Budd-Chiari syndrome

BRITISH LIVER TRUST

Pioneering Liver Health

Budd-Chiari syndrome

This publication is for people diagnosed with Budd-Chiari syndrome and for those who would like to better understand the condition.

The British Liver Trust works to:

- support people with, and affected by, liver disease
- improve knowledge and understanding of the liver and related health issues
- encourage and fund research into new treatments
- campaign for better services and improved patient care
- increase awareness of the risk factors of liver disease and promote earlier diagnosis

All our publications are reviewed by medical specialists and people living with liver disease. Our website provides information and our Helpline gives advice and support on enquiries about liver health. Call the Helpline on **0800 652 7330,** general enquires on **01425 481320**, or visit **britishlivertrust.org.uk**

For the latest updates to this information, please refer to our website **britishlivertrust.org.uk**

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The liver

Your liver is your body's 'factory' carrying out hundreds of jobs that are vital to life. It is able to repair itself (even renewing large sections). **However, the liver's ability to repair itself is limited and continuous harm can lead to permanent scarring.** Your liver is very tough and able to function even when some of it is damaged, which means you may not notice any symptoms until your disease is quite advanced and noticeably affecting your health.

Your liver performs hundreds of functions. Importantly it:

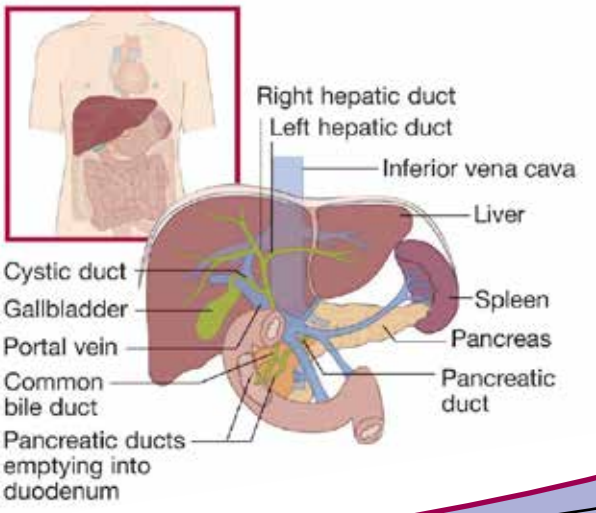
- filters and cleans the blood
- fights infections and disease
- deals with and destroys poisons and drugs
- makes vital proteins which make your blood clot when you cut yourself
- produces bile to help break down food in the gut
- processes food once it has been digested
- stores energy that can be used rapidly when the body needs it most
- regulates fat breakdown and distribution in the bloodstream
- stores sugars, vitamins and minerals, including iron
- gets rid of waste substances from the body
- produces and maintains the balance of some hormones
- produces chemicals – enzymes and other proteins – responsible for most of the chemical reactions in the body, for example repairing tissue
- repairs damage and renews itself (up to a point).

How liver disease develops

Your liver responds to harm by becoming inflamed. Any inflammation of the liver is known as hepatitis, whatever its cause. Sudden inflammation of the liver is known as acute hepatitis. When inflammation of the liver lasts longer than six months, it is known as chronic hepatitis.

Inflammation is part of the process of repairing damaged tissue. In a similar way to a scab forming over a skin wound, a temporary fibrous 'scaffold' forms while new liver cells regenerate. If your liver is repeatedly harmed, new liver cells cannot regenerate fast enough and the fibrous scaffold remains as a scar. This is called fibrosis, and can take a variable amount of time to develop.

