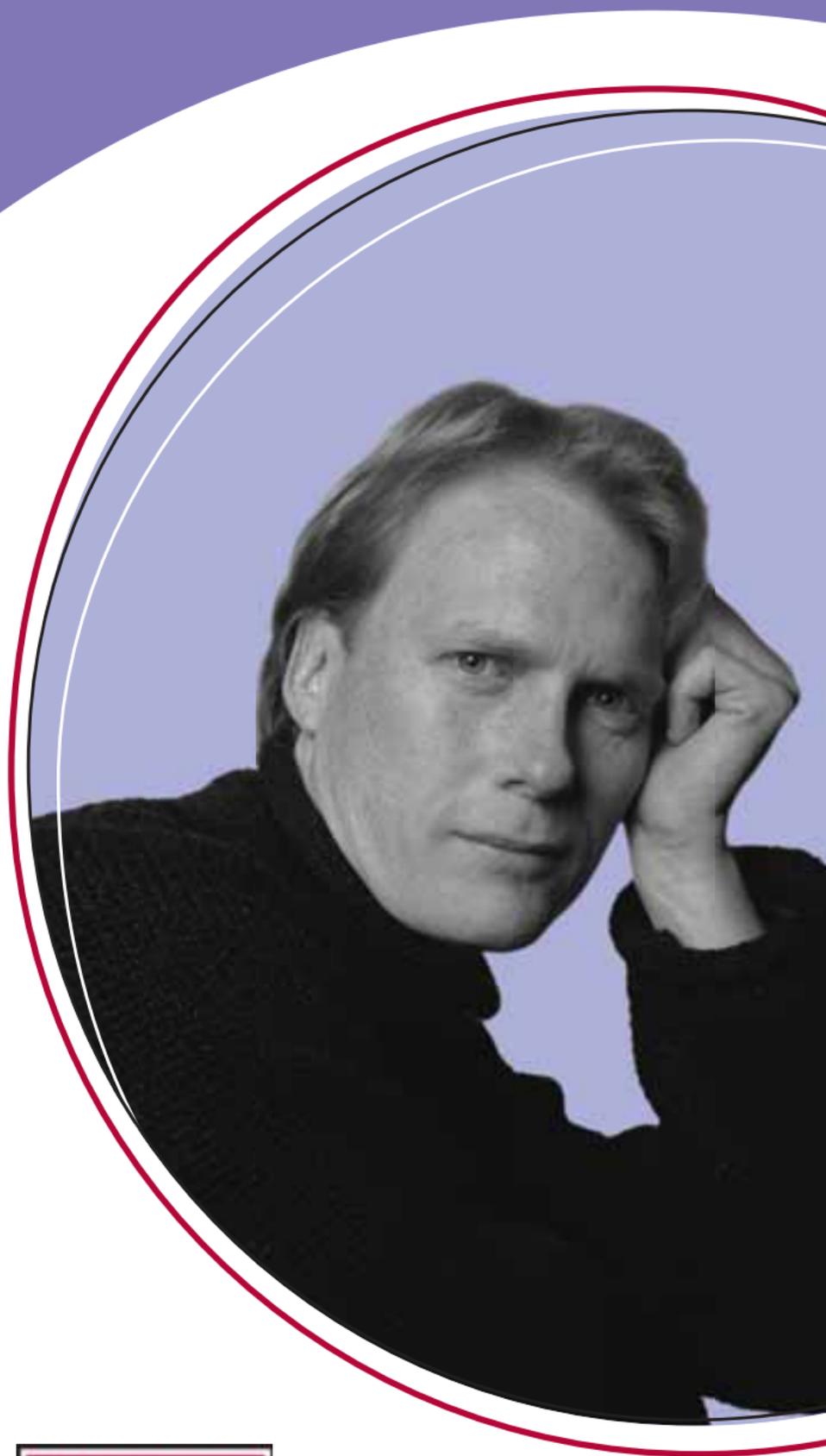


Budd-Chiari syndrome



BRITISH
LIVER
TRUST

Fighting liver disease

Budd-Chiari syndrome

The British Liver Trust works to:

- support people with all kinds of liver disease
- improve knowledge and understanding of the liver and related health issues
- encourage and fund research into new treatments
- lobby for better services.

