Budd-Chiari syndrome

Children's Liver Disease Foundation
fighting childhood liver disease

A guide for parents, families, carers and healthcare professionals

How to say it: bud-kee-ah-ree

What is Budd-Chiari syndrome?

Budd-Chiari syndrome is a rare disease that affects the liver. It is caused by a blockage in the hepatic veins that carry blood out of the liver. In children, this mainly happens due to blood clots forming in the hepatic veins. Unfortunately, it means that blood cannot flow out of the liver in the normal way. This blocks blood flow from the liver to the heart and causes blood to back up inside the liver. It can lead to scarring (fibrosis) of the liver that gets worse over time.

It is also known as: Hepatic venous outflow tract obstruction

Budd-Chiari syndrome mostly begins in adults between the ages of 20 and 40. It affects 1.4 to 4 adults per million in western countries like the UK. We do not know the exact number of children and young people affected. But we do know it is even rarer in this age group. Budd-Chiari syndrome affects boys and girls in equal numbers.

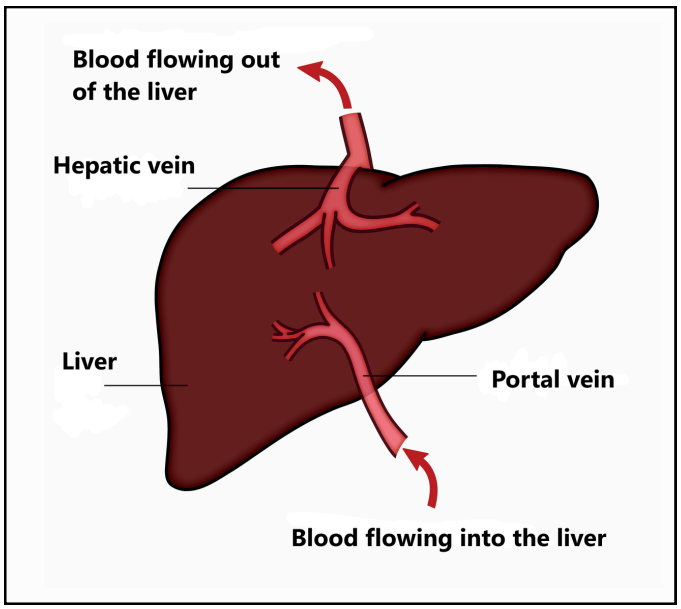
In children, Budd-Chiari syndrome is usually a long-term (chronic) condition. This means that a blockage in the veins carrying blood out of the liver develops slowly over time. But Budd-Chiari syndrome can also happen suddenly.

The symptoms of Budd-Chiari syndrome will vary from child to child. Some children will have no signs or symptoms. Sadly, other children will have severe symptoms.

Budd-Chiari syndrome is a serious condition that needs prompt treatment. This is because scarring (fibrosis) of the liver can become worse over time. This can lead to severe scarring, known as cirrhosis. Early diagnosis and treatment in children offers the best chance of a good outcome.

What causes Budd-Chiari syndrome?

Budd-Chiari syndrome is caused by a blockage in the hepatic veins that carry blood out of the liver. When blood has passed through the liver, it flows out through the hepatic veins. It then passes into the inferior vena cava. This is a large blood vessel that carries blood back to the heart.



Blockages in the hepatic veins are usually caused by either:

- a blood clot (thrombosis) or
- a web (a thin film) which has formed in the space inside the vein

Around 3 out of 4 children with Budd-Chiari syndrome have an underlying blood condition. You may hear this called thrombophilia. This is where blood in the body clots too easily. It increases the risk of blood clots in the blood vessels.

There are other possible causes of Budd-Chiari syndrome. But they are uncommon in children:

- cancer (often liver cancer)
- cysts or abscesses that put pressure on veins
- inflammation of a vein (phlebitis)
- hormonal factors (e.g. use of oral contraceptives)
- inflammatory diseases (e.g. Behçet's disease, Sjögren's syndrome or inflammatory bowel disease)
- liver trauma

What are the signs and symptoms of Budd-Chiari syndrome?

Signs and symptoms can vary a lot from child to child. Some children will have no symptoms. Some will have mild symptoms. Sadly, other children will have severe liver disease. The severity of symptoms will depend on the number of veins affected by blood clots. It will also depend how quickly or slowly they are blocked.

Signs and symptoms in children may include:

- tummy (abdominal) pain
- enlarged liver (hepatomegaly)
- fluid in the tummy (ascites)
- enlarged spleen (splenomegaly)
- tiredness (fatigue)
- itching (pruritus)
- feeling sick (nausea)
- being sick (vomiting)
- yellowing of the skin and whites of the eyes (jaundice)
- black, “tarry” poo (stools)
- swelling of the legs and ankles (oedema)
- vomiting blood (haematemesis)
- visible veins on the tummy wall

What are the possible complications of Budd-Chiari syndrome?

Portal hypertension, varices and ascites

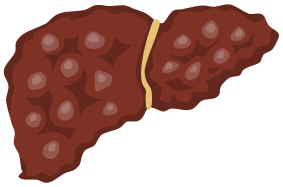
In Budd-Chiari syndrome, blood cannot flow out of the liver in the normal way. Blood backs up inside the liver and this causes high pressure in the portal vein. This is a vein that carries blood from organs in the tummy (abdomen) to the liver. This complication is called portal hypertension.

