

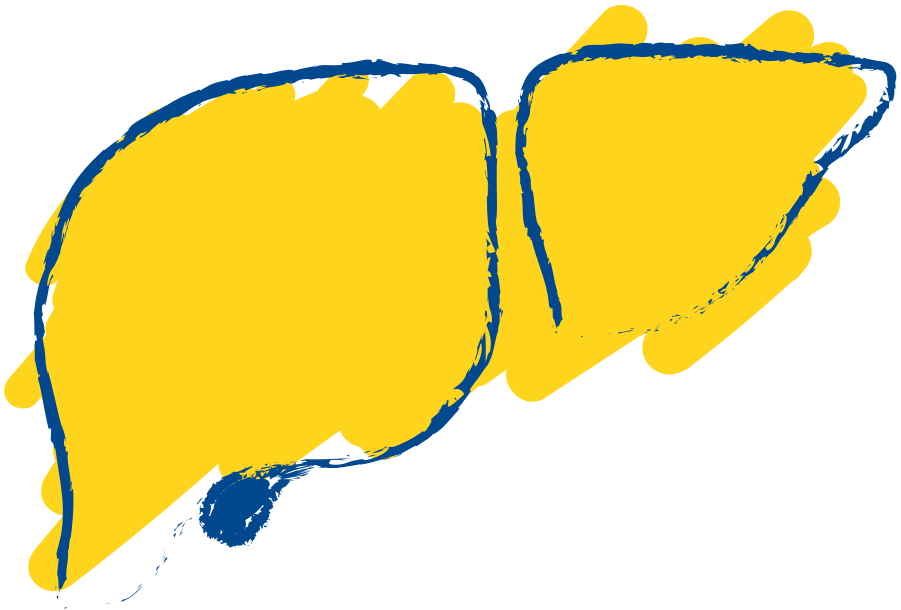


Children's Liver
Disease Foundation

fighting childhood
liver disease

Introduction to Liver Disease

A Guide



A general guide to liver disease in
children and young people

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There are over 100 different liver conditions which can affect children. This leaflet is designed to help you understand the basics of liver disease in children.

It includes information on:

- The liver—what it is, what it does and what can go wrong.
- Routine tests for liver disease—tests which are used to diagnose liver disease and to monitor the health of the liver.
- All of the key medical terms which may be used to talk about liver disease in children.

If you have any questions about anything mentioned in this leaflet you can talk to the medical team looking after your child or you can get in touch with CLDF's Families Team for further advice and support. Contact details are on the back page.

What is the liver?

Diagram 1: Location of the liver

The liver is a large organ found at the top of the abdomen (tummy area) on the right side. It is made up of cells; blood vessels and bile ducts. The main cells in the liver are known as hepatocytes.

