Portal Hypertension and Varices
a guide
Welcome

This leaflet has been written specifically for:

■ Parents/carers of a child with portal hypertension
■ Young people with portal hypertension

Others who may find this leaflet helpful are:

■ Relatives and friends of families
■ Healthcare and allied professionals, school, college, university and nursery teams

This leaflet aims to:

■ Explain portal hypertension
■ Explain the symptoms, diagnosis and treatments

You may find it helpful to also read the following CLDF leaflets:

■ Guide to the liver
■ Glossary of terms
■ Routine investigations for liver disease

Children’s Liver Disease Foundation (CLDF) also has leaflets in its support series which are available to download from our website – childliverdisease.org. Leaflets can be mailed to UK patients free of charge, our contact details are on this leaflet. You may find it helpful to have a copy of CLDF’s making the most of an appointment leaflet which will help you prepare for appointments and meetings following discharge.

We also have the following information packs available, free of charge:

■ Essential 5 – CLDF's starter kit of vital information
■ Education – a pack supporting children and young people under 18 in an educational setting
■ GP Practice – a pack for families to give to their GP providing a range of information on childhood liver disease
■ Friends and Relatives
■ Yellow Alert – a pack to support CLDF’s Yellow Alert Campaign for early diagnosis of liver disease in newborn babies

All are available on request to UK families and young adults.

Overseas families should contact CLDF to discuss their literature needs.
Introduction

To understand portal hypertension, it is helpful to look at the way blood is circulated around the body. There are two pathways which are particularly significant to this leaflet:

- The systemic circulation — Arteries carry blood rich in oxygen from the lungs to all other parts of the body, this ‘arterial’ blood is at relatively high pressure. This blood then returns to the heart through veins at a lower pressure.

- The portal circulation — Blood from the heart goes through the digestive system absorbing the nutrients from digested food and carries them into the liver via the portal vein. The blood is processed by the liver and then returns to the heart. See diagram 1.

What is portal hypertension?

Many people suffer from ‘hypertension’; this usually means abnormally high blood pressure in the systemic circulation. Systemic blood pressure can be measured by placing an air filled cuff around the upper arm, which is connected to a monitoring device. This is not related to portal hypertension.

Portal hypertension refers to abnormally high blood pressure in the portal circulation; this cannot be measured using a cuff. The degree of portal hypertension is assessed by the symptoms it causes.

What causes portal hypertension?

Portal hypertension is caused by obstruction of the blood flow into, through or out of the liver. This obstruction leads to ‘backpressure’ (see diagram 2).
The main causes of obstruction are:

- A blockage in the portal vein, often due to thrombosis (blood clot). This can occur as a complication of severe illness in the first few weeks of life or because of a genetically inherited disorder, which causes the blood to clot more easily than it should, or as the result of an unknown cause. In most cases, the liver continues to function normally giving normal liver function test (LFT) blood results. See diagram 4.

- Scarring in the liver, often as a result of cirrhosis (destruction of liver cells) which occurs in many types of liver disease. Usually, liver function and LFTs deteriorate as a result of progressive cirrhosis. See diagram 5.

- Increased resistance to blood leaving the liver, because of an obstruction in some or all of the hepatic veins, or as a result of heart disease. Although the liver may often be healthy to begin with, it will quickly be affected by the obstruction and deteriorate. See diagram 6.
What are the signs and symptoms of portal hypertension?

The signs and symptoms of portal hypertension result from the increased pressure on the other organs in the portal circulation. These may include any, or all, of the following:

- **An enlarged spleen (splenomegaly).**
  The backpressure on the spleen causes it to increase in size. One of the jobs of the spleen is to break down old blood cells as new ones are made. When the spleen is enlarged it removes more cells from the circulation than it should. This particularly affects the platelets that help with blood clotting. When the platelet count is low, bruising and nosebleeds are more likely and they may be more severe.

