LIVER UK

The new name for the British Liver Trust and Children's Liver Disease Foundation

Biliary atresia

Help, support and guidance for children and families

Liver UK would like to thank Mirum Pharmaceuticals for their kind financial support towards the production of this information.

Mirum Pharmaceuticals have had no editorial involvement.

Contents

This leaflet contains a lot of information.

Don't feel you need to read it all at once.

You may want to read just the key facts and come back to the full content later.

The Liver UK Children and Families team are also here to provide support, advice and information.

Welcome

Welcome to your guide to understanding biliary atresia. It provides information on causes, diagnosis, symptoms, treatment and living with biliary atresia.

This information has been written for:
Parents and carers of babies, children and young people with biliary atresia

Others who may also find this information useful:

- young people with biliary atresia
- adults with biliary atresia
- healthcare professionals

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

- Introduction to liver disease
- Nutrition
- Pruritus
- Portal hypertension and ascites
- Liver transplantationn

Use this QR code to view our resources or visit **liveruk.org/biliary-atresia**

How to say it:
Biliary atresia
BILL-ee-air-ee
a-TREE-zee-uh

Useful words
Stool = poo / faeces
Urine = wee

Key facts about biliary atresia

1. Biliary atresia is a rare disease of the liver and bile ducts. It presents very early in life and requires urgent treatment.

2. Biliary atresia happens in around 1 in 10,000 to 1 in 20,000 births in the UK.

3. There are several different types of biliary atresia.

4. We still don't know exactly why biliary atresia happens, but doctors think it could be caused by a mixture of things.

5. Babies with biliary atresia usually appear well and healthy when they are first born. But signs usually start to appear during the first few weeks of life.

6. Tests for biliary atresia include liver blood tests, an ultrasound scan and sometimes a liver biopsy.

7. A surgical operation called a Kasai procedure can create a new path for bile to flow from the liver. This is best done in the first 2 to 3 months of life.

8. If a baby does not have a Kasai procedure in the first 2 to 3 months of life, the success rate of the operation is very low.

9. Complications following a Kasai procedure may include cholangitis, itching, ongoing jaundice and poor growth. Sadly, in some children, liver disease can get worse and may lead to more severe complications and liver failure.

10. More babies with biliary atresia are growing to reach adulthood without needing a liver transplant. But it is likely that they will need a liver transplant at some point in their adult life.

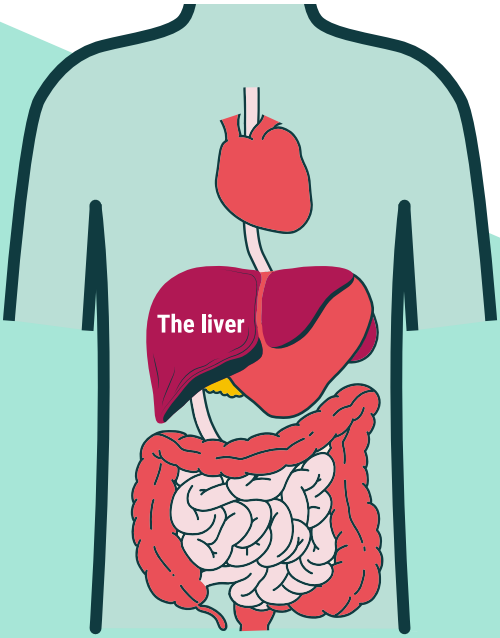
About the liver

The liver is one of the most important organs in the body. It is on the right side of the tummy, just under the ribs. It works like the body's factory and carries out hundreds of important jobs every day.

The liver is a dark reddish-brown colour.

It is divided into two main sections called "lobes". There is a right lobe and left lobe. The right lobe is a bit bigger.

The liver is joined up to some important arteries and veins that bring blood into and out of the liver.



The liver has over 500 jobs. These include:

- digesting food
- producing and storing energy
- storing vitamins, iron and other nutrients
- fighting infections
- breaking down toxins and other harmful substances
- helping you heal

About biliary atresia

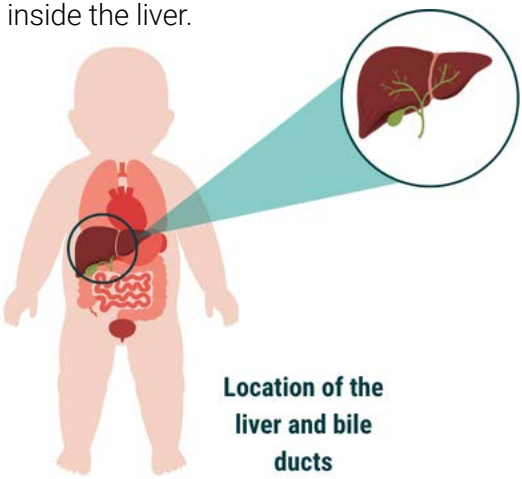
This section explains what it is, what causes it and how it may affect your child.