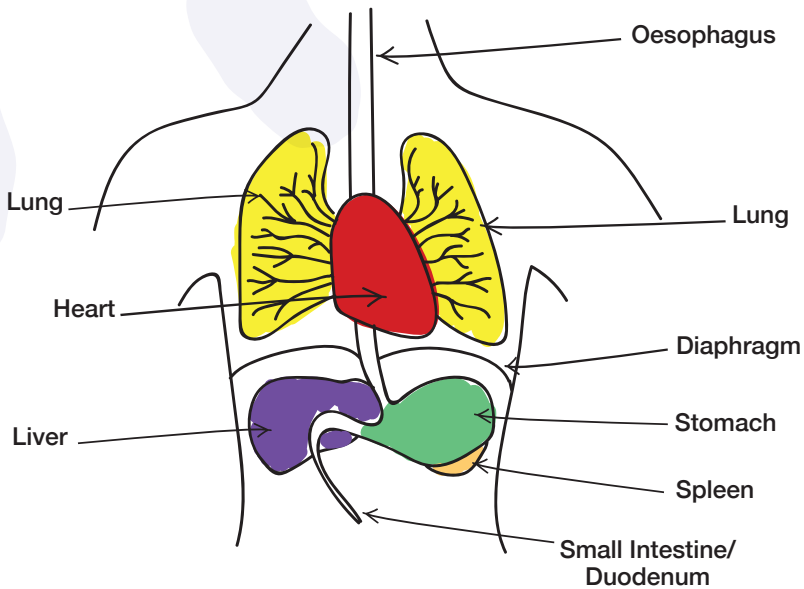
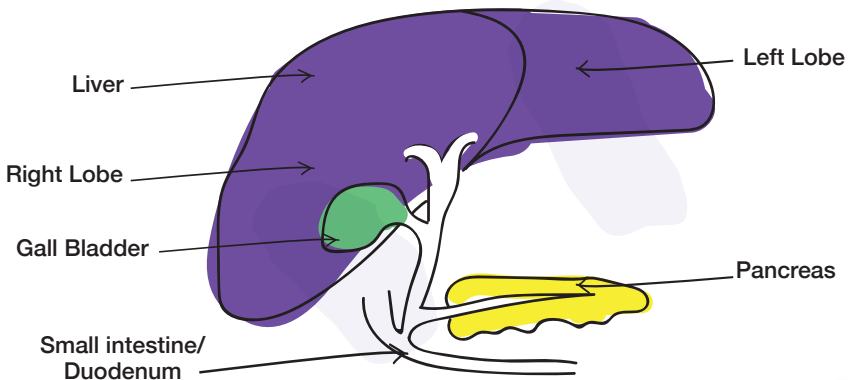


Diagram 2: The Liver

The liver is made up of two main parts: the right lobe and the left lobe. The right lobe is large, whilst the left lobe is smaller.



What does the liver do?

The liver is a complex organ. It has lots of different functions which are essential to live healthily.

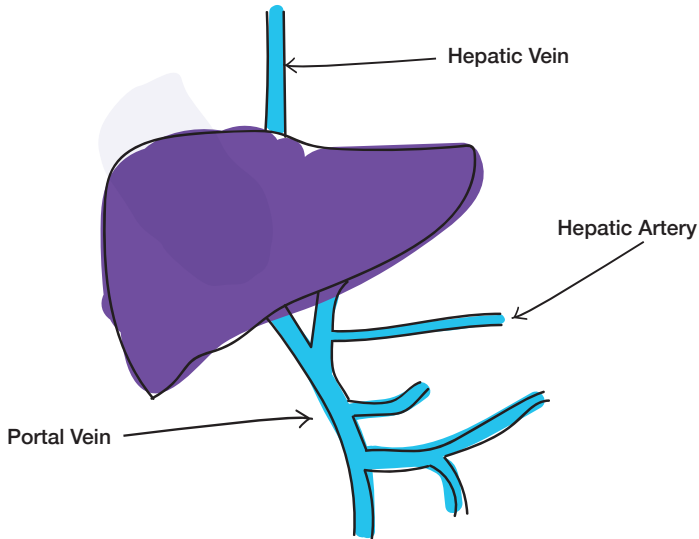
The liver filters blood

It receives blood from two sources:

- From the heart (via the hepatic artery)
- From the intestine (via the portal vein)

Diagram 3: The liver has an important role in the digestive system

Blood from the liver goes back to the heart through the hepatic vein.



The liver processes and stores nutrients from the blood to allow the body to use them. The blood entering the liver from the intestine contains fats, carbohydrates, vitamins and other nutrients.

Carbohydrates are broken down into sugars which provide energy. Excess sugars are stored in the liver as glycogen, for release when needed.

The liver makes lots of different substances

The liver produces lots of different substances that the body needs including:

- Blood plasma proteins such as albumin. Albumin controls how much fluid is in each part of the body
- Clotting factors which help to stop bleeding
- Cholesterol
- Vitamin D
- Immune factors which help to fight infection
- Some hormones
- Bile

What is Bile?