- **Abdominal swelling** — This may be due to the enlargement of the spleen, and/or liver, in which case it occurs gradually and is not severe. A more sudden increase in tummy size may occur if ascites develops. Ascites is an abnormal collection of fluid in the abdomen. It may occur in portal hypertension due to the increased pressure in the blood vessels forcing out some of the fluid part of the blood through the thin blood vessel walls into the spaces between the abdominal organs. CLDF has a leaflet on ascites.

- **Prominent veins under the skin of the tummy** — This is often because of the abdominal swelling but there can be other reasons. Blood from the portal circulation needs to return to the heart (see diagram 2). However, when the usual route is blocked, additional veins can develop to ‘bypass’ the obstruction and link into the systemic circulation. If this occurs near to the skin surface the veins can be easily seen.

- **Varices** — The ‘bypass veins’ described above, also known as collaterals, can occur in many places through the digestive tract and may swell to cause varices, which look similar to varicose veins that some people have in their legs. When these varices occur in the oesophagus or stomach, they have thin walls and can bleed.

- **Signs of bleeding** — Bleeding can cause tiredness, breathlessness and a pale appearance. There may be blood in the child’s vomit, or stools can be blood stained or black. If there is a sudden large bleed, abdominal pain, dizziness and sometimes collapse can occur. Any of these symptoms must be reported to their doctor immediately.

- **Diarrhoea, poor weight gain** — The blood vessels in the lining of the intestine may become swollen as the blood flows through them under pressure. This can reduce absorption of digested food, leading to poor weight gain, frequent stools and diarrhoea.

How is the diagnosis made?

A range of tests can be carried out to make a diagnosis:

- **General physical examination.**

- **Blood tests including full blood count (FBC), blood clotting tests (PT/PTT) and liver function tests (LFTs).**

- **Ultrasound scan to measure: the size of liver and spleen, the direction and speed of blood flow in veins and HARI — hepatic artery resistance index.**

- **Endoscopy** — sometimes called oesophago-gastro-duodenoscopy (OGD). This may be done under sedation or general anaesthetic. A flexible tube (endoscope) is passed through the mouth, down the gullet, through the stomach and into the gut, allowing the
doctor to look directly at the lining for signs of varices.

- Sigmoidoscopy or proctoscopy. In the same way as an endoscopy, another tube can be passed up the anus to examine the lower gut. This is only done if the patient has passed fresh red blood in their stools and will be performed under sedation or general anaesthetic.

For children who have portal hypertension without associated liver disease, there are some other investigations that may be required:

- Angiography. Under anaesthetic, a fine tube is passed into a blood vessel in the groin; a special dye is injected into this. X-rays are then taken to give a ‘map’ of the child’s portal vein and its branches.

- Magnetic resonance imaging (MRI). An injection is given into a vein in the child’s arm. The child must then lie still on a bed inside a long circular tube while computerised pictures are taken. Sedation or anaesthetic are not normally required provided the child is able to stay still.

What is the treatment of portal hypertension?

If the symptoms are mild, then no treatment is needed. If there is an underlying condition contributing to the portal hypertension that will be treated appropriately. If the condition leads to bleeding, this will require treatment.

If the portal hypertension causes diarrhoea or poor weight gain, the dietitian will advise you on changes to the diet. This is done to try and ensure your child absorbs adequate nutrients and calories for growth. Sometimes an alternative method of feeding will need to be considered.

Does portal hypertension always lead to bleeding?

No, portal hypertension can be present for years without causing any problems. The ‘bypass veins’ or collaterals may develop in areas of the body where they will not bleed, so the blood is safely diverted into the systemic circulation, naturally reducing the pressure in the portal circulation. Bleeding is only likely if varices form in places where the walls of the veins are thin, such as the oesophagus or stomach.

What is the treatment for varices?

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

What is the treatment for active bleeding?

If a child/young person is vomiting blood or passing blood in their stools, they must be taken to the NEAREST hospital for immediate treatment. Unless someone else can drive and the hospital is nearby, it is best to call 999 and go by ambulance. REMEMBER they may vomit again. Make sure they do not choke on the vomit, especially if they are very young. If possible, take a sample of the vomit or stool with you to the hospital. Remember to tell them about the diagnosis of portal hypertension, as well as the present symptoms.