What is biliary atresia?

Biliary atresia is a rare disease of the liver and bile ducts. It is a serious condition that a baby is born with. It therefore presents very early in life and requires urgent treatment.

In newborns with biliary atresia, bile ducts are missing, incomplete, blocked or damaged. This stops the flow of bile from the liver to the gallbladder and small intestine. Bile backs up and gets trapped inside the liver. This is known as cholestasis.

It may cause serious damage to the liver as cells die and are replaced with scar tissue (fibrosis).



Location of the liver and bile ducts

Bile ducts are tiny tubes that connect different organs, including the liver and gallbladder. They are part of the digestive system. Bile ducts carry bile from the liver to the gallbladder and small intestine.

Bile is a green/yellow liquid that is made in the liver and stored in the gallbladder. It helps the body:

- **digest food by breaking down fats**
- **absorb vitamins A, D, E & K**
- **get rid of waste products such as bilirubin and leftover cholesterol**

Babies with biliary atresia may also be born with other problems affecting the heart, blood vessels, spleen, intestines and kidneys. This is called **biliary atresia splenic malformation syndrome (BASM)**.

Biliary atresia happens in around 1 in 10,000 to 1 in 20,000 births in the UK.

Types of biliary atresia

There are 3 different types of biliary atresia. The type is based on the location of the blockage and the amount of damage to the bile ducts. In all types, bile ducts inside the liver are damaged.

Type I:

This type happens in around 5-10% of newborns with biliary atresia. In this type, the blockage happens in the common bile duct, but some of the bile ducts near the liver may stay open. Bile is usually found in the gallbladder.

Type II:

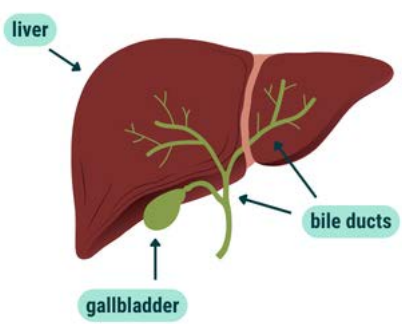
This type is rarer and happens in around 1-2% of newborns with biliary atresia. In this type, the blockage happens in the common hepatic duct, but some of the bile ducts near the liver may stay open.

Type III:

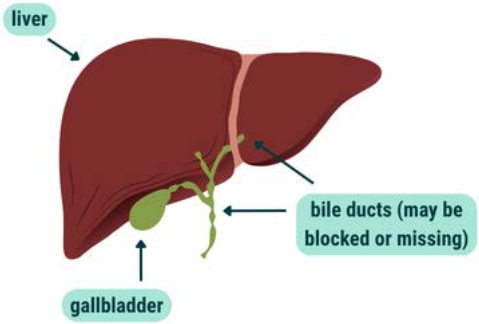
This is the most common type of biliary atresia and happens in more than 90% of newborns with the condition. You may also hear it called extrahepatic biliary atresia. In type III, all the bile ducts outside the liver are missing, blocked or damaged. This causes a total and permanent blockage of bile flow.

There are other sub-types of biliary atresia. This includes biliary atresia splenic malformation syndrome (BASM).

The treatment for all types is the same, but the results are slightly better if your baby has type I or type II biliary atresia.



Normal liver and bile ducts



Bile ducts in biliary atresia

"We had never heard of this disease and in all honesty we were really quite terrified. I made the mistake of googling things I didn't fully understand … and truly expected the worst" – Parent

What causes biliary atresia?

We still don't know exactly why biliary atresia happens. But lots of research has been carried out and doctors think it could be caused by a mixture of things, including:

- genes
- toxins in the environment
- viral infections
- an immune system response
- developmental issues

We know that parents often feel guilty that they may have, in some way, contributed to their child having biliary atresia. But there is no way that the condition could have been prevented. There is no evidence to suggest that biliary atresia is hereditary (can be passed on to children by their parents) or due to anything parents might have done. Biliary atresia does not run in families and children with biliary atresia will not pass the condition on to their own children later in life.

