



The new name for the British Liver Trust
and Children's Liver Disease Foundation

Cystic fibrosis and liver disease in children



Help, support and guidance
for children and families

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Welcome

Welcome to your guide to understanding **cystic fibrosis-related liver disease**. It provides information on causes, symptoms, diagnosis, complications and treatment.

This information has been written for:

- parents and carers of children and young people with cystic fibrosis

Others who may also find this information useful:

- young people with cystic fibrosis
- healthcare professionals

You may also find it helpful to read the following booklets:

- Introduction to liver disease in children
- Nutrition
- Portal hypertension and ascites
- Liver transplantation

Use this QR code to view our resources or visit **www.liveruk.org**



Key facts about CF-related liver disease

1

Cystic fibrosis is a common genetic condition.

6

The symptoms and severity can vary from child to child.

2

Cystic fibrosis can make bile in the body become thicker. This may lead to blockage of the bile ducts, which can damage the liver.

7

In a small number of cases, damage to the liver can become worse over time.

3

Most children and young people with cystic fibrosis will not develop any serious problems with their liver.

8

There is no cure, but dietary support, medicines and medical procedures can help reduce the symptoms and complications.

4

We cannot predict who will get cystic fibrosis-related liver disease.

9

Only a small number of children develop advanced liver disease that will lead to a liver transplant.

5

Tests for cystic fibrosis-related liver disease include physical examination, blood tests, scans and occasionally liver biopsy.



What is cystic fibrosis?

Cystic fibrosis (CF) is a condition in which thick, sticky mucus builds up in the body's tubes and passageways. The mucus causes problems in the lungs, pancreas, digestive system, liver, and sweat glands.

Cystic fibrosis is a common genetic condition. This means it is caused by changes in a person's genes.

Currently, around 11,100 people in the UK have cystic fibrosis. It is estimated that 1 in every 2,500 babies born in the UK will have the condition.

The main symptoms of cystic fibrosis are:

- lung infections
- frequent cough
- shortness of breath
- trouble gaining weight

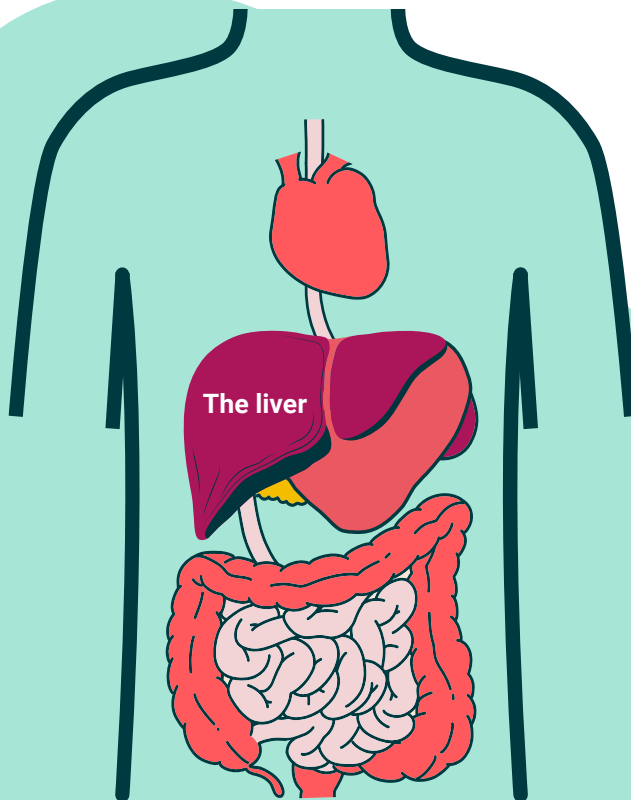


How can cystic fibrosis affect the liver?

The liver is a large organ found at the top of the tummy (abdomen) on the right side. It is made up of cells (hepatocytes), blood vessels and bile ducts.

The liver has many important jobs in the body including:

- cleaning (filtering) the blood
- making blood plasma proteins, clotting factors, cholesterol, vitamin D, hormones and bile
- processing waste products



People with cystic fibrosis have a gene which means they can't move salt and water between their cells properly. This causes fluids in the body to become thicker. The fluids can block the body's tubes and passageways.