All our publications are reviewed by medical specialists and people living with liver disease. Our website provides information on all forms of adult liver disease and our Helpline gives advice and support on general and medical enquiries. Call us on **0800 652 7330** or visit **www.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 very tough and able to continue to function when most of it is damaged. It can also repair itself – even renewing large sections.

Your liver has around 500 different functions.

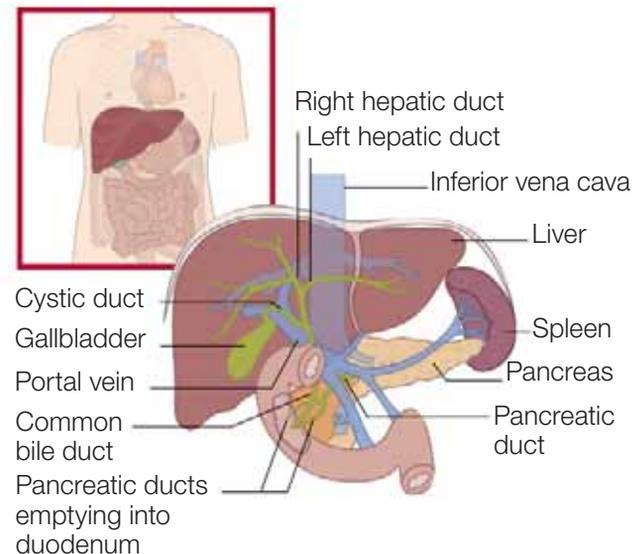
Importantly it:

- fights infections and disease
- destroys and deals with poisons and drugs
- filters and cleans the blood
- controls the amount of cholesterol
- produces and maintains the balance of hormones
- produces chemicals – enzymes and other proteins – responsible for most of the chemical reactions in the body, for example, blood clotting and repairing tissue
- processes food once it has been digested
- produces bile to help break down food in the gut
- stores energy that can be used rapidly when the body needs it most
- stores sugars, vitamins and minerals, including iron
- repairs damage and renews itself.

How liver disease develops

Liver damage develops over time. Any inflammation of the liver is known as hepatitis, whatever its cause. Sudden inflammation of the liver is known as acute hepatitis. Where inflammation of the liver lasts longer than six months the condition is known as chronic hepatitis.

Fibrosis is where scar tissue is formed in the inflamed liver. Fibrosis can take a variable time to develop. Although scar tissue is present the liver keeps on functioning quite well. Treating the cause of the inflammation may prevent the formation of further liver damage and may stop or reverse some or all of the scarring.



Cirrhosis is when inflammation and fibrosis has spread throughout the liver and disrupts the shape and function of the liver. Even at this stage, people can have no signs or symptoms of liver disease. When the working capacity of liver cells has been badly impaired and they are unable to repair or renew the liver, permanent damage occurs.

Cirrhosis can lead to liver failure or liver cancer. All the chemicals and waste products that the liver has to deal with build up in the body. The liver is now so damaged that the whole body becomes poisoned by the waste products and this stage is known as end stage liver disease. In the final stages of liver disease the building up of waste products may cause multiple organ failure and lead to death.

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.

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 BCS, this flow is partially blocked, sometimes referred to as 'an obstruction of hepatic venous outflow'.

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 blocks the large blood vessel which returns blood to the heart from the lower half of the body (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.

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 other veins to enlarge (varicose veins) to provide an escape route for the obstructed blood. In portal hypertension, blood may return to the heart by using extra veins lining your oesophagus 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.

There are a range of treatment options available for BCS, and it is essential that it is diagnosed early before it causes liver damage or even liver failure.

However, the disorder is very rare. Its prevalence is not exactly known but one person in 100,000 is the most common estimate. BCS occurs in people from all ethnic backgrounds and affects both sexes equally.

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

How does blood normally flow to and from the liver?

The liver normally receives 70% to 80% of its blood from the portal vein, with the remaining blood received via the hepatic artery. The portal vein brings blood from the intestines and spleen for processing within the liver before circulating to other parts of the body, and the hepatic artery brings oxygenated blood straight from the heart. Blood leaves the liver through the hepatic veins.

Most people have two groups of hepatic veins:

- An upper group consisting of three large veins.
- A lower group of generally smaller veins.

Between them, these veins drain the blood from the liver into the inferior vena cava, which travels up behind the liver taking blood back to the heart.

