



Children's Liver  
Disease Foundation

fighting childhood  
liver disease

# Baby Jaundice and Liver Disease

a guide

Medical Information Series



# Welcome

This leaflet has been written for:

- Parents/carers of a baby being investigated because they have high levels of conjugated bilirubin (conjugated Hyperbilirubinaemia) in their blood and do not have signs of acute liver failure

Others who may find this leaflet helpful are:

- Relatives and friends

This leaflet aims to:

- Explain the discussions you are likely to have with professionals
- Explain what is likely to happen next
- Outline possible treatments

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

- Jaundice in the Newborn Baby (part of CLDF's Yellow Alert campaign)
- Guide to the liver

Children's Liver Disease Foundation has a website specifically for jaundice in the new born baby. For more information go to [yellowalert.org](http://yellowalert.org).

Children's Liver Disease Foundation (CLDF) also has leaflets in its support series which are available to download from our website – [childliverdisease.org](http://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.

## Glossary for this leaflet

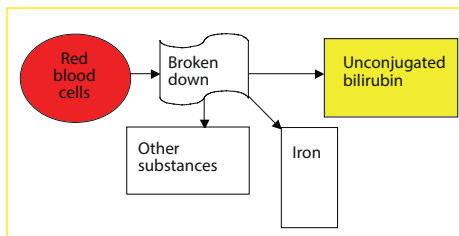
<b>Bile</b>	A yellow/green liquid which is excreted from the liver, stored in the gall bladder and passes into the duodenum to help the digestion of food by breaking down fat.
<b>Jaundice</b>	A condition where the eyes and/or skin become yellow due to an abnormally high level of bilirubin in the blood.
<b>Bilirubin</b>	This is the breakdown product from old red blood cells. The life of a red blood cell is normally 4 months in adults. In babies it is 2 — 3 weeks. <b>Unconjugated bilirubin</b> is the first type of bilirubin produced which is changed into <b>conjugated bilirubin</b> in the liver and excreted in the bile.
<b>Hyperbilirubinaemia</b>	Abnormally high concentrations of bilirubin in the bloodstream, resulting in jaundice.

### ? Jaundice is common so why is it a concern for my baby?

- There are two types of bilirubin, **unconjugated** and **conjugated**, both of which can cause jaundice.
- A raised level of **unconjugated** bilirubin occurs in the relatively common, physiological jaundice seen in many babies in the first two weeks of life especially if they are breast-fed and/or premature. See the CLDF leaflet “Jaundice in the newborn baby”.
- Blood tests already done have shown your baby has a high level and also a significant percentage of **conjugated** bilirubin compared to the total bilirubin. This indicates there is a problem in the liver.
- In some cases the problem will clear itself but in other cases specific treatment is needed to prevent further problems. It is necessary to do a number of special tests to find out which babies need treatment.

### ? How is unconjugated bilirubin produced?

- Unconjugated bilirubin is sometimes called indirect bilirubin.
- Unconjugated bilirubin is produced when red blood cells break down — it is yellow in colour.



**Diagram 1: The breakdown of old red blood cells produces unconjugated bilirubin, iron and other substances**

- This production line is happening in all of us, all of the time, as old red blood cells are broken down. New red blood cells are constantly being produced in the bone marrow, even in young babies.

- Babies are born with a higher number of red blood cells than they need so this ‘breakdown’ is faster in the first two weeks of life.
- Some abnormalities affecting the blood can also cause a more rapid breakdown of red blood cells and thus a raised level of unconjugated bilirubin.
- It is very rare for a raised level of **unconjugated bilirubin** to be caused by a problem in the liver unless there is an inherited problem with the way the liver processes or **conjugates** the bilirubin.

