

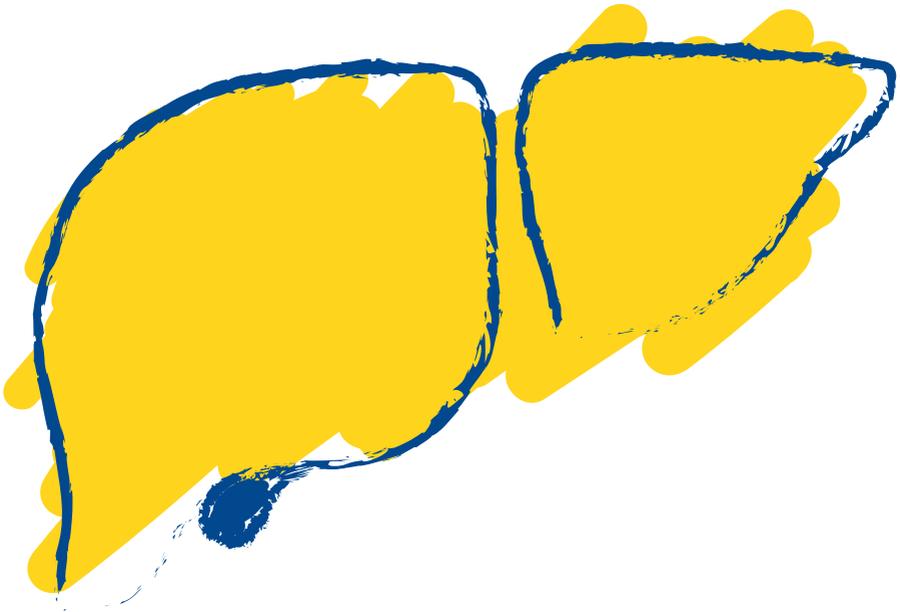


Children's Liver
Disease Foundation

fighting childhood
liver disease

Portal Hypertension and Ascites

A Guide



Information on portal hypertension and
ascites in children and young people

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This guide has been written for:

- Parents/carers of children with liver disease

It may also be helpful for:

- Young people with liver disease
- Healthcare professionals who would like to find out more

It provides information on:

- What portal hypertension and ascites are
- What causes them
- Signs and symptoms
- Complications
- Treatment options

You may also find it helpful to read the following:

- An Introduction to Liver Disease

What is portal hypertension?

As blood travels around the body it goes through the digestive system to pick up the nutrients from food which has been eaten. This blood then takes these nutrients to the liver through a blood vessel called the portal vein. This is called the portal circulation. The liver processes the blood before it goes back to the heart.

Hypertension means high blood pressure. When the pressure of the blood in the portal circulation is unusually high this is known as **portal hypertension**.

What causes it?

Portal hypertension is most often caused by difficulty in blood flow into the liver which then causes a build-up of pressure.

The main causes of this happening are:

- A blockage in the portal vein. This can happen if a blood clot (thrombus) stops blood from passing through the portal vein freely. This is called portal vein thrombosis. In this situation the liver may be completely normal and healthy.
- Scarring in the liver due to cirrhosis or fibrosis. Cirrhosis occurs as liver cells are damaged and die. They are then replaced with scar tissue (fibrosis).

Occasionally, a blockage of the hepatic vein draining blood from the liver can cause increased pressure in the portal vein, but this is not common in children.

What are the signs and symptoms?

Often portal hypertension will not cause any symptoms, but the onset of any symptoms relating to portal hypertension will be monitored. If portal hypertension causes troublesome symptoms, it may be referred to as severe or complicated portal hypertension.

The increased pressure in the affected blood vessels can affect other areas of the body. Potential problems include:

An enlarged spleen (splenomegaly)

The spleen is connected to the liver by a vein which joins the portal vein. This means that if there is high pressure in the portal vein, this may affect the spleen, causing it to enlarge.

The spleen's job is to break down old blood cells. When it gets larger it removes more blood cells than it should. This includes a type of blood cell called platelets which help to stop bleeding. Not having enough platelets can lead to bruising and nosebleeds.