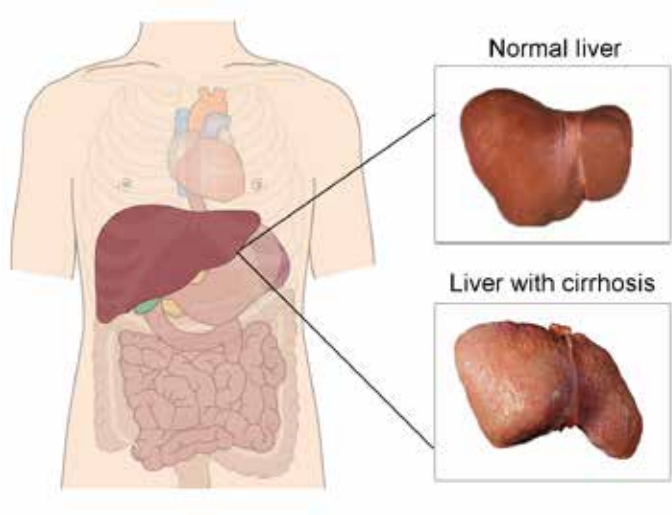
When fibrosis is present, your liver may be able to keep functioning quite well. Removing or treating the cause of the inflammation may reverse some, or all, of the fibrosis and prevent further liver damage.



If the harm to your liver continues, the inflammation and fibrosis can spread throughout your liver, changing its shape and affecting how well your liver cells work. This is known as **compensated** cirrhosis. Even at this stage, people can have no obvious signs or symptoms.

The scar tissue in cirrhosis interrupts the blood flow through the liver. As a result, the blood pressure in the veins in your abdomen is increased and may result in bleeding. Scar tissue in cirrhosis is difficult to remove and may be permanent. However, further progression can be halted and your cirrhosis stabilised, if the cause of the liver damage is removed.

Cirrhosis increases your risk of liver cancer and can lead to liver failure. If damage to your liver continues, it will become unable to function sufficiently (**decompensated** cirrhosis) and start to fail; this is sometimes referred to as 'end stage liver disease'. At this stage chemicals and waste products can build up in the body, commonly causing jaundice, ascites (a build-up of fluid in the abdomen) and hepatic encephalopathy (confusion and memory loss). In the final stages of liver disease the build-up of waste products may lead to multiple organ failure and loss of life.



What is Budd-Chiari syndrome?

Budd-Chiari syndrome (BCS) is a disorder affecting the liver and blood vessels, where blood flowing into the liver has difficulty in being able to flow out, leading to serious complications.

Blood flows into the liver from two blood vessels – the portal vein and the hepatic artery. After blood has passed through the liver, it flows out through the hepatic veins and into the inferior vena cava, a large blood vessel that carries blood back to the heart. In Budd-Chiari syndrome, this flow is partially blocked.

The immediate result is a build-up (congestion) of blood in the liver, as there is more flowing in than flowing out. The liver becomes swollen, tender to the touch and a source of discomfort. The congestion causes fluid to leak from the liver into the abdominal cavity. The resulting build-up of fluid in the abdominal area is called ascites, one of the commonest symptoms of Budd-Chiari syndrome. It may be visible as a bulge in your tummy area and can be uncomfortable and make it hard to breathe and eat normally.

If the blockage is extensive and also affects the inferior vena cava, there may be other places that become swollen. This is particularly likely in the ankles and legs and is called peripheral oedema.

Budd-Chiari syndrome also frequently results in yellowing of the skin and eyes, known as jaundice. This is due to the congestion and swelling of the liver.

Another effect is less visible but is also serious. This is called portal hypertension, where the blood pressure in the portal vein is increased, because of the blockage in the blood coming out of the liver. As happens elsewhere in the body, blocked veins cause collateral vessels to enlarge (dilate) to provide an escape route for the obstructed blood. In portal hypertension, blood may return to the heart by using extra (dilated) veins lining your oesophagus (gullet) and stomach where they are known as varices. These veins have fragile walls, which cannot easily handle the increased blood flow and may burst. This leads to internal bleeding and is referred to as 'bleeding oesophageal varices' which is a medical emergency. A rare side effect of severe portal hypertension is the development of confusion or even coma in severe Budd-Chiari syndrome. This is known as hepatic encephalopathy. See our leaflet on Hepatic Encephalopathy for more information.