The liver produces bile which is a green/yellow liquid. Bile is formed from bilirubin, which comes from old red blood cells which have been broken down in the spleen. Bile helps the body to digest food by breaking fats down so they can be absorbed and enables the absorption of fat-soluble vitamins. Bile also helps the body get rid of waste products such as bilirubin and excess cholesterol which it passes out into the stool (poo).

After bile has been produced by the liver it is transported to the gall bladder where it is stored. When food is eaten the gall bladder releases bile through bile ducts into the small intestine to help with digestion and remove waste products.

The liver processes waste products

The liver gets rid of waste such as the breakdown products of old red blood cells, ammonia (which comes from proteins), medicines and drugs.

What problems can affect the liver?

- The bile flow out of the liver may be blocked
- An infection/virus–this can cause inflammation/swelling
- Metabolic diseases–problems with the way the cells make energy
- Drugs and poisons
- Poor blood supply
- Sometimes the cause is unknown–these are known as idiopathic or cryptogenic

What signs are there that a child may have liver disease?

There are many different signs and symptoms and they depend on the underlying cause. It's rare for them to all happen at once. Liver damage can happen slowly over time, but sometimes it can happen very suddenly.

Signs and symptoms include:

- Jaundice (yellowing of the skin and the whites of the eyes)
- Nausea, vomiting and loss of appetite
- Yellow urine in newborns, dark urine in older children
- Pale coloured stools
- Change in sleep patterns in older children
- Vomiting blood or passing blood in the stool
- Tiredness
- Abdominal swelling
- Itching (known as pruritus)

- Poor weight gain or weight loss
- Abdominal pain
- Nosebleeds and bruising easily

What tests are used to diagnose and monitor liver disease?

Blood tests

Liver function tests (LFTs) are carried out on blood samples in a laboratory. They are a common way of seeing how well the liver is working. They test the levels of particular proteins and enzymes in the liver as well as the level of bilirubin.

Liver function tests are also used to monitor a child's liver disease over time to see if the liver is getting healthier, getting worse or staying the same.

Other blood tests will also need to be done to diagnose the cause of the liver disease.

To see a chart explaining the different blood tests which are carried out, including the LFTs, with their expected results, turn to page 11.

Urine and stool

The colour of urine (wee) and stool (poo) can provide information about the liver. As mentioned previously pale stool can be a sign of a problem with the liver, along with dark urine. Sending urine samples to the laboratory can give more information for the diagnosis.

Abdominal ultrasound

This is a scan of the abdomen. It's the same as the scan that pregnant women have to see their baby and doesn't hurt at all.

The ultrasound can show the size and texture of the liver and other organs such as the gall bladder, bile ducts, spleen and kidneys. It can also show the blood flow into and out of the liver.

Liver biopsy

A liver biopsy may be recommended if tests show that there could be a problem with the liver. The results of the biopsy may indicate if there is a problem with the liver or what it is. A biopsy may not be able to provide a diagnosis but may provide a clue to what other tests may be needed. They can also be used to monitor the condition.

A liver biopsy involves inserting a very thin needle through the tummy and into the liver. The needle takes a very small sample of the liver. That sample of liver tissue is then sent to the laboratory to be examined under a microscope.

Children may either have the procedure done with a local anaesthetic or with general anaesthetic (so they are asleep). When young people move to adult services they will have the biopsy done whilst they are awake using a local anaesthetic.

Liver function tests (LFTs) explained...

It's important to note that every child will have their own 'normal' and whilst these levels are a guide, it doesn't necessarily mean a result outside of these ranges is dangerous. Always talk to your child's medical team if you have any worries about test results.

LFTs show the amount of inflammation in the liver, whether the bile ducts are affected and how well the liver is functioning.

How well the liver is functioning is assessed by measuring what the liver is making, for example, albumin (a type of protein) and clotting factors all of which are made by the liver.