In some people, cystic fibrosis affects bile. This is a fluid made by the liver. The bile becomes thicker than normal and blocks the bile ducts. Blocked bile ducts can cause damage and scarring to the tissues around them. This is called biliary fibrosis. It is like the damage which can happen in the lungs of people with cystic fibrosis.



**My son had no symptoms of liver disease...
no one had mentioned livers to us before, so
it was quite a shock.**

Parent

In some people, the damage spreads throughout the liver as the cells start to die. The healthy cells are replaced with scar tissue (fibrosis). This makes it hard for blood to flow through the liver. It may eventually lead to severe scarring of the liver, known as cirrhosis.

If cystic fibrosis affects the liver, it is called cystic fibrosis-related liver disease (CFLD).



Use this QR code to find out
more about the liver or visit
liveruk.org/about-the-liver/



Which children with cystic fibrosis get liver disease?

Around 40 out of 100 people with cystic fibrosis have changes involving their liver.

These changes are often mild. Most people show no signs or symptoms and do not develop serious liver problems.

Around 5-10 out of 100 children with cystic fibrosis develop liver problems that have an impact on their health.

Cystic fibrosis-related liver disease usually develops in the first 10 years of life. We do not know who will be affected. We also don't know why some people with cystic fibrosis get liver disease and some do not.



The amount of damage from cystic fibrosis-related liver disease can vary a lot from person to person.

Head of Children's Services, Liver UK

Some research says that cystic fibrosis-related liver disease may be more common in:

- boys
- children who had a condition called meconium ileus as a baby. This is where a baby's first poo (stool) gets stuck in the small intestine.
- children and young people who have the SERPINA1 gene. This is a gene for another condition, called alpha-1 antitrypsin deficiency, which can also affect the lungs and liver.

How is cystic fibrosis-related liver disease diagnosed?

There are no tests that can tell us who will get cystic fibrosis-related liver disease. It is also hard to catch early because there are often no signs or symptoms. This is why regular check-ups are so important.

Children and young people with cystic fibrosis have annual check-ups. This includes tests to look for liver disease. The medical team want to find any problems at an early stage. This will give them more time to slow down the disease and stop any damage to the liver getting worse.

Tests used to check the liver may include:

Physical examination

A doctor will check your child's body for the following:

- swelling of the liver (hepatomegaly)
- swelling of the spleen (splenomegaly)
- visible veins in the tummy (abdominal) wall
- yellowing of the skin and whites of the eyes (jaundice)



Liver blood tests

To do these tests, a blood sample is taken and tested in a laboratory. They help to see if the liver is injured and how well it is working. The tests are also used to monitor liver disease over time. They can help to see if the liver is getting healthier, getting worse or staying the same.

Abdominal ultrasound scan

An abdominal ultrasound scan uses sound waves to create a picture of the inside of the body. It may be used to check and monitor:

- the size and texture of the liver
- the size of the spleen
- the size of the gallbladder
- blood flow into and out of the liver
- swelling, stones or blockages in the bile ducts

Liver biopsy

A liver biopsy is not commonly used for diagnosing cystic fibrosis-related liver disease. It is only done if there is doubt over the diagnosis or if there may be another cause of the liver disease.

During this test a very thin needle is inserted into the tummy (abdominal) wall and into the liver. The needle takes a small sample of liver tissue. This is sent to a laboratory to be studied under a microscope.

How is cystic fibrosis-related liver disease monitored over time?

If your child is diagnosed with cystic fibrosis-related liver disease they will have regular liver check-ups. This will help the medical team monitor the liver over time and start treatment if needed.

Tests used to monitor the liver may include:

Physical examination and liver blood tests

The medical team will continue to use physical examination and liver blood tests. These tests help monitor the liver over time. They will be used to look for any changes that show the liver disease is getting worse.

Abdominal ultrasound scan

The medical team will continue to use abdominal ultrasound scans. The scans will be done at regular intervals. They will be done sooner if the physical examination or blood test results pick up any changes to the liver.