There are a range of treatment options available for Budd-Chiari syndrome, and it is essential that it is diagnosed early before it causes liver damage or even liver failure. Early treatment can prevent liver failure, which may require an emergency liver transplant.

However, the disorder is very rare. It affects between 1 and 4 people in 1 million in Western Europe. Budd-Chiari syndrome occurs in people from all ethnic backgrounds and affects young females more frequently.

Budd-Chiari Syndrome takes its name from George Budd, the English doctor who described the first cases in 1845, and Hans Chiari, the Austrian pathologist who is credited with providing the first description of how it affects the body in 1899.

What causes Budd-Chiari syndrome?
Blood contains a large number of elements, which maintain the delicate balance between being able to stop bleeding from a wound on the one hand and clotting too quickly or unnecessarily on the other. The obstruction in the veins in Budd-Chiari syndrome is usually due to an imbalance in this system, where the blood clots too readily. Clots in blood vessels are called thromboses and the condition where there is a problem with blood clots is called thrombotic disease.

There may be a genetic cause or evidence from other tests as to why the blood is clotting too readily. Alternatively, it may be the enzymes that regulate blood clotting are not working properly, or the body may be producing too many red blood cells or platelets, making the blood too thick and sticky.

We know that over 80% of patients with Budd-Chiari syndrome have a condition that makes them more prone to thrombosis. Blood (haematological) disorders are the most common causes of Budd-Chiari syndrome. The most common one is a group of conditions known together as myeloproliferative neoplasms (MPN), which affect around half of all patients with Budd-Chiari syndrome. MPN consists of three disorders called polycythaemia vera, essential thrombocythaemia and myelofibrosis. These patients often have acquired gene mutations in their blood cells which can now be easily diagnosed with a blood test. The most common mutation is known as the JAK2 mutation. Other blood disorders include abnormalities of the clotting system caused by inborn or acquired defects of clotting proteins, and very rarely a condition called paroxysmal nocturnal haemoglobinuria.

Finding the cause is important for identifying the right approach to treatment and to prevent further thrombosis. However, in some people there is no obvious explanation for the problem.

A number of non-haematological conditions or factors are also linked with Budd-Chiari syndrome, including:

- tumours, most commonly liver cancer or hepatocellular carcinoma (HCC)
- chronic inflammatory diseases, such as Behçet disease, Sjögren syndrome or inflammatory bowel disease (IBD)
- pregnancy
- high dose oestrogen from using oral contraception.

It is possible that having an abscess, cyst or tumour can place direct pressure on your veins and increase the likelihood of a clot developing. This means that many existing disorders can be complicated by Budd- Chiari syndrome.

Web-like structures, called ‘membranous webs’, are found in the major hepatic veins or inferior vena cava, far more commonly in South-east Asia and the Middle East than in the UK. It is now thought that these represent the last remains of thromboses that have, for the most part, been cleared by the body’s repair systems. These can also be a cause of Budd-Chiari syndrome.

What are the symptoms of Budd-Chiari syndrome?

Budd-Chiari syndrome can appear or 'present' as an acute condition which develops rapidly with the following symptoms:

- abdominal pain in the upper right hand side of your abdomen (referred to as the 'upper right quadrant')
- ascites
- an enlarged liver due to the build-up of blood (hepatomegaly)
- swelling of legs and ankles
- cramp in legs and feet
- itching

More commonly, Budd-Chiari syndrome can develop in a chronic form, where people are likely to have long-standing ascites and an enlarged liver (hepatomegaly).

Very rarely, there is a fulminant form. This is a type of disease with a sudden and severe onset. In fulminant Budd-Chiari syndrome, ascites, hepatomegaly and kidney failure can occur with rapid liver failure.

Ascites are present in the majority of people because of portal hypertension.