In hospital the child/young person, will be assessed and given treatment as required. Some bleeds are very small and do not need a hospital admission. However, in most cases a hospital stay will be needed for observation, and to ensure the bleeding stops.

Some of the following things will be done:

- Monitoring of blood pressure and pulse.
Observation of vomiting and colour of stools.

Blood test to indicate how much blood has been lost.

Giving special fluids/blood intravenously (through a drip) to maintain the blood volume.

Giving medicines intravenously to actively stop the bleeding (e.g. octreotide).

Once the child/young person is stable, an endoscopy may be performed to look at the varices that are bleeding. The child/young person may be transferred to a specialist centre for this to take place. In an attempt to stop the bleeding, the varices can be injected with chemicals that block the small veins (sclerotherapy). This treatment forces the blood to use deeper veins, which cannot bleed. Another possible treatment is called banding, this involves shrinking the varices by placing special rubber bands around them.

To reduce the chance of the varices bleeding again, it may be appropriate to have repeated treatments (sclerotherapy/banding) over future weeks. It may be possible for the child/young person to go home in between treatments.

If the portal hypertension has been caused by a blood clot in the portal vein, can the clot be removed?

No, almost never. By the time portal hypertension has been diagnosed and the blood clot has been identified as the cause of the blockage, the blood clot will usually have settled and the surrounding tissue will have grown into it. This means the vessel that the clot is in would need to be removed completely. In this instance, it may be better to think of the problem as an abnormality rather than a simple blockage. However, some techniques allow a ‘mainly anatomical’ correction of the problem by creating a new blood route towards the liver (shunt surgery).

What are the long-term indications with portal hypertension?

Medicines and endoscopy treatments do not cure portal hypertension but they may prevent bleeding indefinitely. If these measures are not successful, other surgical approaches may be necessary. If these options need to be considered, they will be fully explained to you.

Do children/young people with portal hypertension need to take special precautions?

The most important thing is to be aware of the potential problems, e.g. bleeding, and make sure that any other carers, for example, teachers, are also well informed. If the spleen is enlarged, there is an increased risk of it being damaged if it receives a hard direct blow. Children/young people with portal hypertension may therefore be advised to avoid activities where it is more likely that they could receive a high impact blow to the abdomen.

It is also very important not to limit activities unnecessarily. Try to find suitable activities that they enjoy.

For children/young people with a very low platelet count, below about 50, it may be advisable to avoid any kind of activity where bumps and bruises are likely to occur. This should really be assessed on an individual basis; you are advised to discuss the matter with your specialist nurse or doctor.

Some medicines can irritate the lining of the gut and make bleeding more likely; common examples include aspirin, ibuprofen/Junofen. These products should be avoided. Paracetamol (Calpol) is a suitable alternative and should be given as directed on the container.

A doctor, who is fully aware of the diagnosis, should prescribe all other medicines.
Is there a charity taking action against the effects of childhood liver disease?

Yes. Started by families in 1980, Children’s Liver Disease Foundation (CLDF) leads the way in fighting all childhood liver disease.

CLDF funds vital research, develops information and awareness programmes and supports families, young people and adults diagnosed in childhood who are living day in, day out with a liver condition or transplant. And its work has made a big difference and continues to help save lives.

CLDF has so much to offer you: information, the opportunity to meet other families, events and regular updates. To find out more, call, email or write today:

Children’s Liver Disease Foundation,
36 Great Charles Street, Birmingham,
B3 3JY
0121 212 3839
Main site: childliverdisease.org
Young people’s site: cldf-focus.org
info@childliverdisease.org

What are the roles of CLDF’s Family and Young People’s teams?

CLDF’s Family 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.

Our parents say . . .