At this stage ultrasound scans are useful for checking for:

- scarring of the liver (fibrosis)
- development of a complication called portal hypertension



Liver stiffness measurements

The medical team use special scans to monitor the amount of stiffness in the liver. Healthy liver tissue is soft, so stiffness shows that damage has occurred. The scans are usually only available at the specialist paediatric liver centres.

Endoscopy (also known as oesophago-gastro-duodenoscopy / OGD)

An endoscopy is a test that lets the doctor look inside the body without doing surgery. The medical team may use it if there is a chance your child has a complication called portal hypertension.

An endoscopy is done under general anaesthetic. During the procedure, the doctor passes a flexible tube down the food pipe (oesophagus). It then moves through the stomach and into the first part of the small intestine. The tube has a small light and camera at the end. The doctor uses it to look for swollen blood vessels in the food pipe (oesophageal varices).

The swollen blood vessels are caused by portal hypertension. They can be treated during the endoscopy procedure.

Magnetic resonance imaging (MRI) / magnetic resonance cholangiopancreatography (MRCP)

These scans use strong magnets and radio waves to make pictures of the internal organs. They are particularly useful for looking at the biliary tree.

What are the symptoms of cystic fibrosis-related liver disease?

You may not notice the early signs of cystic fibrosis-related liver disease. This is because your child may show no symptoms.

In most children, any symptoms are mild, and no treatment is needed. If symptoms do occur, the severity can vary a lot from person to person.

Symptoms may include:

Enlarged liver (hepatomegaly)

This is a larger than normal swelling of the liver. It may cause tummy (abdominal) discomfort or lead to your child “feeling full”.



An enlarged liver is the most common symptom of cystic fibrosis-related liver disease. It is usually found at the routine physical examination.

Head of Children's Services, Liver UK

Enlarged spleen (splenomegaly)

This is a larger than normal swelling of the spleen. It may happen as a result of a complication called portal hypertension. It can cause tummy (abdominal) discomfort or lead to your child “feeling full”.

Overactive spleen (hypersplenism)

A possible side effect of an enlarged spleen is an overactive spleen (hypersplenism). As the spleen gets bigger, it traps and destroys platelets from the blood. This makes them unavailable for their role in blood clotting. This may lead to increased bruising and nosebleeds.

Malnutrition and faltering growth

Malnutrition is a common symptom of cystic fibrosis. Children and young people with the condition often have trouble gaining weight. This can have an impact on their growth and development. They have a high need for extra calories and nutrition. Your child may already take supplements to help with this.

Malnutrition and vitamin deficiencies can become worse if your child develops cystic fibrosis-related liver disease. A high calorie diet may not be enough, and your child may need more support. This may involve special formulas, diets, supplements and tube feeding.



Nutritional support and advice will be given by members of the multidisciplinary team. This may include dietitians, play therapists and community nurses.

Head of Children's Services, Liver UK

Jaundice

This happens when bilirubin trapped in the liver passes back into the bloodstream. Bilirubin is a natural waste product made when old red blood cells break down. If there is too much, it causes yellowing of the whites of the eyes (sclera) and skin. Jaundice is a late symptom of cystic fibrosis-related liver disease.

Gallstones (cholelithiasis)

This is the name given to small stones that form in the gallbladder or bile ducts. If a stone blocks a bile duct this can cause severe pain and/or jaundice. Gallstones also increase the risk of infection in the bile ducts (cholangitis). In most cases gallstones are found on routine ultrasound scans but cause no problems.

What are the possible complications of cystic fibrosis-related liver disease?

In a small number of children, damage to the liver becomes worse over time. This causes the liver to become stiff and scarred (fibrosis). It can lead to severe scarring, known as cirrhosis.

The effects of severe scarring (cirrhosis) of the liver are:

Portal hypertension

When damage happens inside the liver, it stiffens. This makes it more difficult for blood to flow through it. This causes high pressure in the portal vein. This is the blood vessel that carries blood from many organs in the tummy (abdomen) to the liver. This pressure is known as portal hypertension.