Diagnosis

Like many liver diseases the symptoms of Budd-Chiari syndrome are non-specific, meaning that they can be caused by many conditions other than Budd-Chiari syndrome. Methods of diagnosis include abnormal liver blood tests and imaging tests using ultrasound and computerised tomography.

Ultrasound directs sound waves through your skin via a probe device as it is passed over your liver area. In Budd-Chiari syndrome it is used to obtain information about blood flow in your arteries and veins and this often gives enough information for the condition to be diagnosed.

A CT or MRI scan may also be used. This can obtain pictures from different angles around your body using computer processing and can also show cross-sections of your tissue and organs. This scan can show an enlargement of the liver and changes in the density of liver tissue due to abnormal blood flow. A liver biopsy may be necessary if other tests have helped doctors to identify the reason for liver problems.

Treatment

The aim of treatment for Budd-Chiari syndrome is to keep your liver function stable by maintaining the flow of blood out of the liver. It is very important that all patients with Budd-Chiari syndrome are treated in specialised hospitals with the required expertise, by a multidisciplinary team. This team of doctors includes liver specialists, blood disorder specialists (haematologists), and radiologists. They will be looking to:

- re-channel the blocked veins if possible
- prevent recurrence or progression of thrombosis by thinning the blood
- ease or 'decompress' the congestion of blood in your liver
- manage your ascites
- prevent further damage to your liver and allow liver cells to regenerate
- treat the underlying disease that caused the thrombosis in the first place.

Doctors will try to pinpoint the exact area where blood flow is obstructed to help them understand how your health may be affected (your 'prognosis'). The severity of Budd-Chiari syndrome can depend on where the clot is located and the number of veins that are affected.

Interventional radiologists will use X-ray techniques known as hepatic angiography or venography to examine arteries and veins directly to determine the location and severity of a clot. If the inferior vena cava is being examined, an X-ray called inferior cavography is used.

This is usually done under local anaesthetic and you are also likely to be given sedation. It is possible you may be asked to stay in hospital overnight.

These procedures involve inserting a thin, flexible tube (catheter) into a blood vessel through an easily accessible vein in the arm, neck or groin. A dye, referred to as a 'contrast dye' or 'contrast medium', is then injected through the catheter to light up the blood vessels to make them easier to see in the X-ray. In some cases the scan shows that only the end portion of the vein is blocked and that much of the vein remains clear. In other cases, the vein is more blocked and doctors will need to get to the vein via a tube put into the liver from the tummy.

Once the tube gets to the blockage, doctors can remove the clot and open the vein. This is called venoplasty or angioplasty. The vein is opened with balloons and the interventional radiologist often decides to place a metal spring (stent) within it to keep it open.

If the clot within the hepatic veins was formed recently and is difficult to remove, the catheter tube may be left in the hepatic vein for a day or two to allow clot-busting drugs (thrombolysis) to get rid of the clot. When venoplasty/stent succeeds it usually leads to rapid improvement in the patient's overall condition.

In some patients, the blockages are too extensive for venoplasty to work. Doctors will look at which treatments are suitable for the symptoms, depending on how severe they are. When ascites or bleeding varices are troublesome, a radiological procedure called TIPSS (see Useful words section) may be offered. This lowers pressure in the portal vein.

In this procedure a metal or plastic tube (stent) is passed across your liver to make a shunt, or bypass, to make your blood travel straight from the portal vein past the blocked hepatic veins into the inferior vena cava which carries the blood back to the heart. This is done using a needle guided by a catheter inserted through a tiny puncture in your neck. This procedure may be performed either under general anaesthesia or with sedation.

Occasionally another operation may be offered which reverses the flow in the portal vein so that it is taking blood out of the liver rather than into it. The liver still receives enough blood from the hepatic artery to function adequately. This operation uses a vein from the neck to make a new connection which allows the blood to escape from the congested liver. The vein from the neck is grafted on between the mesenteric vein, a vein from which blood normally flows into the portal vein and the inferior vena cava, and is called a meso-caval shunt.