Name of Test	Normal Range	What does this show?
<p>Bilirubin (SBR)</p> <p>a) Total bilirubin</p> <p>b) Conjugated bilirubin</p>	<p>a) 3 – 20 mmol/l</p> <p>b) conjugated less than 7 mmol/l</p>	<p>The total bilirubin level corresponds to the level of jaundice present.</p> <p>The amount of conjugated bilirubin compared to the unconjugated bilirubin can show whether jaundice is caused by red blood cells breaking up too quickly (not a liver disease) or is due to a reduced flow of bile out of the liver.</p> <p>Conjugated means that a sugar has been added to the bilirubin.</p>
<p>Aspartate Aminotransferase (AST)</p> <p>Alanine Aminotransferase (ALT)</p>	10 – 40 IU/l	High levels can mean there is liver inflammation present.
Gamma-Glutamyl Transferase (GGT, gGT or Gamma GT)	5-55 IU/L	<p>High levels can mean bile duct inflammation or obstruction.</p> <p>The result can sometimes give information about bile production.</p>
Alkaline Phosphatase (ALP)	Less than 350 IU/L	<p>High levels can mean there is bile duct inflammation</p> <p>BUT</p> <p>ALP is not specific to the liver as it is also produced in other organs e.g. bones. Therefore, abnormal levels can be due to processes occurring in other parts of the body.</p>
Total Protein	60 – 80 g/l	High and low levels can be used in consideration with the other LFT results.
Albumin	35 – 50 g/l	Low levels can mean the liver is not making enough, that protein is being lost through the stool or a child is not well nourished.

Another blood test which is helpful to assess how the liver is working is a blood clotting test. Blood is taken, chemicals are added to it in the lab and the time taken for blood to clot is measured.

Name of Test	Normal Range	What does this show?
<p>Blood Clotting (Coagulation) Test</p> <p>PT (Prothrombin time)</p> <p>INR (International normalised ratio*)</p> <p>*INR is a standardised version of the PT for comparison, they both measure the same thing</p>	<p>12 – 15 seconds</p> <p>0.9 – 1.2</p>	<p>The liver produces substances needed for blood to clot.</p> <p>Raised levels can mean there is less Vitamin K due to a reduced bile flow.</p> <p>If Vitamin K is given and the result is still raised, it may mean there is damage to the liver.</p>

Other blood tests include:

Name of Test	What is measured?
Full blood count (FBC)	<p>Haemoglobin (Hb)</p> <p>White blood cell count (WBC)</p> <p>Platelets (Plt) – levels can fall in liver disease if the spleen is enlarged</p>
Urea and electrolytes	<p>Urea and creatinine are waste products removed by the kidneys.</p> <p>Electrolytes include sodium, chloride and calcium.</p>
Blood glucose	A high or low level of sugar in the blood can sometimes be a sign of liver damage or a metabolic liver disease.
Ammonia	<p>Ammonia is a waste product. It is converted into urea in the liver and is then removed in urine.</p> <p>A higher level than usual may mean there is damage to the liver or an underlying metabolic condition.</p>

Some of these tests may be used to monitor the effects of medications on the body.

What treatments are available?

There are treatments which can control and support the liver. Treatment will depend on the specific liver disease and the age and condition of the child but they can include:

- Medicines
- Special diets
- Operations
- Exercise or lifestyle change
- Liver transplantation

CLDF is committed to funding research into treatments for childhood liver disease. You can read about the current research programme at childliverdisease.org/research.

Glossary of Liver Terms

There are many medical terms which are used by those caring for children and young people with liver disease. It can be really helpful to know these terms which may be used during appointments.

If you come across a medical term you don't know you can look up an explanation here.

A

Acute – sudden and severe.

Alanine aminotransferase (ALT) and aspartate aminotransferase (AST) – enzymes produced mainly in the liver.

Albumin – a protein made by the liver. Controls fluid in the blood and tissues.

Alkaline phosphatase (ALP/ALK-PHOS) – an enzyme produced in the bile ducts and elsewhere in the body.

Anaemia – a lack of iron can lead to a person having fewer red blood cells than usual. There can also be other causes.