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

Portal hypertension can also cause fluid to build up in the tummy (abdomen). This is known as ascites.

Liver scarring (fibrosis)

Problems with blood flow out of the liver can lead to scarring (fibrosis) of the liver. A normal liver is soft and squishy. But as scarring builds up it becomes hard and bumpy. This can get worse over time. It can lead to severe scarring, known as cirrhosis. The speed and severity of scarring will vary from child to child. Liver scarring could happen during childhood. Or it may happen when your child becomes an adult.



Liver failure

Liver failure happens when large parts of the liver become damaged and scarred. The liver can no longer do its job. It is said to be failing. This is a late stage of liver disease. It is difficult for doctors to predict who will develop liver failure. And if this will happen during childhood or adulthood.

Hepatic encephalopathy

This is a rare side effect of severe portal hypertension. It happens when the liver can't remove toxins properly from the blood. This causes a rise in ammonia in the blood, which affects the brain. It can result in symptoms like confusion, forgetfulness and mood changes. In some children, hepatic encephalopathy can be very serious.

How is Budd-Chiari syndrome diagnosed?

Budd-Chiari syndrome can be hard to diagnose. The signs and symptoms can be similar to those seen in other liver conditions. Doctors will do different tests to rule these out before they make a diagnosis of Budd-Chiari syndrome.

Once a diagnosis is made, further tests will be done to help find out where the blood flow is blocked in the hepatic veins. This will help the medical team decide what treatments are possible for your child.

Budd-Chiari syndrome may be found by accident when your child is being tested for something else. Or when they are having tests for a complication called portal hypertension.

Tests used to help with the diagnosis may include:

- physical examination
- liver blood tests (also known as liver function tests / LFTs)
- blood tests to check for problems with blood clotting (thrombophilia)
- abdominal ultrasound scan

- computed tomography (CT) scan
- magnetic resonance imaging (MRI)
- endoscopy (if portal hypertension is suspected)
- hepatic vein catheterisation – to measure the pressure within veins
- angiography / venography – special X-rays to check blood vessels for blockages
- liver biopsy

Some of the tests will only be used if the doctors are unsure about the diagnosis.

How is Budd-Chiari syndrome treated?

Sadly, there is no cure for Budd-Chiari syndrome. The aim of treatment is to keep your child's liver function stable by helping blood flow out of the liver.

Treatment will vary from child to child. Your child will be offered treatments depending on the number of veins that are blocked and the location of the blockage. Early diagnosis and treatment offer the best chance for a good outcome.

The main aims of treatment are:

1. To make sure that blood can flow easily from the liver back to the heart
2. To stop blood clots forming in veins or increasing in size
3. To reduce the build-up of blood in the liver
4. To manage any complications from the condition

Remember:
Help and support is always available from CLDF

Anticoagulants

These are medicines that stop blood clots from forming or growing. They are sometimes called blood thinners. They are often the first treatment used in children. They are also used for long-term (chronic) Budd-Chiari syndrome. Some children will need lifelong treatment with these medicines.

Angioplasty / stent insertion

This treatment widens veins that are blocked by a blood clot. This stops blood building up in blood vessels and the liver. It allows blood to flow around the clot.

A thin plastic tube called a catheter is inserted into the blood vessel. It places a small balloon at the point of the blockage. The balloon is inflated to help blood flow through. A metal spring (stent) may be put in the space to keep it open.

Percutaneous hepatic vein recanalisation (HIVR) or inferior vena cava (IVC) recanalisation

HIVR and IVC recanalisation are types of angioplasty that are used to treat blocked veins inside the liver. They are carried out by a special doctor called an interventional radiologist. They are used if the clot can’t be reached using standard angioplasty. Like angioplasty, they are used to widen veins that are blocked by a blood clot. The exact treatment used will depend on the location of the blockage.

Endotherapy

There are three different types of endotherapy:

- banding
- glue injection
- sclerotherapy

They are used to treat swollen blood vessels in the food pipe (oesophageal varices). The swollen blood vessels are caused by portal hypertension.

Transjugular intrahepatic portosystemic shunt (TIPS)

This procedure is carried out by a special doctor called an interventional radiologist. A metal or plastic tube (stent) is placed between two large blood vessels in the liver (the portal vein and the hepatic veins). This creates a shunt, or bypass. It takes blood past any blocked veins. It may be used in older children. TIPS is used:

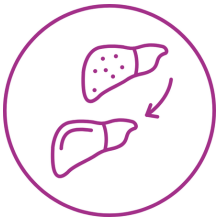
- to help manage severe portal hypertension
- if angioplasty and stenting are not possible
- in young people waiting for a liver transplant
- when a liver transplant is not possible

Meso-caval shunt

This is an operation carried out by a surgeon. It involves using a vein from the body or a synthetic vein to connect two blood vessels (the mesenteric vein and the inferior vena cava). This allows blood to escape from the liver and return to the heart.

Liver transplant

A liver transplant is usually only recommended if other treatments are no longer helpful, or if damage to the liver cannot be reversed. But a liver transplant is rare in children with Budd-Chiari syndrome.



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).

Where can I get further help or support?

CLDF can provide you with support and advice.
Contact us at families@childliverdisease.org.

This resource was reviewed in August 2025
It is due to be reviewed by August 2028

View CLDF resources

Contact us

Trusted Information Creator
Patient Information Forum

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