Children's Liver Disease Foundation

fighting childhood liver disease

Baby Jaundice and Liver Disease

A Guide

An explanation of what baby jaundice is, information about further investigations and possible treatments

Baby Jaundice and Liver Disease

This information has primarily been written for:

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

Others may also find this information useful:

- Healthcare professionals who would like to find out more about liver disease in children.

It provides information on:

- What jaundice is
- The different types of bilirubin
- What happens in the liver
- Possible investigations and treatments

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

- Jaundice in the Newborn Baby
- Yellow Alert Stool Chart Bookmark
- Introduction to Liver Disease – explains the role of the liver, bile, the tests used to diagnose and monitor liver disease, treatments and a glossary of liver terms
- CLDF Essentials Pack

What is jaundice?

Jaundice is caused by a build-up of bilirubin. Bilirubin is produced in all healthy children and adults and is formed when red blood cells are broken down. As old red blood cells are broken down, they are replaced with new cells produced in the bone marrow. Jaundice normally disappears by the time your baby is 10-14 days old. This may take a bit longer if the baby is premature, in which case, it can take about three weeks to go away.

Why is jaundice a concern for my baby?

In most cases jaundice clears up on its own. However, sometimes jaundice can be a sign that there is a problem with a baby's liver.

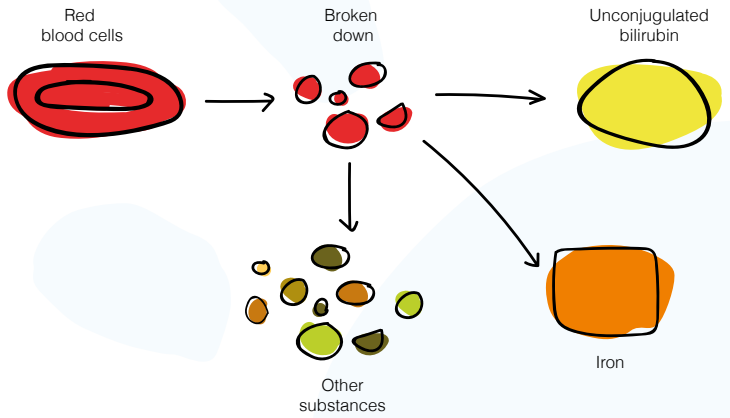
There are two types of bilirubin which can cause jaundice:

- Unconjugated bilirubin
  A raised level of unconjugated bilirubin is relatively common. Babies with physiological jaundice in the first two weeks of life often have high levels of this type of bilirubin. It is especially common in breast-fed and/or premature children.
- Conjugated bilirubin
  Your baby has had some blood tests. These have shown that there is a higher level of conjugated bilirubin than is normally expected in comparison to the level of unconjugated bilirubin. This suggests that there may be a problem with your child's 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 carry out a number of tests to find out what is causing your baby's jaundice and what treatment may be needed.

How is unconjugated bilirubin produced?

Unconjugated bilirubin is produced when red blood cells are broken down in the body — it is yellow in colour.



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 leading to 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 bilirubin.

How is unconjugated bilirubin changed to conjugated 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.

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

How is conjugated bilirubin different?

Conjugated bilirubin is not broken down by phototherapy. This is a special light treatment often used to treat babies who have common physiological (unconjugated) jaundice.

Conjugated bilirubin is unable to pass into the brain and therefore cannot cause brain irritation or damage which can be caused by very high levels of unconjugated bilirubin.

Conjugated bilirubin is soluble in water, therefore when there are high levels in the blood some is passed out with water from the kidneys. This makes a baby's 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?

Each of the cells in the liver is supplied with blood by:

- branches of the hepatic artery, bringing blood from the heart carrying oxygen. This 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.

Numerous complex processes take place in every liver cell.

One of these processes is the conjugation of bilirubin. Other processes are responsible for changing 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 made or changed within the liver cells are removed by:

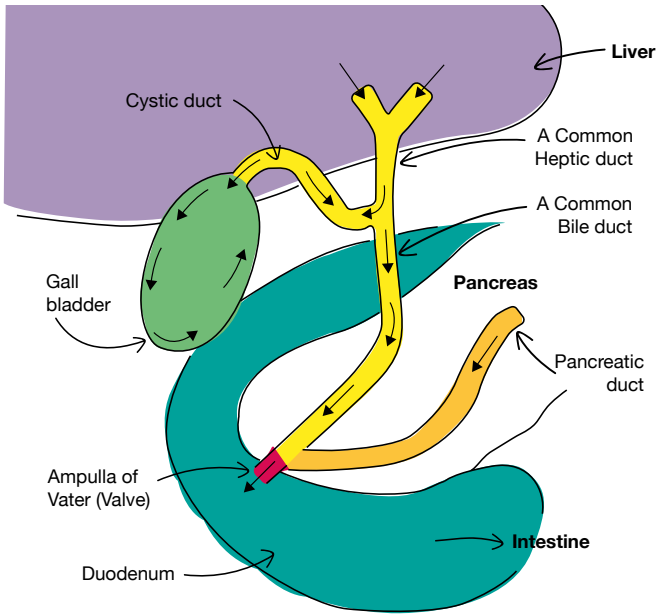
- branches of the hepatic veins which return blood to the heart
- branches of the bile duct system which remove water, conjugated bilirubin, bile acids, cholesterol and other substances. Together these make up the bile.