Anastomosis – a connection made surgically between two tubes.

Angiogram – a scan using x-rays to see blood vessels. It can be used to see the blood vessels which supply the liver.

Antibody – a protein which fights infection.

Ascites – fluid collecting in the abdomen.

Atresia – blocked, destroyed or missing.

Autoimmune (autoantibodies) – an immune response against the body's own tissue.

B

Banding – a treatment for varices. Surgical elastic bands are placed around the varices during an endoscopy.

Bile – a green/yellow liquid containing bile salts, bilirubin and cholesterol.

Bile acids – acids found in bile which can be converted into bile salts.

Biliary tract – the system of tubes which carry bile from the liver and gall bladder and drain into the intestine.

Bilirubin – this is a product of the breakdown of old red blood cells. It has a yellow colour which gives stool and blood their colour.

Bilirubin travels in the blood stream to the liver where it has a sugar added and becomes conjugated bilirubin. Before this sugar is added it is known as unconjugated bilirubin.

Biopsy (liver) – a procedure to take a small sample of the liver to test in the laboratory.

Body Mass Index (BMI) – a test using height and weight to measure whether an individual is a healthy weight.

C

Cannula - a short, soft, narrow plastic tube temporarily put into a vein so medicines/fluids/blood etc. can be given intravenously (IV) as needed.

Cardiac – refers to the heart.

Cardiovascular system – the heart and blood vessels which transport nutrients and oxygen around the body.

Carrier – someone who has a gene which can cause a genetic condition but doesn't have the condition themselves.

Cell – basic structural and functional component of all living things.

Central venous line (CVL) – a central line is similar to a cannula but is a longer tube used to give medicine, fluid, nutrition or blood directly into a larger vein and can also be used to take blood samples. It is a long, narrow plastic tube which can be placed into a vein in the neck, chest or groin and is used for longer than a cannula. It is usually inserted under a general anaesthetic.

Cholangitis – inflammation or infection of bile ducts which can cause poor bile flow from the liver and liver damage.

Cholestasis – reduction or blockage of bile flow.

Chronic Hepatitis – inflammation of the liver that lasts over a period of time. Liver cells may be destroyed by the inflammation.

Chronic – an illness or condition which lasts over a period of time.

Cirrhosis – scarring of the liver when liver cells are damaged and destroyed. Scarring can stop blood flowing through the liver properly leading to further damage. The liver becomes smaller, hardens and can't function properly. Cirrhosis cannot be reversed.

Coagulopathy – refers to any condition where the blood cannot clot properly.

Computerised tomography scan (CT Scan) – x-rays are used to create a detailed picture of different parts of the body.

Congenital – a condition which is present from birth.

Cystic – relating to the gall bladder.

Cytomegalovirus (CMV) – a virus belonging to the herpes virus group.

D

Distension – expansion or enlargement.

Duct – a tube which fluid can pass through from one part of the body to another, e.g. bile ducts.

Dysplasia – the abnormal development of body tissue or an organ.

E

Echocardiogram (Echo) – a scan of the heart using high frequency sound waves, similar to an ultrasound of the abdomen.

Electrocardiogram (ECG) - records the electrical activity of the heart.

Encephalopathy – changes in the brain which can be due to liver failure. The build-up of toxins in the blood can lead to tiredness, irritability and personality changes.

Endoscopic retrograde cholangiopancreatography (ERCP) – a special x-ray used to examine bile ducts under general anaesthetic.

Endoscopy – using a flexible, thin tube with a camera attached to look at the inside of the upper end of the digestive system. It is passed through the mouth and down the oesophagus (food tube).

Enteral feeding – feeding through a tube directly into the stomach or part of the intestine.

Enzyme – substances in the body which help chemical reactions to take place.

Epstein Barr virus (EBV) – a virus which causes glandular fever.

F

Faeces – also known as stool or poo.

Fibrosis – scar tissue which replaces normal tissue which, when extensive, is known as cirrhosis.