What are the signs of biliary atresia?

Babies with biliary atresia usually appear well and healthy when they are first born. They often feed well and show no signs of the condition in the first days of life.

But signs usually start to appear during the first few weeks. In some babies the signs can be very clear. In others they can be much less obvious. This can make a diagnosis difficult for healthcare professionals.

The key signs of biliary atresia are:

Prolonged jaundice

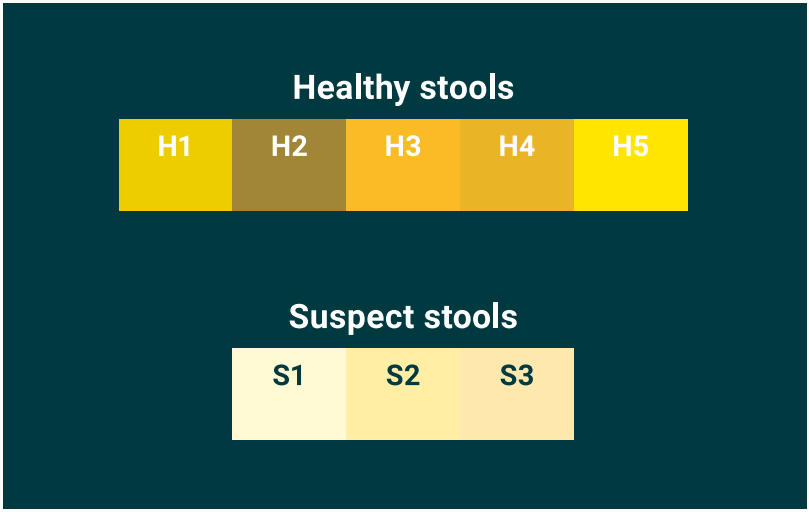
Jaundice is when the whites of the eyes and the skin turn yellow. It happens when bilirubin trapped in the liver passes back into the bloodstream. Jaundice is very common in newborn babies and usually clears within the first 2 weeks of life. **But if jaundice does not go away by 2 weeks of age (3 weeks in a preterm baby), this could be a sign of biliary atresia.** This is called prolonged jaundice.

Bilirubin is a natural waste product in the body. It is made when old red blood cells break down in the spleen. Bilirubin is yellow, which gives stools and blood their colour. Bilirubin travels in the blood stream.

Pale stools (poo)

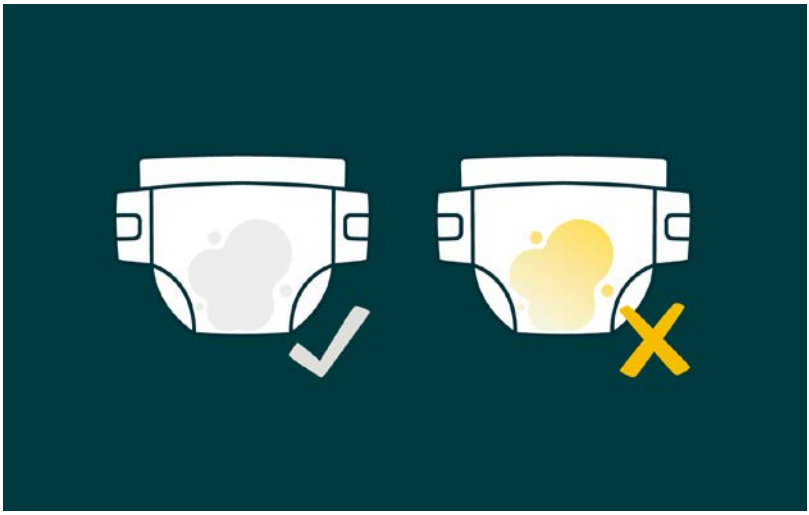
The stool of a baby should be a yellow or mustard colour. If your baby's stool is **pale, grey, white or clay** in colour, this could be a sign of biliary atresia. It happens when there is no bile being emptied into the intestine. Bilirubin in bile gives normal stools their colour. If there is less bile flow, this can result in pale stools.

See our stool chart which shows the colours of healthy and potentially unhealthy stools.