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

All blood leaves the liver cells via tiny branches of the hepatic vein which gradually combine to form three hepatic veins. These take blood from the liver back to the heart, which then pumps it to the rest of the body.

Bile leaves the liver cells via tiny bile ducts which gradually combine to form the two main branches of the biliary tree — the left and right hepatic ducts. These two ducts join to form the trunk of the biliary tree outside the liver.

Bile is then diverted into the gall bladder where it is stored. 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 (duodenum).



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, 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 terminal ileum, most of the bile acids are reabsorbed through the bowel wall into the blood stream and returned to the liver to be reused.

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

What happens when there is a reduction in bile flow (cholestasis)?

When the flow of bile is reduced:

- The conjugated bilirubin level in the blood is raised; 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.
- Stools 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 may cause 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 as 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 (international normalised ratio) and/or prothrombin time are used to measure blood clotting. See the CLDF leaflet "Introduction to Liver Disease" for further information.

How will my baby's jaundice be investigated?

If your baby has high levels of conjugated bilirubin they may be seen at their local hospital or referred to a specialist centre.

The liver 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 an answer is like doing a jigsaw puzzle and each test adds a piece to the final picture. It is also helpful for the team to speak with the parent(s) about the pregnancy and family history. One particular test may not give a clear answer on its own. See the CLDF leaflet "Introduction to Liver Disease" for further information.

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?

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

- **Temporarily stopping breast feeding** 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 breast feeding.
- **Feeding with 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 type of fat called medium chain triglycerides; these are easier to digest when bile flow is poor.
- **Feeding via a naso-gastric tube.** Your baby may be partially or completely fed by a fine tube placed through their nostril and down into their stomach.

These feeds may be changed if a specific cause is found for the jaundice.

See the CLDF leaflet "Nutrition" for further information.

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 (A, D, E and K)–these are less well absorbed from the milk when bile flow is poor.
- 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 causes conjugated jaundice (conjugated hyperbilirubinaemia)?

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

- **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.

- **Metabolic conditions** — an abnormality of one of the complex processes that occur within each liver cell.

  These include:

  abnormalities in the way the cells in the liver make the bile or pass it out into the bile ducts

  abnormalities in the way in which the liver cells metabolise (change) the nutrients in the milk into a form which the body can use

How is conjugated jaundice treated?

- Special feeds and medicines can help to treat the effects of poor bile flow until this has completely resolved.
- Specific treatment relating to the cause if available. This may include a special diet, medicines or an operation.

Is my baby's jaundice serious?

This depends on the cause of the jaundice. Conjugated bilirubin on its own is not serious. If no underlying cause is identified the majority of babies make good progress and gradually clear their jaundice within weeks or sometimes months with no long-term ill effect.

However, a small percentage of children may 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 by a paediatrician.

CLDF produces a wide variety of information resources for children and young people up to the age of 25 with liver disease, their families and the healthcare professionals who look after them. This information can be downloaded or ordered from CLDF's website childliverdisease.org. For further enquiries regarding CLDF's information please contact the Information and Research Hub Manager by email at irhm@childliverdisease.org or call 0121 212 6029.

Thanks

This booklet has been written, edited and reviewed with the help of staff at each of the specialist paediatric liver centres: Birmingham Women's and Children's NHS Foundation Trust, King's College Hospital and Leeds Children's Hospital. Thank you to all the staff involved who have made the production of this leaflet possible.

Disclaimer

This leaflet provides general information but does not replace medical advice. It is important to contact your/your child's medical team if you have any worries or concerns.

Feedback and information sources

Information within this leaflet has been produced with input from the three specialist paediatric liver centres in the UK. To provide feedback on this leaflet, or for more information on the content of this leaflet including references and how it was developed contact Children's Liver Disease Foundation: info@childliverdisease.org.

This leaflet has been reviewed in March 2019. It is due to be reviewed by March 2022.

What is Children's Liver Disease Foundation (CLDF)?

CLDF is the UK's leading organisation dedicated to fighting all childhood liver diseases. CLDF provides information and support services to young people up to the age of 25 with liver conditions and their families, funds vital research into childhood liver disease and is a voice for everyone affected.

Are you a young person up to the age of 25 with a liver condition or a family member? CLDF's Families 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.

If you are a parent/carer or family member then get in touch with CLDF's Families Team:
Phone: 0121 212 6023 **Email:** families@childliverdisease.org

If you are a young person and want to find out more about CLDF's services you can contact CLDF's Young People's Team:
Phone: 0121 212 6024 **Email:** youngpeople@childliverdisease.org

CLDF have a social media platform called HIVE for young people with a liver disease/transplant to make new friends, connect and share stories **childliverdisease.org/young-people/hive**

Would you like to help us support the fight against childhood liver disease? All of CLDF's work is funded entirely through voluntary donations and fundraising. Please help us to continue to support young people, families and adults diagnosed in childhood now and in the future. To find out more about fundraising and how you can join the fight against childhood liver disease you can visit childliverdisease.org/get-involved. Alternatively you can email the Fundraising Team at fundraising@childliverdisease.org or call them on **0121 212 6022**

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