### ? How is unconjugated bilirubin changed to conjugated bilirubin?

- Conjugated bilirubin is sometimes called direct bilirubin.
- Unconjugated bilirubin is carried in the blood stream around the body and eventually to the liver.
- When the unconjugated bilirubin reaches the liver it is ‘processed’ by having a sugar attached to it, this changes its form and it is now called ‘conjugated’ bilirubin. It is still yellow in colour.
- The liver is often not completely mature at the time of birth, even if the baby is full-term, so it may not be able to process the unconjugated bilirubin as quickly as

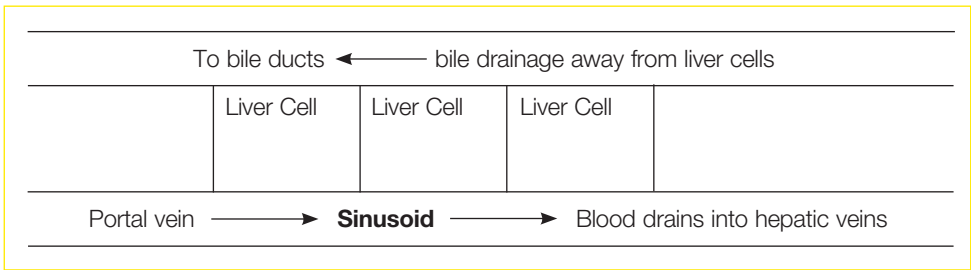
it is made. This can lead to a build-up of unconjugated bilirubin in the blood which passes around the body and makes the baby appear jaundiced (yellow). This explains why so many babies are jaundiced in the first two weeks of life but the jaundice usually clears fairly quickly. This is called **physiological jaundice**.

- An enzyme in breast milk can also slow the rate at which unconjugated bilirubin is converted into the conjugated form which explains why jaundice is more common in breast-fed babies and conjugated jaundice may not be considered.

### ? How is jaundice due to raised levels of conjugated bilirubin different?

- Conjugated bilirubin is not broken down by phototherapy (special light treatment which is often used to treat babies who have the common physiological — unconjugated jaundice).
- The different form of the conjugated bilirubin means it is unable to pass into the brain therefore it CANNOT cause brain irritation or damage which can be caused by very high levels of unconjugated bilirubin.

Diagram 2: Diagrammatic representation of liver cells



- Conjugated bilirubin (unlike unconjugated) is soluble in water, therefore when there are high levels in the blood some is passed out with water from the kidneys making the urine (wee) yellow in colour. The urine of a baby less than three months old should be completely colourless — like water.

## ? What happens in the liver?

In the liver, each of the cells is supplied by:

- branches of the hepatic artery bringing blood carrying oxygen, which is important to keep the cells healthy and able to function
- branches of the portal vein, bringing blood containing nutrients from the digestion of food and drink in the intestines (see diagram 2).

Numerous complex processes take place in every liver cell many of which are organised according to the instructions (blueprint) inherited in the genes from each parent. One of these processes is the conjugation of bilirubin; a sugar is attached to the unconjugated bilirubin to convert it into conjugated bilirubin. Other processes are responsible for the metabolism (changing) of the nutrients we take in as food and drink to a form which the body can use for growth, repair and energy. These are known as metabolic processes or metabolism.

Substances which are made or changed within the liver cells are taken away by:

- branches of the hepatic veins which return blood to the heart
- branches of the bile duct system which remove water, conjugated bilirubin, bile acids, cholesterol and other substances which together make up the bile (see glossary and later in this leaflet for more information about bile).

Some substances pass easily through the walls of the cell, blood vessels and bile ducts and others need the help of special 'pumps'.

- All blood leaves the liver cells via tiny branches of the hepatic vein which gradually combine to eventually form the three 'hepatic' veins which take blood from the liver back to the heart then pumped to the rest of the body.
- Bile leaves the liver cells in tiny bile ducts which gradually combine together to eventually form the two main 'branches' of the biliary tree — the left and right hepatic ducts. These two ducts combine to form the 'trunk' of the biliary 'tree' outside the liver.
- Bile is then diverted into the gall bladder and stored there; some of the water content is reabsorbed. When we eat, the gall bladder automatically contracts, squeezing bile back into the main bile duct and down into the first part of the intestine (bowel) known as the duodenum (see diagram 3).