Portal hypertension can make the spleen get bigger. It can also cause swollen blood vessels in the food pipe (oesophageal varices). These blood vessels have thin walls and may bleed. This may cause your child to vomit blood or pass black tarry poo (stools). Both symptoms need urgent medical attention.

The management of portal hypertension may be complicated by a condition called pancreatic insufficiency. This is common in children and young people with cystic fibrosis.

Fluid in the tummy (ascites)

Ascites is the term for a build-up of fluid in the tummy (abdomen). It is caused by:

- increased pressure in some of the blood vessels.
- the reduced ability of the liver to make an important protein called albumin.

The main symptom of ascites is swelling of the tummy (abdomen).



**Find more information
in Liver UK's booklet: Portal
Hypertension and Ascites**

Fluid in other parts of the body (oedema)

Oedema is the term for a build-up of fluid in the body. This fluid makes the tissues swell. Oedema is most likely to occur around the legs, ankles and feet.

Liver failure

Liver failure happens when large parts of the liver become damaged and scarred. The liver can no longer do its job and is said to be failing. This is a late stage of liver disease. It happens rarely in children and young people with cystic fibrosis-related liver disease.

How is cystic fibrosis-related liver disease treated?

There is no cure for cystic fibrosis-related liver disease. However, dietary support, medicines and other treatments can help slow down the disease. They may also reduce the symptoms and complications.

The medical team will recommend treatments. These will differ from child to child depending on the severity of the symptoms.

Dietary support

Children and young people with cystic fibrosis have a high need for extra calories and nutrition. Your child may already take supplements and follow a high calorie diet. Malnutrition and vitamin deficiencies can become worse if your child develops cystic fibrosis-related liver disease. A high calorie diet may not be enough.

Extra support will be given by dietitians and the medical team and may include:

- **MCT (medium chain triglyceride) formulas and diets**

MCT is a type of fat. It is more easily absorbed and is a good source of energy. Your child's dietitian will prescribe special milk formulas and supplements containing MCT. They will also provide a list of suitable foods for older children.

- **Fat soluble vitamins (vitamins A, D, E and K)**

Your child may already take fat soluble vitamins. If they develop liver disease, the medical team may need to change the formulation or dose. Vitamin K is particularly important in cystic fibrosis-related liver disease. It is needed for blood clotting and good bone health.

- **Nasogastric feeding**

Nasogastric feeding may be used if your child needs more calories or is not growing well. This involves passing a very thin, soft tube up the nose, down the back of the throat and into the stomach. A special milk formula will be given overnight using a pump. Parents and carers can be taught how to do this at home.



I was petrified of needing an NG tube for feeding but was really well supported when that day came. It gave her the energy to enjoy things again.

Parent



**Find more information
in Liver UK's booklet:
Nutrition**

Medicines

Medicines may be used to try to slow down the disease, or to help with symptoms. The choice of medication will depend on the severity of symptoms.

- **Ursodeoxycholic acid**

This is a bile salt that occurs naturally in small amounts in bile. Taking it as a medicine can help make the bile less sticky and allow it to flow more easily from the liver. Some research has shown that the drug improves symptoms and blood test results. However, the long-term benefits are still not clear. So far, research has not shown that it prevents advanced liver disease or the need for liver transplant.

- **Blood pressure medicines**

Drugs may be used to help lower high blood pressure in the portal vein. This will reduce the risk of bleeding from swollen blood vessels in the stomach or food pipe (oesophagus).

- **CFTR modulators**

CFTR modulators are new drugs that are being used in people with cystic fibrosis. They are different to the treatments used in the past because they target the genetic causes of the condition. They may help to ease cystic fibrosis symptoms and slow down its progression.

It is not yet clear if CFTR modulators can help with cystic fibrosis-related liver disease. We do not know if they stop liver disease from happening or slow down its progression. We do know that CFTR modulators cannot reverse scarring caused by advanced liver disease. More research is needed to find out the long-term effects of the drugs.



Invasive procedures

The medical team may use invasive procedures to treat the complications of cystic fibrosis-related liver disease.

Endotherapy

This is used in the treatment of swollen blood vessels in the food pipe (oesophageal varices). There are three different types of endotherapy. The choice of treatment will depend on the results of the endoscopy.