Full blood count (FBC) – a blood test measuring all of the different types of cells in the blood.

Fulminant – a severe, sudden form of liver failure. More commonly known as acute liver failure.

G

Gallbladder – a small sac which stores and releases bile made by the liver.

Gallstones – the stones which can be formed by bile and collect in the gall bladder and bile ducts. They can cause pain and may pass into the common bile duct and cause cholangitis or obstructive jaundice.

Gamma-Glutamyl Transferase (also known as Gamma Glutamyltranspeptidase (Gamma GT (GGT)) – an enzyme produced mostly by the bile ducts.

Gastroenterology – study of the digestive system.

General anaesthetic – an individual is put into a state of controlled unconsciousness.

Genes – genes are made up of DNA and provide instructions for the body.

Graft – an organ, tissue or cells used for transplantation.

H

Haematemesis – vomiting (sickness) blood.

Haemoglobin – part of a red blood cell which carries oxygen around the body.

Hepatic – referring to the liver.

Hepatic artery – the blood vessel which brings blood with oxygen to the liver.

Hepatic vein – the blood vessel which takes blood away from the liver.

Hepatitis – inflammation of the liver.

Hepatology – study of the liver.

Hepatomegaly – enlargement of the liver.

Hypo/hypertension – low/high blood pressure.

I

Idiopathic – something which doesn't have a known cause.

Immune system – the body's defence which fights against foreign substances in the body such as bacteria and viruses.

Immunity – protection against a particular disease. This can happen naturally or due to vaccination.

Immunoglobulin – a protein which plays a role in the immune system.

Immunosuppression – medications which can be taken which make the immune system less active.

Incidence – the number of new cases of a disease over a certain period of time in a set population.

International normalised ratio (INR) – a measure of how well blood clots.

Intravenous – into the vein. Some drugs are delivered straight into the veins.

J

Jaundice – a high level of bilirubin in the blood leading to the yellowing of the skin and/or the whites of the eyes.

K

Kasai portoenterostomy – an operation to get bile flowing from the liver to the intestine in biliary atresia. Bile ducts are removed and a piece of intestine is used to replace the ducts.

L

Liver function tests (LFT) – blood tests which can show how well the liver is working.

Liver transplant – an operation to replace a damaged liver with a donor liver.

- Auxiliary liver transplant – where part of a donor liver is placed alongside the child's own liver.
- Orthotopic liver transplant – where the child's liver is replaced by all of part of a donor liver.

Lymph – a clear, watery liquid derived from body tissues which carries white blood cells and fats. It travels through the lymphatic system of the body.

M

Magnetic resonance imaging (MRI) – a detailed scan of the body using magnets rather than x-ray.

Malabsorption – the body doesn't absorb nutrients properly when we eat them.

Metabolism – the processes in the body which break down and build up different chemicals.

Metabolic disorder – a condition in which metabolism is affected.

N

Nasogastric (NG) feeding – giving food through a tube which goes through the nose and down the throat to the stomach.

Nasogastric tube – the tube used during NG feeding above.

Nasojejunal (NJ) tube – a small tube that is passed up the nose and down the throat, through the stomach, and into the small intestine used for feeding.

O

Oedema – also known as fluid retention, fluid builds up in the tissues affected.

P

Parenteral nutrition (PN) – feeding nutrients directly into the vein when an individual cannot eat normally.

Percutaneous Transhepatic Cholangiography (PTC) – a special x-ray which looks at the bile ducts. It is done under a general anaesthetic using a needle which is inserted through the skin of the abdomen into the bile ducts. It can be used to diagnose and treat a variety of bile duct issues.

Phlebotomist – a person who takes blood samples.

Platelets – part of the blood and important for clotting the blood. They can be low in people with liver disease.

Polymerase Chain Reaction (PCR) – a test which checks if a virus is present in the blood. A positive result means the virus is present.

Portal Hypertension – high blood pressure in the portal vein which may occur due to scarring of the liver or a blockage of the portal vein.

