Welcome

This leaflet has been written for:

■ Parents/carers of a child/young person diagnosed with cystic fibrosis who have developed problems with their liver
■ Young people diagnosed with cystic fibrosis who have developed problems with their liver

Others who may find this leaflet helpful are:

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

This leaflet aims to:

■ Explain what happens to the liver
■ Explain tests and treatment

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

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

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 new born babies

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

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

Some children and young people with cystic fibrosis may have had prolonged jaundice as a baby. This usually disappears a few months after birth. However, a number of children and young people with cystic fibrosis develop liver problems later in life regardless of whether they had prolonged jaundice as a baby.

Liver disease is very variable and progresses at different rates. Each child or young person therefore needs individual assessment and follow-up.

Which children or young people with cystic fibrosis get liver disease?

It is not possible to predict who will be affected. Boys seem to be affected more than girls.

The development of liver disease may be more likely in children/young people who have had “meconium ileus” as a baby.

What happens to the liver in cystic fibrosis?

Damage to the liver starts in the small bile ducts. The bile produced by the liver cells into these ducts becomes stickier than normal and blockage of the ducts follows (this very occasionally causes jaundice). The surrounding liver tissue then becomes damaged and scarred, which may be called biliary fibrosis. This is not unlike the underlying process occurring in the lungs. In time the fibrosis progresses and eventually spreads throughout the liver. This causes the liver to become hard and the blood flow through it more difficult.

What tests are used to diagnose a liver problem?

Most children and young people with cystic fibrosis have regular check-ups which include:

1. Physical Examination

   Signs of liver disease may be found during a routine examination. These include:
   - Enlargement of the liver (hepatomegaly).
   - Enlargement of the spleen (splenomegaly) or both (hepatosplenomegaly).
   - Jaundice is very rare and is usually only seen when liver disease is very advanced.
   - Visible veins on the abdominal wall which suggests poor blood flow through the liver (portal hypertension). CLDF has a leaflet on portal hypertension.

2. Liver Function Tests (Blood Tests)

   Blood tests can be useful in monitoring the liver function but they may remain normal despite progression of liver disease.

3. Abdominal Ultrasound

   This shows the size and texture of the liver, the blood flow through it and any obvious abnormalities of the bile ducts including swelling and the presence of stones or blockages. It also enables accurate measurement of the size of the spleen and gall bladder.
If a liver problem is suspected following the above tests then occasionally the doctor may recommend a liver biopsy. This procedure, performed under anaesthetic, involves removing a small piece of liver with a special needle. This may help to show the extent of the damage to the liver. CLDF has a leaflet on routine investigations for liver disease.

What are the main effects of liver disease in children with cystic fibrosis?

1. **Portal Hypertension**

The hardening of the liver makes it more difficult for the blood to flow through it. Some of the blood supply may re-route through other blood vessels outside the liver, these are in the intestine, through the spleen and in the abdominal wall. The increased flow of blood through these vessels and spleen causes them to swell, and the enlarged vessels within parts of the intestine are called “varices”. This situation is called portal hypertension. Varices lie just under the lining of the oesophagus (gullet) and/or stomach, and their walls are thin and can bleed easily. This may cause vomiting of blood which needs urgent medical attention. Blood which has passed through the intestines turns black and the stools may turn black in colour. It is vital that a child is taken immediately to hospital for treatment if he or she vomits blood or passes black stools. Avoidance of medicines that may cause bleeding such as aspirin and brufen is very important.

CLDF has a leaflet on portal hypertension and oesophageal varices which is available on request.

2. **Enlarged spleen**

Portal hypertension also causes enlargement of the spleen. As the spleen slowly enlarges it traps and destroys platelets making them unavailable for their role in blood clotting — this is called hypersplenism. A low platelet count can be identified on a routine blood test. Easy bruising and prolonged bleeding can occur and may show for example as nosebleeds or heavy periods. In most circumstances, however, the blood can clot adequately despite relatively low numbers of platelets.

3. **Disturbance of the normal working of the liver**

If the liver becomes severely damaged its ability to perform its essential functions is impaired. The ways in which this affects the child may include:

- Reduced absorption of dietary fat soluble vitamins A, D, E and K.
- Excessive fluid may collect in the abdomen (called ascites) and, less often, in the legs (oedema) — due to impaired production of albumin and/or as a complication of portal hypertension.

CLDF has leaflets on ascites and portal hypertension which are available on request.

- An increased tendency to bruise/bleed due to impaired production of clotting factors by the liver.
- Poor growth. Liver disease can make the reduced absorption of food and poor appetite in children with cystic fibrosis worse. Extra nutrients are often required in the diet and advice is normally given by a dietitian. Despite such intervention, children can become underweight, lack energy and have poor growth.
What treatments may be considered?

1. Medication
   - Supplements of fat soluble vitamins may be required and in most cases are essential.
   - Ursodeoxycholic acid (Urso)
     Urso occurs naturally in small amounts in the bile. Increasing the amount of Urso in the bile by taking it as a medicine helps to make the bile less sticky, enabling it to flow more easily from the liver. As a result, the liver damage may be reduced. It is a relatively new medicine. An improvement in abnormal liver function tests has been shown but research continues as to whether the deterioration of the liver is prevented.

2. Nutrition
   Calorie and protein requirements are high and supplementary feeding is often indicated. Occasionally feeding directly into the stomach using a nasogastric tube is necessary. A dietitian will give advice.

3. Diagnosis and Treatment of Varices
   - Endoscopy
     An endoscope is a flexible tube which allows the medical team to look directly at the oesophagus and the stomach. An endoscopy will be performed if portal hypertension is suspected and can be performed at the same time as a liver biopsy.
   - Sclerotherapy
     This is the name given to the injection treatment of oesophageal varices. This is normally done by using a special attachment on the endoscope and is therefore often done at the same time as an endoscopy. This treatment may need to be repeated several times (see CLDF portal hypertension leaflet for details).
   - Banding
     This is another form of treatment to reduce bleeding from varices by placing a tight band around them to make them shrivel up.
   - TIPS
     This is a radiological procedure which reduces blood flow through the varices by directing it through a channel created in the liver.
   - Medication
     Ranitidine and/or sucralfate may be prescribed to reduce gastric irritation.

4. Liver Transplantation
   Liver transplantation can be an effective treatment for children with cystic fibrosis who have severe liver disease. A full assessment of nutrition, lungs and liver disease and associated complications is needed to decide whether the liver alone needs to be replaced, or the liver and the lungs. With careful assessment, preparation and excellent support, transplantation is a procedure that can offer great improvement in quality of life for many years.

Remember!

Only a small proportion of children with cystic fibrosis develop advanced liver disease that will cause major problems. The aim is to detect those at risk at an early stage and try to prevent the progression of liver damage. CLDF has a leaflet on routine investigations for liver disease, available on request.
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

© Children’s Liver Disease Foundation: April 2002
Updated: November 2005; April 2006; August 2008; February 2011; April 2012; August 2013; June 2014
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.
I confirm that I have paid or will pay an amount of Income tax and/or Capital Gains Tax for each tax year (6 April to 5 April) that is at least equal to the amount of tax that all the charities or CASCs that I donate to will reclaim on my gifts for that year.

I understand that: the charity will reclaim 25p of tax on every £1 that I give, I may cancel this declaration at any time, and I must inform CLDF if I change my name and/or address whilst this declaration is in force. All gift aid details will be confirmed 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