- **Banding (variceal band ligation)**

Banding is done during an endoscopy. The doctor places small rubber bands around the swollen blood vessels to make them close. This stops blood flow and reduces the risk of bleeding. Once a tiny scar has formed the rubber bands fall off. They may appear in your child's poo (stool) after a few days.



12 years on his liver is still doing what livers do and the portal hypertension has been controlled by the banding programme.

Parent

- **Glue injection (glue obturation)**

This treatment is done during an endoscopy. A medical glue is injected into the swollen blood vessels to seal them off and prevent bleeding.

- **Sclerotherapy**

This treatment is done during an endoscopy. It will be used if your child is too small for a banding procedure or if the swollen blood vessels are in the stomach wall (gastric varices). A medicine (sclerosant) is injected into the swollen blood vessels. This irritates the inside of the blood vessel wall. A scar forms, which blocks the blood flow and reduces the risk of bleeding.

TIPS (Transjugular intrahepatic portosystemic shunt)

This treatment may be considered for older children. It may be used:

- to help manage severe portal hypertension that cannot be controlled with endotherapy
- in young people waiting for a liver transplant
- when a liver transplant is not possible

During the procedure, a small metal tube is placed between the vein supplying blood to the liver (portal vein) and the veins draining blood from the liver back to the heart (hepatic veins).

This new channel or 'bypass' reduces pressure in the portal vein and allows blood to flow more freely. It makes swollen blood vessels (oesophageal varices) less likely to bleed. It also reduces the build-up of fluid in the tummy (ascites).

TIPS can be done as a planned treatment or as an emergency treatment if your child has a bleed.



Liver transplant

A liver transplant is used for the treatment of complex portal hypertension or liver failure. Only a small number of children with cystic fibrosis develop advanced liver disease that will lead to a liver transplant.

A liver transplant is an operation to remove a liver that does not work. It is replaced with a healthy liver from another person (donor). A child may receive a whole, split or reduced liver from a deceased donor. It is also possible that they may receive part of a liver from a living donor. A successful liver transplant will help a child survive when the liver is failing. It will also significantly improve a child's quality of life.

A liver transplant is a major operation. It will only be done after the benefits and risks have been carefully weighed up. The medical team will also assess and monitor your child's lungs. This is done to make sure that they are healthy enough to cope with major surgery and the recovery process. Following a successful liver transplant children need lifelong medication and follow up.

Cystic fibrosis-related liver disease will not return after a liver transplant. However, a liver transplant will not cure cystic fibrosis or the effects of the condition in other organs.

**Find more information
in Liver UK's leaflet:
Liver Transplantation**



Useful resources

Useful websites



Cystic Fibrosis Trust

www.cysticfibrosis.org.uk

A UK-wide charity providing support for people with cystic fibrosis.

Medicines for children: advice and guides

www.medicinesforchildren.org.uk/advice-guides/

A website with general advice on medicines in children.

How Liver UK can help



A diagnosis of liver disease can be worrying, and you may have a lot of questions. The Liver UK Children's and Families Team are here for you and for your family and friends.

Whether you have questions or just need someone to listen, we can help.

Find out how we can help you:
liveruk.org/support

Call our helpline to talk to one of our team: **0800 652 7330**



How we produce information

Feedback and information services

Your feedback is important to us and will help us improve our information. To provide feedback, email: **patient-info@liveruk.org**

For more information on how our information was developed, visit our website: **www.liveruk.org**



Patient Information Forum

We have gained PIF TICK accreditation for our information production process.



Thanks



All our information resources are reviewed by medical experts and families.

We would like to thank all our clinical experts and parent and family reviewers.

Disclaimer

This resource provides general information but does not replace personal medical advice. It is important to contact your medical team if you have any worries or concerns.



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www.liveruk.org/donate

Liver UK is the UK's leading charity dedicated to supporting adults, children and young people affected by liver disease or liver cancer. Our mission is to transform liver health by raising awareness, promoting prevention, and improving care and support for everyone affected.



liveruk.org
info@liveruk.org
0300 1311 292

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