Dark urine

A baby's urine should be colourless. But if your baby is jaundiced and their urine is persistently **yellow or dark** in colour this could be a sign of biliary atresia. This happens when a type of bilirubin called conjugated bilirubin passes out in the urine and makes it look darker than normal.



Other signs

Other signs of biliary atresia may appear in some babies and include:

- feeding more often (your baby may seem very hungry)
- poor weight gain
- enlarged liver
- enlarged spleen
- trouble with bleeding or clotting (coagulopathy) – this will be more likely in babies who have not been given the routine vitamin K injection at birth

Some signs may only appear in babies where an early diagnosis is missed and a few more weeks have passed.

How is biliary atresia diagnosed?

Biliary atresia is diagnosed very early in life. If your baby has any signs of the condition, they will go to hospital for tests to help confirm the diagnosis. These tests are completed over the course of a few days, so your baby may be admitted to a ward.

Tests may include:

Liver blood tests

(also known as liver function tests/LFTs)

A blood sample will be taken and tested in a laboratory. This is a common way to check if the liver is damaged and how well it works.

Stool and urine tests

Your baby's stool will be checked to look for a pale, grey, white or clay colour. A sample of urine will also be taken and tested in a laboratory.

Ultrasound scans

Ultrasound scans use sound waves to make pictures of the inside of the body. An ultrasound scan has no side effects and won't be painful for your baby. These scans are used to check and monitor:

- the size and texture of your baby's liver
- blood flow to and from your baby's liver
- your baby's gallbladder and bile ducts
- the size of your baby's spleen

Liver biopsy

A liver biopsy may be needed to confirm the diagnosis. This is a short procedure that will be done under general anaesthetic. During a liver biopsy a very thin needle goes through the tummy wall and into the liver. The needle collects a small sample of liver tissue. This is sent to a laboratory to be studied under a microscope.

How can biliary atresia be treated?

There is no cure for biliary atresia and it cannot be treated with medicines.

If tests show that your baby is very likely to have biliary atresia, they will need surgery as soon as possible. The operation needed for biliary atresia is called a **Kasai procedure**. This is named after the Japanese surgeon who first performed the operation in the 1950s.

Needing surgery so early in life can be a big shock for families. But it is very important that the operation is done as early as possible, because this will give your child the best chance of a good outcome. Remember – your hospital team and the Liver UK Children and Families team are here to support you every step of the way.

Your baby will be treated at one of 3 paediatric liver centres in the UK:

- King's College Hospital, London
- Birmingham Women's and Children's Hospital
- Leeds General Infirmary

Use this QR code to view our resources or visit **liveruk.org/biliary-atresia**

"Although it was extremely overwhelming, the team (at the hospital) did everything they could to put our worries at ease, my son's health and happiness was always a priority and I felt confident his care was in the best hands" - Parent

Before the Kasai procedure

Blood tests will be done and will include checking your baby's blood group in case they need blood before, during or after surgery. Your baby may be given special medicines to prepare their gut for surgery.

Your baby will not be fed milk for a number of hours before the operation. The team caring for your baby will explain what this involves. If you are breast feeding and want to express and store milk, speak to your nursing team.

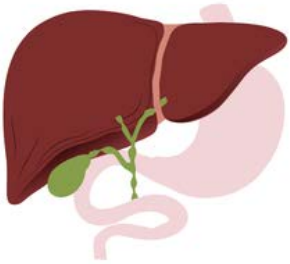
You will be allowed to go down to the operating theatre with your baby and stay until just before your baby is asleep.

During the Kasai procedure

The medical team will start the operation by carrying out an investigation called an intraoperative cholangiogram. This is done under general anaesthetic when your child has been put to sleep and will confirm the diagnosis of biliary atresia.

A cholangiogram is an X-ray that uses a special dye to show the bile ducts. It checks for blockages in bile ducts outside the liver.

If biliary atresia is confirmed, the surgical team will continue the operation with the Kasai procedure (also known as a Kasai portoenterostomy). This is usually done under the same general anaesthetic at the same time. The aim of the operation is to help restore bile flow by making a drainage tube that allows bile to drain from the liver into the gut.



Before surgery

After surgery

During the operation, the gallbladder and all the damaged bile ducts outside the liver are removed. The surgeon finds smaller bile ducts along the surface of the liver that are still open and draining bile. They attach a loop of the baby's own intestine to this part of the liver. This allows bile to flow from the remaining healthy bile ducts found inside the liver into the intestine.

