inherited) condition that is present from birth. It affects the lungs and digestive system. Treatments can help ease the symptoms and prevent complications but the condition usually gets worse as people get older. There is no cure and people with cystic fibrosis have a reduced life expectancy.

What treatments work?

Cystic fibrosis (CF) affects the lungs and digestive system by causing them to become clogged with thick mucus so that they can't work properly. For more background information, see our leaflet *Cystic fibrosis: what is it?*

There is no cure for CF. Instead, the aims of treatment are to ease the condition as much as possible by:

- doing as much as possible to prevent common health problems linked to CF
- dealing with the complications of CF, such as issues with nutrition, and lung problems.

Treatments for lung problems

Lung problems are the most common cause of difficulty for people with CF. If the lungs become badly congested (clogged) there are several ways of helping to clear them, including physical treatments and drug treatments.

Physical treatments include:

- manual physiotherapy. This involves a physiotherapist helping you get into positions where mucus can drain from the lungs more easily, then gently tapping parts of the body to loosen the mucus
- special breathing exercises
- 'vest' therapy. You wear a special inflatable vest that vibrates gently to loosen mucus
- a flutter valve, which is a small breathing device that makes the lungs vibrate to loosen mucus. You use it whenever you need to

- a positive expiratory pressure (PEP) mask or mouthpiece. The device makes you work harder to breathe out, which helps loosen mucus.

Drug treatments that you breathe in through an inhaler include:

- mucolytics. These are drugs that loosen mucus
- bronchodilators. These are drugs that help open up your airways
- antibiotics. Your doctor might suggest these if you have a long-term chest infection
- corticosteroids (steroids for short). These are drugs that relieve inflammation (swelling) in the lungs. These tend to be used in people who have asthma as well as CF.

Oral drug treatments (tablets and medicines that you swallow) include:

- corticosteroids. As with the inhaled version, these are often only recommended for people with other chest problems such as asthma
- antibiotics to help prevent chest infections
- anti-inflammatory drugs such as ibuprofen.

Newer drugs called CF transmembrane conductance regulator (CFTR) modulators are intended to treat the underlying causes of CF rather than the symptoms. But they are not available everywhere and they only work for people with certain gene problems.

A **lung transplant** operation might be an option for some people if all other treatments don't work well enough. But it can take a long time for suitable lungs to become available, if they become available at all.

Lung transplants are also not safe for people with CF who have certain other health problems such as multiple organ problems, and severe gastro-oesophageal reflux.

Treatments for nutrition problems

People with CF need to take supplements that contain:

- vitamins that they can't absorb well enough from food, and
- enzymes that help them to absorb nutrition from food.

You take these supplements before meals. A nutritionist will explain when and how to take them, and how much you need at different times.

Treatments for intestine (gut) blockages

People with CF can have various digestive problems, including diarrhoea and gastro-oesophageal reflux. But the intestine can also sometimes become partially blocked with mucus. This is treated with enemas.

After an episode of intestinal blockage some people may need to take medicines for a while, such as laxatives, or medicines that soften the stool. This can help stop the problem happening again.

If your intestine becomes completely blocked you may need to have surgery to remove the blockage. But this is rare.

Other problems

Some people with CF also need to take medicines to help with gastro-oesophageal reflux. And about 1 in 3 people with CF also develop liver disease and need to take medicine to treat it. Some people with CF will develop advanced liver disease, which can be fatal.

What will happen?

CF is a serious condition that shortens people's lives. There is no cure. But the outlook is changing.

A few decades ago most people with CF died in childhood. Most people with CF now live into their late 30s, and many live much longer. And advances in treatments are still being made.

The ongoing care of people with CF is also improving. If you have CF you should be seen in a specialist centre every 3 months, or more often depending on your health. You can discuss how you are doing with a doctor and nutritionist and have tests to check if you are getting the right nutrition and how well your lungs are working.

One thing that has been shown to help people with CF stay healthier and live longer is regular exercise. Exercise helps keep the lungs working well. You can talk with your doctor about what type and how much exercise you (or your child) should do. But staying as active as possible is a good rule of thumb.

There are various charities and support groups to help people with CF and the parents of children with the condition. You should be able to find them easily online. For example, in the UK, the Cystic Fibrosis Trust (cysticfibrosis.org.uk) provides information and help with many aspects of the condition.