These surgical procedures and TIPSS can be effective, however, there is a risk they can create an additional problem. The shunts mean that less blood goes through the liver to be cleaned of toxins. As a consequence, there is a risk these toxins will build up and this can cause a condition called hepatic encephalopathy. The symptoms of this can include mental confusion, tremors or drowsiness. Hepatic encephalopathy can be treated using laxatives such as lactulose, or antibiotics, to help your body remove these toxins. In very few cases the TIPSS stent can be altered in size to try and reduce the encephalopathy.

Where membranous webs are the cause of Budd-Chiari syndrome, angioplasty may be used to relieve the obstruction. This is a technique in which a catheter with a small balloon at the end is inserted into the blocked artery. The balloon is then inflated to widen the artery and allow the blood to flow more freely. This may have to be carried out in a number of blood vessels.

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Haematological Treatment

Anticoagulation

Most experts now recommend that patients with Budd-Chiari syndrome should receive life-long anticoagulation therapy, because Budd-Chiari syndrome often means that people's blood has a tendency to clot too readily. Anticoagulation therapy involves taking medicines and being closely monitored with regular blood tests to check that the levels of clotting are right, to make sure the blood does not clot too readily (which could cause thromboses or blockages) or not well enough (which could cause bleeding). Anticoagulation also treats conditions with clotting defects and paroxysmal nocturnal haemoglobinuria

Treatment of Myeloproliferative Neoplasia (MPN)

In patients with MPN, the haematologist will assess the patient's disease type and the risk of further thrombosis in order to recommend appropriate treatment. Assessment includes a molecular test from the blood for acquired mutations, such as JAK2, and a biopsy of the bone marrow. The latter takes no more than 5 minutes and is performed under local anasthaesia (using the same drug as at the dentist). The treatment for MPN usually involves taking drugs long term to control the activity of the bone marrow and blood counts. This type of treatment is called cytoreductive therapy and consists of either taking tablets or injections, which require monitoring on a regular basis.

Liver transplantation

A liver transplant is usually only recommended if other treatments are no longer helpful and your life is threatened by end stage liver disease. In Budd-Chiari syndrome, a liver transplant may be required when:

- an onset of fulminant Budd-Chiari syndrome causes your liver to fail
- your liver stops performing all of its functions adequately, a condition called decompensated cirrhosis
- shunt procedures cannot prevent a further deterioration in your condition.

Liver transplantation is a major operation and if it is not an emergency treatment, you will need to plan it carefully with your medical team, family and friends. Liver transplants offer a good prospect of a full recovery.

Prognosis

Being diagnosed with Budd-Chiari syndrome can be worrying and frightening, as it is a serious disease. For many people, the diagnosis can be the end to a long process of feeling something was wrong. Diagnosis also gives patients an opportunity to have effective treatment that can improve quality of life. Treatments aim to reduce the risk of the disease progressing to a serious outcome, and to improve the quality of life. How well you respond to treatment will depend on your age and the severity of the disease at diagnosis. Over 80% of patients do respond well and are still alive 10 years after diagnosis. You should speak to your own doctor about your prognosis.

Looking after yourself

Like other people with liver disease, you have a higher risk of infection and a higher risk of infections leading to serious complications. You can help protect yourself by maintaining high standards of hygiene and getting vaccinated before travel and for infections such as flu and pneumococcal infection. You may also find that you get tired more easily and need to plan your day to ensure you have enough energy for all your activities. To manage your ascites you will be given diuretics and placed on a low-sodium diet. This will mean no added salt on your food and also only eating foods that are very low in salt, avoiding processed and cured foods and snacks such as crisps and biscuits. A dietitian can help you adjust your diet.

