



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

Portal hypertension and ascites in children



Help, support and guidance
for children and families

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Welcome

Welcome to your guide to understanding **portal hypertension and ascites**. 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 liver disease

Others who may also find this information useful:

- young people with liver disease
- healthcare professionals

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

- Introduction to liver disease in children

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



What is portal hypertension?

Hypertension means high blood pressure. Portal hypertension is high blood pressure in the portal vein. This is a major vein going into the liver.

What causes it?

Portal hypertension happens when blood has difficulty flowing through the liver. This causes pressure to build up in the blood vessels that carry blood to the liver.

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. 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?

Portal hypertension often causes no symptoms. However, if it causes serious symptoms or complications, it may be described as severe or complicated portal hypertension.

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

An enlarged spleen (splenomegaly)

The spleen is connected to the liver by a blood vessel that joins the portal vein. When there is increased pressure in the portal vein, this can also affect the spleen and cause it to become enlarged.

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)

Ascites can cause your child's tummy to become swollen or suddenly increase in size. Ascites is a build-up of fluid in the tummy (abdomen). Portal hypertension can cause ascites because the increased pressure in the blood vessels can push fluid into the space around 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 may become more visible under the skin of the tummy when it is swollen. They can also become more noticeable when blood flow through the portal vein is blocked. The blood then finds other routes back to the heart, causing new veins to develop. When these veins are close to the surface of the skin, they may be visible.

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. If bleeding occurs, your child may become tired, short of breath or look pale.

Blood might appear when your child is sick and may also appear in their stool (poo). This can make it appear bloodstained or black.

If there is heavy bleeding, your child may have tummy pain, feel dizzy or faint, or 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 your 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 your 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, and through the stomach and 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 your child is completely asleep) or under sedation (where your child has drugs to make them sleepy and relaxed).

Colonoscopy

This is similar to an 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 a blood test to check the number of platelets in their blood.



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 a drip will be inserted into a vein in your child's arm or hand. This is used to pass a special dye into the bloodstream. Your child will then lie on a bed that moves through a large, circular scanner while computerised pictures are taken.

Sedation or a general anaesthetic is not usually needed if your child is able to lie still during the procedure. However, younger children may need a general anaesthetic.

How is it treated?

If there are no symptoms or complications, then treatment may not be required, although your child's condition will be monitored.

Treatment may be needed if the varices are significant in size 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.

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.

Unlike in 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 your child, as well as any complications, 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. It 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 your 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 (a thin, flexible tube with a camera) is passed down the oesophagus (food pipe). A small needle is then passed through the endoscope to inject a special chemical into or next to the varices to help stop or prevent bleeding.

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

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

In hospital, your 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 observation. An endoscopy will be organised in the liver centre.

The hospital team caring for your child may:

- **monitor their blood pressure and pulse**
- **keep an eye on your child's vomit and poo, including any changes in colour or signs of blood**
- **find out how much blood has been lost**
- **give your child fluid or blood through a drip**
- **give medicines to stop the bleeding**
- **contact your liver team and discuss a 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 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. 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. Your child's medical team will be able to provide specific advice on a case by case basis.

If your child has portal hypertension and is travelling or flying, it is important to tell the medical team well in advance. Your child may need treatment before going away.

Other complications of portal hypertension

In rare cases, portal hypertension can also affect the lungs.

Some children may develop hepatopulmonary syndrome, which can cause:

- Low levels of oxygen in the blood. This happens when the blood vessels in the lungs become wider than usual, allowing oxygen-rich and oxygen-poor blood to mix.
- Portopulmonary syndrome, where there is increased blood pressure in the blood vessels of the lungs.

If either of these conditions are suspected, your child will need heart and lung tests to confirm the diagnosis. Liver transplantation is usually the preferred treatment when these complications are severe.



What is Ascites?

Ascites is a collection of fluid which gathers around abdominal organs and gives children a swollen tummy.

It can be caused by portal hypertension. It can also 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 feet, ankles and legs.

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 blood pressure in the veins around the abdominal organs, which leads to fluid leaking into the abdomen.

Salt being retained in the body

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

What are the signs and symptoms of ascites?

Signs and symptoms may include:

- a 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
- visible veins 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 a doctor examining your child or by using an ultrasound scan.



What are the possible complications?

Poor nutritional progress

Fluid in the abdomen can make children feel full very easily. They may eat smaller portions and may vomit because there is less space in the abdomen. A dietitian will be able to advise and support you.



Use this QR code to view our leaflet on nutrition or visit liveruk.org/nutrition-children



Infection

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

Any fever accompanied by pain or tenderness in the abdomen 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. This means they may not develop as quickly as expected. A physiotherapist can advise you on exercises and activities to help support your child's development and 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 help manage ascites, although they won't cure the underlying cause.

Medicines

A diuretic may be prescribed. This is a medication which helps the body get rid of extra fluid and will mean your child needs 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 recommend changes to your child's diet to make sure they are getting enough nutrients and energy to support healthy growth. It is also important to reduce the amount of salt in their diet. Too much salt can make ascites worse. Try not to add salt to food and avoid foods that are high in salt, such as crisps.

Fluids

Your child may be advised to drink less fluid. This should only be done if recommended by your child's medical team. A doctor and dietitian will let you know if this is needed and will advise you on how much your child should drink.

Albumin replacement

If the level of albumin in the blood is very low it might be necessary to give your 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.

Useful resources

Useful websites



Liver UK

www.liveruk.org

Information and support for everyone living with a liver condition.

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**

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.



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