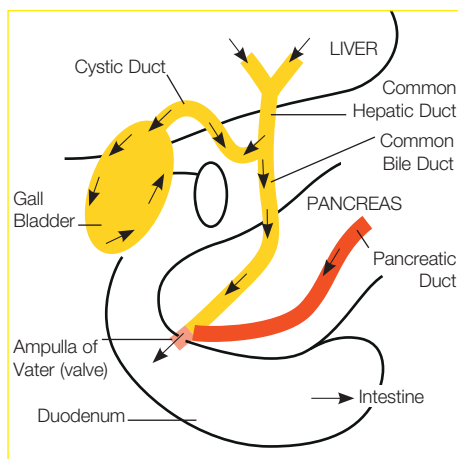


Diagram 3: The Bile Duct System or 'Biliary Tree'

## ? What is bile and why is it important?

Bile is a yellow/green bitter tasting liquid which is made in the liver. It contains:

- Bile acids
- Conjugated bilirubin
- Cholesterol
- Other lipids (fats)
- Water
- Other waste products

And small amounts of other substances.

Bile acts like the detergent in a washing up bowl breaking down the fat globules. It plays an important part in breaking down the fat in the food we eat into very small particles, so that we can digest it, use the energy in it and absorb the fat-soluble vitamins it contains.

Further down the intestine, in the section known as the terminal ileum, most of the bile acids are reabsorbed through the bowel wall into the blood stream and return to the liver to be reused.

Bile is responsible for colouring the stool and making it less smelly.

## ? What happens when there is a hold-up of bile flow (called cholestasis)?

When bile flow is reduced:

- The conjugated bilirubin level in the blood is raised; the total bilirubin should be less than 20µmol/l.
- The baby is likely to be jaundiced; this is most noticeable as yellow colouring of the whites of the eyes. The yellowing of the skin colour is more noticeable in babies who have naturally pale skin colour.

- Some conjugated bilirubin is passed out in the urine making it more coloured.
- A baby's urine should be completely colourless like water for the first few months.
- The stools (poo) may appear more bulky and/or greasy because some of the fat passes through the intestines without being completely digested.
- Stools may be paler in colour because there is less bile to colour them.
- Increased fats in the stool (because the lack of bile means fat cannot be digested) causing your baby to be very hungry and want more milk than normal.
- As the baby gets older they may find it difficult to gain the usual amount of weight.
- The liver may be larger than usual due to inflammation and swelling; the trapped bile irritates the liver.
- Vitamin levels may be low (especially the fat-soluble vitamins A, D, E and K) — due to reduced absorption of these vitamins, babies quickly use up the supply stored in the liver before birth.

Vitamin K is especially important as low levels can reduce the ability of the blood to clot effectively. Signs of affected blood clotting may include bruising (usually very rare in the first few months), prolonged bleeding from the umbilical cord stump (belly button) and nose bleeds. Blood tests including INR and/or Prothrombin time are used to measure blood clotting. See the CLDF leaflet "Routine Investigations".

## ? How will my baby's jaundice be investigated?

Some babies will be seen at their local hospital; others may be referred to a specialist centre.

The team will try to find the reason for the high level of conjugated bilirubin and advise what, if any, special treatment is required. Trying to find the answer is like doing a jigsaw



puzzle and each test puts a piece in the final picture. It is also most helpful for the team to speak with the parent(s) about the pregnancy and the family history. One particular test or piece of information may not give a clear answer on its own. The result may indicate which further tests are needed, eliminate some possibilities or just help to build the overall picture. There is a further CLDF leaflet "Routine investigations". The investigations may include the following:

- An examination by the doctor
- Blood tests
- Urine tests
- Stool examination/tests
- Ultrasound scan
- Other specialised scans
- Liver biopsy
- X-rays
- Eye test
- Heart (cardiac) test
- Bone marrow test

In some liver related conditions, there may be significant signs in other parts of the body so it is common for the examination and tests to cover all parts of the body.

