Children's Liver Disease Foundation
fighting childhood liver disease

Cystic Fibrosis and Liver Disease

A Guide

An overview of cystic fibrosis-related liver disease, including the causes, diagnosis, symptoms, complications and treatment

This information has been written for:

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

Others who may also find this information useful:

- young people with cystic fibrosis
- healthcare professionals who would like to find out more about cystic fibrosis-related liver disease

It provides information on:

- how cystic fibrosis can affect the liver
- which children/young people get the condition
- diagnosis
- symptoms
- possible complications
- treatment

You may also find it helpful to read the following CLDF leaflets:

- Introduction to Liver Disease
- Nutrition
- Portal Hypertension and Ascites
- Liver Transplantation

Use this QR code to view our resources

Key facts about cystic fibrosis-related liver disease

1. Cystic fibrosis is a common genetic condition that causes thick, sticky mucus to build up in the body's tubes and passageways.
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.
3. Most children and young people with cystic fibrosis will not develop any serious problems with their liver. Around 5-10% will develop liver problems that have an impact on their health.
4. We cannot predict who will get cystic fibrosis-related liver disease.
5. Tests for cystic fibrosis-related liver disease include physical examination, blood tests, scans and occasionally liver biopsy.
6. The symptoms and severity of cystic fibrosis-related liver disease can vary from child to child.
7. In a small number of cases, damage to the liver can become worse over time.
8. There is no cure for cystic fibrosis-related liver disease but dietary support, medicines and medical procedures can help reduce the symptoms and complications.
9. Only a small number of children with cystic fibrosis develop advanced liver disease that will lead to a liver transplant.
10. Support services for children, young people and their families are available from Children's Liver Disease Foundation.

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 (mutations) 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

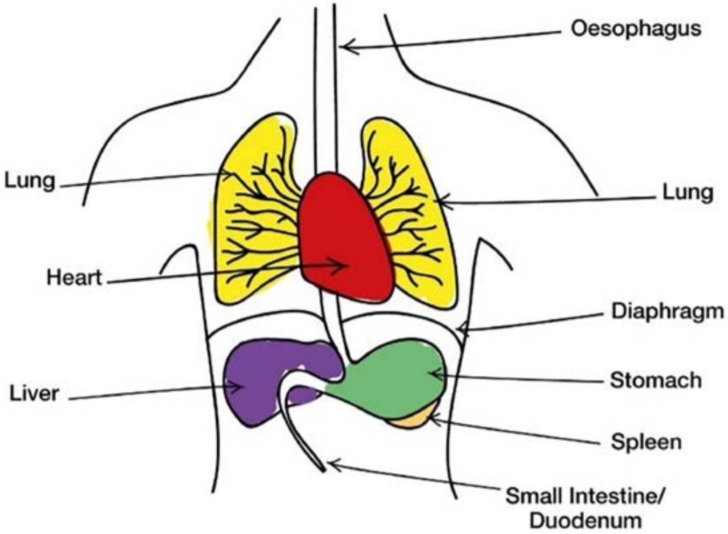

Figure 1: Location of the liver

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).

Find more information on the role of the liver in CLDF's leaflet: An Introduction to Liver Disease.

Which children or young people 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 do not 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.**
>
> **- CLDF Head of Services**

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
(also known as liver function tests/LFTs)

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.

Find more information in CLDF's leaflet: An Introduction to Liver Disease.

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 (see page 16).

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.**
>
> **- CLDF Head of Services**

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.**
>
> **- CLDF Head of Services**

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 and young people, 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 CLDF's leaflet: 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 may 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/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 CLDF's leaflet: 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 CLDF’s leaflet: Liver Transplantation.

Useful resources

Glossary of liver terms

Many medical terms may be used by those caring for children and young people with liver disease. Follow this QR code for a list of terms.

Other CLDF resources

CLDF produces a wide range of information for children and young people up to the age of 25 with liver disease, their families and the healthcare professionals who look after them.

Information can be downloaded or ordered from:
Website: **www.childliverdisease.org**
Email: **info@childliverdisease.org**
Tel: **0121 212 3839**
Follow this QR code to see our resources.

Information from other organisations

Cystic Fibrosis Trust

www.cysticfibrosis.org.uk

A UK-wide charity providing support for people with cystic fibrosis. They fund research, improve care, provide a network of support, and produce a range of information resources.

How we produce information

Our information resources are written, edited and reviewed with the help of medical experts and families living with liver disease.

Thanks

We would like to thank staff at each of the specialist paediatric liver centres: Birmingham Children's Hospital, King's College Hospital and Leeds Children's Hospital. We would also like to thank the families who helped us produce this information.

PIF TICK accreditation

We have gained PIF TICK accreditation for our information production process. Follow this QR code for more details.

Disclaimer

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

Feedback and information sources

Your feedback is important to us and will help us improve our information. To provide feedback, or for more information on how it was developed, email info@childliverdisease.org.

This leaflet was reviewed in July 2024

It is due to be reviewed by July 2027

Notes

Children's Liver Disease Foundation

Children's Liver Disease Foundation (CLDF) is the UK's leading organisation dedicated to fighting all childhood liver diseases.

CLDF provides information and support services to young people up to the age of 25 with liver conditions and their families and is a voice for everyone affected.

CLDF is here for you, whether you want to talk, and share with others, or just belong to a group which cares, knows what it's like and is fighting to make a difference.

Are you a young person up to the age of 25?

Contact the Young People's Team:

Phone: 0121 212 6024 **Email:** youngpeople@childliverdisease.org

Are you the parent or carer of a child or young person with a liver condition?

Contact the Families Team:

Phone: 0121 212 6023 **Email:** families@childliverdisease.org

Would you like to help us support the fight against childhood liver disease? Please help us to continue to support children, young people and families now and in the future.

Contact the Fundraising Team:

Email: fundraising@childliverdisease.org

Tel: 0121 212 3839
Email: info@childliverdisease.org
Website: www.childliverdisease.org

Registered Charity Number: 298858
Charity registered in Scotland: SC042140

Trusted Information Creator
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Children's Liver Disease Foundation
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