During the operation, the surgeon will also check the liver for damage or other problems. They will take a tiny piece of the liver to send to the laboratory for further tests. This is called a wedge biopsy.

The Kasai procedure will last a whole morning or afternoon. Putting your baby to sleep, giving your child pain management and then waiting for your baby to wake up will take longer.

"I reached out to other families who had been through similar experiences. Hearing their stories helped me feel less alone and gave me hope during a very challenging time" - Parent

After the Kasai procedure

You will be able to see your baby after surgery in the recovery area. There will be a tube going into your baby's nose and down into their stomach. This is called a nasogastric tube. This keeps your baby's stomach empty, which helps prevent sickness. It is important to keep your baby's stomach empty for 48-72 hours after surgery.

You will not be able to feed your baby for a couple of days. This is because the surgeon operates on the bowel, which stops it from working for a short time. When the bowel recovers, you can start feeding again. Until this time, your baby will be fed using an intravenous drip. This is where fluids and nutrients are put straight into your baby's body through a vein.

It can be very emotional to see your baby after surgery. There will be lots of tubes and machines and this can feel very overwhelming. The medical team will explain what is happening and will answer any questions you may have. It will also be possible for you to hold your baby the day following the surgery.

Some commonly used devices you will see after surgery include:

Central line

A long, thin, hollow tube called a central line will be placed into one of the bigger blood vessels in your baby's neck during surgery. This will be used to give your baby fluids, medicines, antibiotics and blood products if needed.

Urinary catheter

Your baby will have a urinary catheter in place, which drains urine from the bladder. This helps the medical team measure the amount of urine your baby is making. It will tell them how well their kidneys are working.

Abdominal drain

Your baby may have an abdominal drain put in place during surgery. This drains excess wound fluid, blood or bile from around the liver. The drain will be removed as the amount of liquid reduces. A dressing will cover the wound on your baby's tummy for the next few days. The stitches will dissolve after the wound has healed.

Medicines

Medicines are used after the Kasai procedure to help with bile flow and to manage symptoms. The medicines used will be different for each baby and will depend on their condition and progress. The medical team will help you understand which medicines your baby needs to take and when.

Some commonly used medicines are:

Pain relieving medicines

Your baby will be given pain relief after surgery to keep them comfortable. Medicines are given via the central line or via a cannula (a tube inserted into a vein).

Sometimes pain relief is given through a small tube inserted into your baby's back at the time of the operation. This is similar to those used for women in labour. It is called an "epidural catheter".

Antibiotics

Antibiotics are given after the Kasai procedure to reduce the risk of infection in the bile ducts (cholangitis). Your baby will be given antibiotics through a vein for 3-5 days after surgery. Further antibiotics may be given if needed.

Phenobarbital

This medicine may be used to help increase the flow of bile. It might make your baby sleepy at first, so it is usually given as a single dose in the evening.

URSO (Ursodeoxycholic acid)

URSO is a bile salt found naturally in small amounts in bile and helps to increase the flow of bile. Your baby may start taking this medicine before the Kasai procedure. Or they may be given URSO when they start feeding again after surgery.

Steroids

Steroid medicines may be given for 4-6 weeks after surgery. They help to reduce inflammation and promote bile flow.

Vitamins

Poor bile flow can make it difficult for your baby to absorb vitamins from their feeds. They will be given vitamin supplements to help with this. This may be before or after the Kasai procedure.

Spironolactone

This medicine will be used if your baby needs help to get rid of extra fluid that has collected in their abdomen (ascites).

Anti-viral therapy

When doing tests for biliary atresia, the medical team sometimes find a virus called cytomegalovirus (CMV). This happens in around 10% of newborns with biliary atresia. The medical team may need to treat the virus with medicines that are put straight into your baby's body through a vein. This will be a course of treatment lasting around 4-6 weeks. Your baby may have blood tests to check that the virus has cleared once they are home.

Other medicines may be prescribed in certain circumstances. Your hospital team will explain why these medicines are needed.

"He had the Kasai procedure at eight weeks and has been doing brilliantly ever since. He always surprises the doctors and consultants when we have our check-ups as he is such a strong, vibrant and healthy boy who is much bigger than most of his friends at nursery!" - Parent

Are there any other problems linked with biliary atresia?