Abdominal swelling

Ascites can be the reason for a child's tummy suddenly getting bigger. Ascites is a collection of fluid in the tummy (abdomen). Portal hypertension can cause this because the high pressure in the blood vessels pushes fluid out of these blood vessels into the space between the organs.

Abdominal swelling can also be caused by the spleen and liver getting larger. If this is the cause the tummy is not normally as swollen as it is with ascites.

Noticeable veins under the skin of the tummy

Veins can appear under the skin of the tummy because it is swollen. They can also appear because blood returning to the heart through the portal vein is blocked. The blood is then forced out of its usual route and new veins form to get around the blockage. When this happens near the surface of the skin the veins may be seen.

Varices

As well as veins becoming visible over the surface of the tummy, other veins can become enlarged, especially in the digestive tract (the oesophagus, stomach and intestine). These are called varices. They look similar to varicose veins which people can have in their legs. When these form in the food pipe (oesophagus), stomach or rectum they can bleed as they have thin walls.

Signs of Bleeding

Varices have thin walls and may bleed. Bleeding can cause tiredness and can cause a child to feel short of breath. It can also cause a pale appearance.

Blood might appear when a child is sick and may appear in their stool (poo) which can be bloodstained or black.

If there is a large bleed a child may have tummy pain, be dizzy and may collapse. **Whatever the sign of bleeding, seek emergency medical advice by calling 999.**

Diarrhoea and poor weight gain

The blood vessels which line the intestine can become swollen as blood flows through them under a high pressure. This can lead to fewer nutrients and less fluid being absorbed from food travelling through the intestine leading to diarrhoea and poor weight gain.

How is portal hypertension diagnosed?

There are tests which can be done to find out whether a child has portal hypertension. These include:

A general physical examination

An ultrasound scan

This is the same scan pregnant women have. It is used to measure the size of the liver and the spleen. If the spleen is larger than expected, this may suggest the child has portal hypertension. The scan can also be used to check the direction of the blood flow in the veins around the liver.

Endoscopy

A flexible tube, called an endoscope, is passed into the mouth, down the oesophagus, through the stomach and the intestine. This allows the doctor to look directly at the lining of the digestive tract to see if there are varices. This is usually done under general anaesthetic (where the child is completely asleep) or under sedation (where the child has drugs to make them sleepy and relaxed).

Colonoscopy

This is similar to the endoscopy but the tube is passed up the anus to examine the lower intestine. It is often not needed to make a diagnosis.

Blood test

Your child will have blood taken and the levels of platelets will be reviewed.

There are some cases where a child has portal hypertension without having a known liver disease. These children may need more tests, including:

Angiography

Under anaesthetic, a thin tube is passed into a blood vessel in the groin or neck and a special dye is injected. X-rays are then taken to give a 'map' of the portal vein and its branches and also measure the blood pressure in the liver vessels.

Magnetic resonance imaging (MRI)

A cannula or drip is inserted, which is used to pass dye into the vein. It then involves lying on a bed in a long circular tube whilst computerised pictures are taken and also measure the blood pressure in the liver vessels.

Sedation or anaesthetic is not normally required provided the child is able to stay still, but in young children an anaesthetic may be needed.

How is it treated?

If there are no symptoms or complications, then treatment may not be required, although the child's condition will be monitored. Treatment may be needed if the varices are significant in size according to international grading criteria and if there is an underlying liver condition which has caused the portal hypertension. If portal hypertension leads to varices which bleed, treatment of the varices will definitely be required.

The main treatment options for varices are called banding and sclerotherapy. These are explained further on in the leaflet.

Children with portal hypertension may also have varices in gut areas that are not accessible by endoscopy and endoscopic treatment may not be possible. However, other non-endoscopic treatment options are available.

In contrast to adults, there isn't currently strong evidence on medicines that can help in the management of portal hypertension in children.