Portal Vein – main vein carrying blood from the intestine to the liver.

Pruritus – itching of the skin.

Pulmonary – refers to the lungs.

R

Rejection – when the body's immune system attacks a transplanted organ. It can be acute (sudden) or chronic (over a period of time).

Renal – refers to the kidneys.

Respiratory – refers to breathing.

S

Sclerotherapy – a treatment for varices where a solution is injected into them during an endoscopy.

Screening – testing for a condition or the genes for a condition.

Spider Naevi – small, broken veins under the skin which can look like red spiders.

Spleen – an organ which removes old blood cells. Its blood supply is connected to the liver.

Splenomegaly – an enlarged spleen.

Split bilirubin test – a blood test which measures how much conjugated and unconjugated bilirubin is in the blood. See 'bilirubin' for more information.

Steroids – medication which reduces the activity of the body's immune system.

T

Thrombosis – a blood clot in a vein or artery.

TPR – an abbreviation for temperature, pulse and respiration.

Triglycerides – a type of fat.

TTA and TTO – abbreviation for ‘to take away’ or ‘to take out’ referring to medicines which are taken home.

U

Ultrasound scan (USS) – a scan which takes pictures of organs and blood vessels inside the body.

Urea and electrolytes – may be called ‘U & E’s or kidney function tests. They are monitored to see how well the kidneys are working.

V

Varices – veins in the lining of the intestine, oesophagus and stomach which can be enlarged and swollen due to portal hypertension and can bleed if not treated.

Viral load – a viral load test is a lab test that measures the number of virus particles in a millilitre of blood.

X

Xanthomas – cholesterol deposited under the skin which can look like pale warts.



CLDF produces a wide variety of information resources for children and young people up to the age of 25 with liver disease, their families and the healthcare professionals who look after them. This information can be downloaded or ordered from CLDF's website childliverdisease.org. For further enquiries regarding CLDF's information please contact the Information and Research Hub Manager by email at irhm@childliverdisease.org or call 0121 212 6029.

Thanks

This booklet has been written, edited and reviewed with the help of staff at each of the specialist paediatric liver centres: Birmingham Children's Hospital, King's College Hospital and Leeds Children's Hospital. Thank you to all of the staff involved who have made the production of this leaflet possible.

Disclaimer

This leaflet provides general information but does not replace medical advice. It is important to contact your/your child's medical team if you have any worries or concerns.

Feedback and Information Sources

Information within this leaflet has been produced with input from the three specialist paediatric liver centres in the UK. To provide feedback on this leaflet, or for more information on the content of this leaflet, including references and how it was developed, contact Children's Liver Disease Foundation: info@childliverdisease.org.

This leaflet has been reviewed in December 2020. It is due to be updated in December 2023.

What is Children's Liver Disease Foundation (CLDF)?

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, funds vital research into childhood liver disease and is a voice for everyone affected.

Are you a young person up to the age of 25 with a liver condition or a family member? CLDF's Families and Young People's teams are here for you, whether you want to talk about issues affecting you, meet and share with others, or just belong to a group which cares, knows what it's like and is fighting to make a difference. You are not alone.

If you are a parent/carer or family member then get in touch with CLDF's Families Team:

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

If you are a young person and want to find out more about CLDF's services you can contact CLDF's Young People's Team:

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

CLDF have a social media platform called HIVE for 13 – 24 year olds with a liver disease/transplant to make new friends, connect and share stories.

childliverdisease.org/young-people/hive

Would you like to help us support the fight against childhood liver disease?

CLDF's work relies on voluntary donations and fundraising. Please help us to continue to support children, young people and families now and in the future.

To find out more about fundraising and how you can join the fight against childhood liver disease you can visit **childliverdisease.org/get-involved**. Alternatively you can email the Fundraising Team at **fundraising@childliverdisease.org** or call them on **0121 212 6022**

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Children's Liver
Disease Foundation

fighting childhood
liver disease