Some babies with biliary atresia have additional problems linked with the condition. They may include:

Problems with their spleen

Some babies with biliary atresia have problems with their spleen. They may have no spleen (asplenia) or multiple small spleens (polysplenia). This can affect the way the immune system works to fight off infections.

> The spleen is a small organ which sits in the upper left abdomen next to the stomach. It helps fight infection and filter the blood.

Problems with their heart

Some babies with biliary atresia have problems with their heart. We don't know the exact number of babies affected, but it is thought to be around 7% of babies with biliary atresia in the UK.

Having heart problems alongside biliary atresia can make the condition more serious for your baby. The medical team will do extra tests, such as heart scans to diagnose any problems. Your baby will be referred to the cardiology team to explain the findings and what treatment is needed. If your baby needs surgery to treat heart problems, this may be done before the Kasai procedure, where possible, as it may help improve your baby's outcome.

Problems with their blood vessels

Some babies with biliary atresia have problems with their blood vessels, particularly those around the liver and spleen. This can make the Kasai procedure more difficult for the surgeon.

Problems with their intestines and digestive system

Babies with biliary atresia may have problems with the intestines or other parts of the digestive system. Some problems can be corrected at the same time as the Kasai procedure.

If some or all of these complications occur, it is a condition known as biliary atresia splenic malformation syndrome (BASM). This condition does not affect how a baby responds to the Kasai procedure. However, the link with heart problems can affect the outcome of surgery.

Babies with BASM may need additional tests. They may also need to take daily antibiotics to reduce their risk of infection.

What happens when my baby leaves hospital?

If there are no problems after the Kasai procedure, your baby will be allowed to go home around 7-10 days after the operation. This will happen once you and the medical team are happy with your baby's condition.

Before going home, the medical team will make sure that your baby's wound is healing and there is no suggestion of infection anywhere in their body. You will be given or sent an outpatient appointment, plus a supply of medicines and feeds.

Your baby will need life-long follow-up by doctors, specialist nurses and dietitians. The hospital staff at the liver centre will advise your health visitor, GP and local hospital about your baby's condition and any special care they may need. This means they can offer you support when you are at home. Your Clinical Nurse Specialist (CNS) will stay in touch with you, and you will be able to ring them for advice or reassurance whenever you are worried, so you are not alone.

When you get home you should try and treat your baby as normally as possible. We know it can be hard settling into everyday life after such a difficult experience, and you will be worried at times. This is completely normal. But remember that babies with biliary atresia get common illnesses just like other babies. There is also help and support available.

Remember – your hospital team and the Liver UK Children and Families team are here to support you every step of the way.

Use this QR code to view our support services or visit **liveruk.org/support**

Medicines at home

Your baby will leave hospital with a supply of medicines. After this, more medicines will be arranged through your GP. It is important to note that some medicines are not available straight away from pharmacies. You may need to order them several days in advance so you do not run out.

As with all babies, it is important that your child gets all their usual immunisations at the correct times. But sometimes your baby's vaccinations will need to be delayed. Your medical team will advise you when immunisations can be given.

Remember: DO NOT give your child any medicines containing aspirin or ibuprofen. If your child needs medicine to reduce a high temperature, please use paracetamol (e.g. Panadol/ Calpol). Speak to your medical team if you are unsure.

Feeding and nutrition

If your baby was being breast fed before you found out they had biliary atresia, it may be possible to start breast feeding again. It is important that your milk has been maintained by expressing milk while your baby was unable to feed.

Most babies with biliary atresia are unable to gain enough weight on breast milk or standard formula alone. Your baby may also need a special milk formula suitable for babies with liver disease. These milks contain fats which are more easily digested and absorbed. Formulas with high calories may also be used to help your baby gain enough weight. It is often helpful for your baby to be weighed on a weekly basis to check how much weight they gain. A dietitian will be available to support you with feeding options.

Your doctor will also prescribe extra vitamins for your baby. This is because babies with biliary atresia have problems absorbing vitamins A, D, E and K.

Use this QR code to view our leaflet on nutrition or visit **liveruk.org/nutrition-children**

"He now has an NG feeding tube to help him gain some weight. I was initially upset at the thought of it but having reached out to other BA parents, I was assured that this would benefit him and they were right – it's been fantastic!" - Parent

Symptoms to look out for

When your baby leaves hospital, you will have regular follow-ups. There will also be support from your GP and health visiting team. But it is important to seek further advice from your GP or hospital if your baby shows any of the following symptoms:

- a raised temperature of 37.8ºC or more
- an unknown illness with a raised temperature
- jaundice with pale stools and dark urine
- your baby is generally unwell

If you have any questions, do not hesitate to contact your Clinical Nurse Specialist at the liver unit treating your baby.