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 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. The body may be producing too many red blood cells or platelets. It may not be producing enough of the substances that regulate blood clotting, or these may not be working properly. Finding a cause may be useful in identifying the right approach to treatment. However, in some people there is no obvious explanation for the problem.

A number of other conditions or factors are linked with BCS, including:

- tumours, most commonly the liver cancer 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 BCS.

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 BCS.

What are the symptoms of Budd-Chiari syndrome?

BCS 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, BCS 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 BCS, 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 BCS are non-specific, meaning that they can be caused by many conditions other than BCS.

Methods of diagnosis include liver function tests (LFTs) and imaging tests with ultrasound and computerised tomography (CT).

Ultrasound, the same technology used to confirm all is well in pregnancy, directs sound waves through your skin via a probe device as it is passed over your liver area. Anything solid will bounce back as a reflected sound wave via the probe and will be turned into an image that can be seen on a screen. In BCS, a type of scan known as 'Doppler' ultrasound is used to obtain information about blood flow in your arteries and veins and this often gives enough information for BCS to be diagnosed.

A CT scan may also be used. This can obtain pictures, called tomograms, 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 (thickness) of liver tissue due to abnormal blood flow.

A liver biopsy may be necessary if other tests have not helped doctors to identify the reason for liver problems. During a liver biopsy a tiny piece of the liver is taken for study. A fine hollow needle is passed through the skin into the liver and a small sample of tissue is withdrawn. The test is usually done under local anaesthetic and may mean an overnight stay in hospital, although some people may be allowed home later the same day.

Treatment

The aim of treatment for BCS is to keep your liver function stable by maintaining the flow of blood out of the liver. Doctors will be looking to:

- re-channel the blocked veins if possible
- prevent recurrence or progression of thrombosis
- 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.

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 BCS can depend on where the clot is located and the number of veins that are affected.

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 doctor may decide 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 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 what treatments are suitable for the symptoms, depending on how severe they are. When ascites or bleeding varices are troublesome, a surgical procedure called TIPSS 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 is not painful in itself but you will be given a local anaesthetic and usually some sedation.

Occasionally another operation may be offered which reverses 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 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.

Where membranous webs are the cause of BCS, 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.

Anticoagulation

Most experts now recommend that patients with BCS should receive life-long anticoagulation therapy, because BCS 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).

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 BCS, a liver transplant may be required when:

- an onset of fulminant BCS 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.

Looking after yourself

Being diagnosed with BCS 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, including the risk of death and to improve the quality of life.

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 dietician can help you adjust your diet.

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.

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 condition 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.

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.

Who else can help?

AntiCoagulation Europe

PO Box 405

Bromley

Kent BR2 9WP

Tel: 020 8289 6875

Fax: 0208 464 2417

Email: anticoagulation@ntlworld.com

Web: www.anticoagulationeurope.org

A registered charity committed to the prevention of thrombosis and the provision of information and support for people already receiving anticoagulant and anti-platelet therapy.

Further information

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*
- *Life after liver transplant*

Contact us for more information:

Tel: 0800 652 7330

Email: info@britishlivertrust.org.uk

Web: www.britishlivertrust.org.uk

This leaflet is for information only. Professional, medical or other advice should be obtained before acting on anything contained in the leaflet as no responsibility can be accepted by the British Liver Trust as a result of action taken or not taken because of the contents.

Special thanks

Professor Elwyn Elias, Consultant Physician,
Queen Elizabeth Hospital, Birmingham.

Can you make a difference?

Liver disease is increasing alarmingly and the need to do more is greater than ever before...

For the British Liver Trust to continue its support, information and research programme, we need your help. We raise funds from many sources and a large proportion is donated by voluntary contributions. If you would like to send a donation it will enable us to continue providing the services that people need.

If you can help, please fill in the form on the page opposite.

If you wish to help us further with our work by organising or participating in a fundraising event or becoming a **“Friend of the British Liver Trust”** please:

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Make a donation via our website at
www.britishlivertrust.org.uk

or write to
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2 Southampton Road,
Ringwood, BH24 1HY

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I am interested in leaving the Trust a legacy. Please send me more information

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Your name and address will be added to our computer database ensuring you are sent the latest information. If you do not wish to receive further information, please tick here.



This patient information leaflet was produced with support of a donation from The James Tudor Foundation.

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