Banding

When portal hypertension is confirmed, an endoscopy will be planned to assess whether varices are present. The timing of this will depend on the age and size of the child, as well as any complications that are known to be present, such as an enlarged spleen and low platelet count. During the endoscopy, if varices are present, the doctors will grade them according to their size. If necessary, the varices will be treated at the time of the endoscopy. Banding involves placing a medical rubber band over the affected blood vessels (the varices).

This causes the blood in the varices to clot or thrombose. This means that blood cannot flow through the varices which reduces the risk of bleeding. The elastic band will fall off and may be seen in a child's stool (poo) after a few days, although it is tiny and may not be noticed.

Banding can be done urgently when your child is an inpatient, or it can be a planned procedure. Repeated endoscopies are usually needed to improve the varices, usually with intervals of weeks or months between each session.

Sclerotherapy

Another treatment for varices is called sclerotherapy. This is performed when banding may not be possible, for example, in a very small child.

An endoscope is passed down the oesophagus, a needle is passed through the endoscope into or next to the varices and a chemical is injected. The chemical causes the varices to clot and close off.

Does portal hypertension always lead to bleeding?

No. A child can have portal hypertension for years without having any related problems.

The varices which form may develop in areas of the body where they will not bleed. These veins help the blood flow back into the main circulation via a new route so the blood pressure in the portal circulation is lowered.

Bleeding is more likely if varices form in places where the walls of veins are thin, including the oesophagus and the stomach.

If the varices have inflamed the lining of either the oesophagus or the stomach, then a medicine will be given to protect the lining and help it to heal.

What should I do if I think my child may have a bleed?

If a child is vomiting blood or passing blood in their stools, then ring 999 straight away.

The team at the hospital should be told that the child has a diagnosis of portal hypertension and be given a description of the symptoms. The medical team will give families a letter to carry with them to show the emergency doctor.

In hospital the child will be assessed and treated if needed. Some bleeds may be very small but in general it will be recommended that your child remains in hospital for observations and an endoscopy will be organised in the liver centre.

The hospital team caring for your child may:

- monitor the child's blood pressure and pulse
- observe the vomiting/colour of stools
- find out how much blood has been lost
- give the child fluid/blood through a drip
- give medicines to stop the bleeding
- contact you liver team and discuss management plan
- perform an endoscopy to look at the varices which are bleeding
- treat the varices by putting rubber bands around them (banding) or by injecting the varices with a chemical to stop the bleeding (sclerotherapy)

What will happen to a child in the long term?

Children can be managed for years on banding programmes. If the programme no longer manages the varices effectively, then further treatment options are available. These options can be surgical in the form of creating a bypass in the portal system (if there is a blockage), a shunt (if there is high pressure in the system) or replacing the liver and its vessels (liver transplantation).

Do children with portal hypertension need to take special precautions?

Medicines such as aspirin and ibuprofen should be avoided as they can make bleeding in the gut more likely. Paracetamol (Calpol) is a safer alternative.

It is important to be aware of the potential issues which could occur, e.g. bleeding. It is vital that all carers and teachers are aware of what to look for and what to do if they notice any signs that there may be a problem.

If the spleen is enlarged there is a higher chance of it being damaged if it receives a hard blow so children may be advised to avoid activities where they may receive a blow to the abdomen, e.g. contact sports such as kickboxing and rugby. Most physical

activity however is to be actively encouraged and children should participate in PE at school.

It's important not to limit activities unnecessarily and the child's medical team will be able to provide specific advice on a case by case basis.

If a child with portal hypertension is travelling/flying it's important that their medical team are informed well in advance as the child may need treatment before going away.

Other complications of portal hypertension

In rare cases of portal hypertension the veins in the lungs can also be affected.

Children may develop hepatopulmonary syndrome characterised by:

- low oxygen levels in the blood due blood vessels in the lungs dilating and mixing of oxygenated with non-oxygenated blood
- portopulmonary syndrome. This is where there is high blood pressure in the pulmonary veins due to resistance to blood flow through the circulatory system.

Heart and lung investigations will be required to establish the diagnosis and liver transplantation is usually the treatment of choice.

Ascites

What is ascites?