“. . . We don’t know how we would have coped without CLDF’s care and support. They have been just fantastic from the outset — tremendous people, who are compassionate and so positive. They really care about families and children struggling with liver disease.”

“When Emily was very ill we felt we were on the sidelines, knowing we couldn’t influence the outcome and not in control. Getting involved in fundraising is something you can control and achieve a positive result. I really took comfort from that.”

Our young people say . . .

“Knowing CLDF is there is what I need. I can call whenever I want. Whatever I think and feel is listened to. Even when I called to tell them it was my birthday!”

“I feel really well. It’s great that CLDF has given us the chance to meet other young people outside of the hospital and have a fun time. I want them to do more things like this.”

Families Team
families@childliverdisease.org
0121 212 6023

Young People’s Team
youngpeople@childliverdisease.org
0121 212 6023
Donation, Regular Gift & Gift Aid Declaration Form

To make a one-off gift or set up a direct debit gift online, go to childliverdisease.org

I’d like to make a gift to CLDF

How much?  £10 □  £20 □  Other £ ............

□ I enclose a cheque made payable to Children’s Liver Disease Foundation

□ I wish to pay by card — MASTERCARD / VISA / DEBIT CARD (delete as appropriate)

Card No. ......................................... Expiry Date ...... /...... /......

Name on Card  ............................................. Security Number: ................ (back of card)

I’d like to make a regular gift by direct debit to CLDF

How much?  £5 □  £10 □  £20 □  £25 □  £50 □  other £ ...........

How often?  □ monthly  □ quarterly  □ half-yearly  □ annually

My bank details:

Bank name: ...................................................... Branch name: ..............................................................

My bank address: ......................................................................................................................................

.................................................................................................................. Postcode: ............................

My bank sort code: □□ □□ □□ □□ My bank account number: ............................................................

Please pay to Children’s Liver Disease Foundation, account no. 00181442, sort code: 12-05-65

Starting on ...... /...... /...... until further notice. My signature: ..............................................................

Are you a UK taxpayer? Yes / No  If yes, please give your gift under Gift Aid.

This means that CLDF can claim the basic rate of tax you have already paid on the amount you are donating. It will not cost you anything.

In order to qualify you must have paid enough UK income or capital gains tax to cover all your charitable donations. Other taxes such as Council Tax or VAT do not apply. We will confirm all Gift Aid details in your acknowledgement letter.

□ Yes, please treat this and any future donations as given under Gift Aid. Date: ..............

About you:

First name: ............................ Surname: ..........................Title: Mr / Mrs / Ms / Miss / ..........

My address is: ............................................................................................................................................

.................................................................................................................. Postcode: ............................

Home telephone: ............................ Mobile: ...............................................................

Home email: ............................ Work email: ...............................................................

To claim Gift Aid we are required to have your full name and address including postcode.

Please return your completed form to CLDF, address below. Thank you.

Children’s Liver Disease Foundation, 36 Great Charles Street, Birmingham B3 3JY
Children’s Liver Disease Foundation is the UK’s leading organisation dedicated to taking action against the effects of childhood liver disease.

It provides free of charge:

- A huge selection of literature and online animations on the working of the liver available in print and online
- Information packs for a wide range of audiences, including young people, parents/carers, GP practices, schools and nurseries, friends and relatives
- Families and young people’s teams providing services in person, online, facebook, text and phone
- Developing services for adults diagnosed with a liver disease in childhood
- Website – childliverdisease.org
- Young people’s website – cldf-focus.org
- National event programme for families and young people to meet, share and have fun
- Secure online message board – childliverdisease.org/forum

Around 75% of CLDF’s annual income is derived from voluntary donations. Please help us to continue to support young people, families and adults diagnosed in childhood by making a donation. You can do this online or by completing the donation form in this leaflet. Even better, a regular direct debit gift will enable us to plan our work more fully.

Thank you.

Children’s Liver Disease Foundation
36 Great Charles Street
Birmingham
B3 3JY

0121 212 3839

info@childliverdisease.org

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Registered charity number 1067331