## ? How will my baby be fed?

While tests are being carried out you are likely to be advised about the best feeding regime for your baby. One or more of the following changes may be advised:

- **Temporarily stopping breastfeeding** (if given) while some specific metabolic conditions are ruled out. If your baby has one of these conditions continuing to give breast milk could cause further damage to the liver. You will be advised about expressing your milk to maintain your supply as these conditions are very rare and your baby is most likely to be able to restart at least partial breastfeeding.

- **Giving a special milk formula.** If your baby's weight gain is not as good as it should be your baby will be given a formula containing a special type of fat called medium chain triglycerides; these are easier to digest when bile flow is poor. CLDF has a leaflet on this called "MCT Feeds. What are they?".
- **Naso-gastric tube feeding** — partial or complete feeding by a fine tube placed through your baby's nostril and down into the stomach may be required if they are unable to take the amount or type of feed they need by mouth. CLDF has a leaflet on this called "An introduction — Tube feeding".
- If a specific cause is found for the jaundice the feeds given may be changed again.

## ? What medicines will my baby be given?

Your baby may be given medicines to help to reduce the effects of the poor bile flow, these may include:

- **Vitamins** — these are less well absorbed from the milk when bile flow is poor. Vitamin K is especially important as the liver needs a good supply in order to make the proteins which control the ability of the blood to clot.
- **Ursodeoxycholic Acid (Urso)** — a medicine that can improve bile flow in some cases.

If a specific cause is found for the jaundice additional medications may be needed.

## ? What are the common causes of conjugated jaundice (hyperbilirubinaemia)?

There are many different reasons for this but they broadly fall into the following groups:

1. **Bile duct (drainage) problems** — the bile ducts are abnormal in some way. This may affect the bile ducts inside the liver, outside the liver or both. One condition which it is important to consider quickly is biliary atresia. This condition is rare but does require early surgery to obtain the best outcome. In biliary atresia some unknown process affects the bile ducts inside and outside the liver and the bile ducts outside the liver collapse or disappear causing complete obstruction to bile flow. CLDF has a leaflet on biliary atresia.
2. **Metabolic conditions** — an abnormality of one of the complex processes that occur within each liver cell. These include:
  - a) abnormalities in the way the cells in the liver make the bile or pass it out into the bile ducts;
  - b) abnormalities in the way in which the liver cells metabolise (change) the nutrients in the milk into a form which the body can use.
3. **Unknown** — despite the techniques available to modern medicine there are still many cases of high conjugated bilirubin (up to a third of all babies tested) where no clear explanation is found. This is more common in those babies who have had other medical problems soon after birth.

## ? What is the treatment for conjugated jaundice?

1. Special feeds and medication, as above, to assist with the effects of poor bile flow until this has completely resolved.
2. Specific treatment relating to the cause if available, this may include:
  - a) Special diet
  - b) Medication
  - c) Operation

## ? Is it serious?

The severity of the condition depends on the cause. The conjugated bilirubin on its own is not serious. If no underlying cause is identified the majority of children make good progress gradually clearing the jaundice over weeks or sometimes months and having no long-term ill effect from it. There are however a small percentage of children who continue to have ongoing liver problems and require further treatment — for this reason all children with high levels of conjugated bilirubin must be followed up.



## ? 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](http://childliverdisease.org)

Young people's site: [cldf-focus.org](http://cldf-focus.org)

[info@childliverdisease.org](mailto: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](mailto:families@childliverdisease.org)

0121 212 6023

## Young People's Team

[youngpeople@childliverdisease.org](mailto:youngpeople@childliverdisease.org)

0121 212 6023

**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](http://childliverdisease.org)
- Young people's website – [cldf-focus.org](http://cldf-focus.org)
- National event programme for families and young people to meet, share and have fun
- Secure online message board – [childliverdisease.org/forum](http://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.**

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