Ascites is a collection of fluid which gathers around abdominal organs and gives children affected a swollen tummy. It can be caused by portal hypertension. It can be associated with oedema which is swelling in tissues in other areas of the body. Oedema is more likely to occur around the eyes and face and in the foot, ankle and leg.

What causes it?

Ascites can be caused by:

Less albumin (a protein) being made by the liver

Albumin helps to keep fluid within blood vessels. If the liver produces less albumin, its levels fall and fluid leaks out of blood vessels and collects in cavities and tissues in the body. When the fluid collects in the abdomen it is known as ascites.

The fluid can also leak into the space between the lungs and the chest wall which is known as pleural effusion.

Increased pressure within blood vessels

Portal hypertension raises the blood pressure in the veins and around abdominal organs which leads to fluid leaking into the abdomen.

Salt being retained in the body

This can contribute to the build-up of fluid in the tissues and cavities.

What are the signs and symptoms of ascites?

- protruding tummy. Signs of this can be the need for bigger clothes or nappies. Sometimes the belly button can be pushed outwards (umbilical hernia).
- shiny, tight skin over the swollen tummy
- prominent veins visible beneath the skin on the tummy
- quick weight gain which is not due to normal growth
- shortness of breath
- loss of appetite (eating less)

How is a diagnosis made?

A diagnosis can be made by the doctor examining the child or by using an ultrasound scan.

What are the possible complications?

Poor nutritional progress

The fluid in the abdomen can make children feel full very easily. Children may eat smaller portions and can vomit due to limited space in the abdomen. A dietitian will be able to advise and support you. CLDF has a leaflet on nutrition with further information.

Infection

The fluid in the abdomen can become infected by bacteria. This is called spontaneous bacterial peritonitis.

Any fever with pain or tenderness over the abdomen with ascites should be reported to the medical team immediately. This is usually treated with intravenous antibiotics.

Restricted movement

The movement of small children and toddlers can be restricted due to their enlarged tummies so they may not develop as quickly as expected. A physiotherapist will be able to advise on exercises to encourage developmental progress.

Breathing difficulties

Ascites can sometimes make breathing difficult as there is less room for the lungs to expand. It's important to inform the medical team if this is a problem. Sitting in a more upright position can help as well as using extra pillows when in bed.

How is it treated?

There are different treatments available to try and deal with ascites, although these won't cure it.

Medicines

A diuretic may be prescribed. This is a medication which helps the body to get rid of extra fluid and will lead to needing to urinate (wee) more often. A diuretic called spironolactone is commonly used.

Children taking diuretics have an increased risk of becoming dehydrated (not having enough fluid) if they have vomiting or diarrhoea. This should be reported to their doctor, as well as the fact that they are on diuretics and have liver disease.

Nutrition and diet

A dietitian may advise changes to a child's diet. It's important to ensure the diet has enough nutrients and energy to support growth and to reduce salt. Too much salt can make ascites worse so it's important not to add salt to food and to avoid salty foods such as crisps.

Fluids

In some cases a reduction in the amount of fluid being drunk may be advised. This must not be done without medical advice and a dietitian and doctor will advise if this is necessary.

Albumin replacement

If the albumin in the blood stream is very low it might be necessary to give a child albumin. This is done through a drip and it may need to be repeated on several occasions.

Drainage of excess fluid (ascitic tap)

A thin needle is passed into the cavity in the abdomen. This is then attached either to a drain or to a syringe. A local anaesthetic is used to numb the area where the needle will be placed. This procedure is done using an ultrasound scan for guidance.

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

The 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, 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 was reviewed in December 2020. It is due to be reviewed by 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, is the voice of children, young people and young adults with a childhood liver disease and their families and funds vital medical research into childhood liver disease.

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 dedicated Facebook page called HIVE/HIVE+ for 13 – 24 year olds with a liver disease/transplant to make new friends, connect and share stories [childliverdisease.org/young-people/hive](https://www.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 www.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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Registered Charity Number: 1067331

Charity registered in Scotland: SC044387



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