Underlying the development of Budd-Chiari syndrome, there may be another blood problem that results in the blood being unusually sticky and likely to form clots. This means you will end up having to cope with a separate diagnosis and receive two sets of lifelong treatments. However, the treatments you receive can make you feel better and prevent further blood clots.

Living with a long-term serious illness means making adjustments to your way of life and these often take time and patience to achieve. These will affect your family and friends so it may be useful to keep them informed about your conditions and feelings (as you judge appropriate), so that they are best able to help and support you. Some people find it useful to join support groups or online forums for people with liver disease or other life-limiting conditions. The British Liver Trust website has more details – please visit: britishlivertrust.org.uk/find-support/ or email helpline@britishlivertrust.org.uk

Complementary and alternative medicines

Many complementary and alternative medicines are available that may ease the symptoms of liver disease. But certain medications used in non-liver related disease can damage the liver. At present, healthcare professionals are not clear on the role and place of some therapies in managing liver disease. More research needs to be done on the use of these therapies. You may wish to discuss the use of these therapies with your doctor.

Useful words

Anticoagulation – therapy to reduce blood clotting and thrombosis.

Acute – a short sharp illness that may be severe but from which most people will recover in a few weeks without lasting effects.

Ascites – an accumulation of fluid in the peritoneal cavity which surrounds the bowel, leading to enlarged, swollen and tender abdomen.

Chronic – an illness that lasts a long time (more than six months), possibly for the rest of a person's life.

Clot – as in blood clot, a lump made from blood cells that form to prevent bleeding also referred to as a thrombus.

Cyst – an abnormal, fluid-filled balloon-like structure (sac) that can grow in any part of your body.

Enzyme – a substance, usually a protein, produced by the body to help speed up a chemical reaction.

Fulminant – sudden and severe onset of symptoms.

Hepatic – anything relating to the liver.

Inferior vena cava – the large vein that carries blood back to the heart from the lower part of the body.

Inflammation – the first response of the immune system to infection, commonly characterised by heat, swelling, pain and tenderness.

Membranous – made of or similar to the tissue of a membrane, i.e. thin, pliable and semi-transparent.

Occlusion – obstruction or closing off.

Portal vein – the vein that carries blood from the bowel and the spleen to the liver.

TIPSS – stands for transjugular intrahepatic portosystemic stent shunts, a surgical procedure to lower the pressure in the portal vein.

Thrombosis – the formation or presence of a blood clot (thrombus) inside a blood vessel, obstructing the flow of blood through the circulatory system. People with a thrombosis may be said to have thrombotic disease.

Tumour – an abnormal lump or swelling of tissue caused by an uncontrolled build-up of cells.

Vascular – a term relating to veins, arteries and smaller blood vessels in the body.

Further information

Please refer to the Trust website for details of patient organisations and support groups specialising in specific liver conditions, that you may find helpful.

The British Liver Trust publishes a large range of leaflets about the liver and liver problems, specially written for the general public.

Leaflets that you may find particularly helpful include:

- *Cirrhosis of the liver*
- *Diet and liver disease*
- *Liver disease tests explained*
- *Liver transplantation*

Contact us for more information:

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We hope you have found this publication helpful

All our publications are reviewed by medical experts and people living with liver disease. If you have any feedback on this publication please email the Trust at **info@britishlivertrust.org.uk**

The British Liver Trust is proud to be recognised as a provider of expert liver health information, but to do this we must depend on the kind donations of our supporters. The Trust receives no government aid, yet strives to fill the growing need for liver health information in the UK.

We are a small charity, and your donation can make an important difference.
A gift of £5 could help us answer patient calls to our helpline

A gift of £20 could help us to set up a new patient support group

A gift of £50 could support the costs of a new patient guide or leaflet

Gifts can be made:
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Remember to indicate your Gift Aid preference.

By texting LIVR10 £10 (or other amount) to 70070

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If you have questions about making a donation, please call **01425 481320** or email **fundraising@britishlivertrust.org.uk**

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