How do I know if the Kasai procedure has been successful?

At the time of the operation the surgeon will not be able to tell if the Kasai procedure has been successful. It usually takes around 3 to 6 months to know if bile is draining well from the liver to the intestines.

The success of the Kasai procedure will also depend on:

- the age that your baby had the Kasai procedure
- the amount of scarring and damage that happened to your baby's liver before the operation

After surgery, the medical team will monitor your baby's stools to see if the colour changes from pale/grey/white/ to a yellow/ mustard colour. They will also monitor your baby's urine to see if it becomes clearer and less yellow. If your baby is jaundiced, this should gradually fade in the 2 to 3 months after surgery. Blood tests will hopefully show that bilirubin levels drop towards normal levels.

What happens if biliary atresia is not treated or treatment is unsuccessful?

If a baby does not have a Kasai procedure in the first 2 to 3 months of life, the success rate of the operation is very low.

Sadly, a small number of babies are diagnosed late, and a Kasai procedure is not possible. Bile produced in the liver builds up, causing too much damage and scarring in the liver. At this stage, a liver transplant is the only treatment available. In the UK, less than 5% of babies with biliary atresia go straight to liver transplantation.

Even if a Kasai procedure happens in the first few months of life, it is not always successful. In some babies, bile does not drain successfully, and damage keeps happening to the liver. With no other treatment available to stop this ongoing damage to the liver, around 40% of babies and children will need a liver transplant before 2 years of age.

It is important to remember that the Kasai procedure is not a cure for biliary atresia. If successful, it allows some children to live for many years, sometimes decades, with their own liver. But most children will still develop complications that need to be managed. Sometimes, these complications can be severe and may make a liver transplant necessary. Most children will still need a liver transplant at some point in their life.

Just over half of children with biliary atresia will need a liver transplant within 10 years

Around 60% of children and young people with biliary atresia will need a liver transplant by 20 years of age

What other complications might occur?

Complications can happen in any baby with biliary atresia, even if they have a Kasai procedure and clear their jaundice completely.

Complications can often be managed using medicines and other treatments. But sometimes the complications become very severe, even life-threatening, despite medical treatment. At this point, the medical team may consider a liver transplant.

Cholangitis

Cholangitis is an infection of the bile ducts. It is a common complication after the Kasai procedure and needs urgent treatment. Cholangitis most often happens in the first year or two after the Kasai procedure, but it can happen at any time. It can also happen more than once or keep returning.

The symptoms of cholangitis include fever and jaundice. If your child has a temperature over 37.8ºC or an unknown illness, contact your child's medical team. Cholangitis can be treated with antibiotics, but they can only be given at your local hospital or the liver unit treating your child. Antibiotics will be given intravenously. This is where the medicine is put straight into your baby's body through a vein. It is often followed by a long course of oral antibiotics (antibiotics given by mouth).

Intrahepatic biliary cysts (bile lakes)

Intrahepatic biliary cysts, or "bile lakes" are pools of bile that form in the bile ducts. They can block the bile ducts and increase the risk of cholangitis. Bile lakes are fairly common in biliary atresia, with around 1 in 4 children developing at least one bile lake. If the lakes are large or cause problems, they can be drained.

Itching (pruritus)

Some children with biliary atresia experience itching of the skin. The level of itch varies from child to child. It can range from being mild to causing severe discomfort. If you think your child may be affected, talk to your GP or liver unit. They may be able to offer medicines to help with this symptom. The Liver UK Children and Families Team also has lots of advice for managing itching.

Use this QR code to view our leaflet on pruritus or visit **liveruk.org/pruritus**

Vitamin deficiencies

Babies and children with biliary atresia often have trouble absorbing vitamins A, D, E and K, even after a successful Kasai procedure. They will be monitored for vitamin deficiencies and given extra vitamins if needed.

Faltering growth

Many babies and children with biliary atresia struggle to gain weight, even after a successful Kasai procedure. They may grow more slowly than other children and may need extra nutritional support. A dietitian will be available to support you with feeding options.

Portal hypertension and variceal bleeding

Portal hypertension is high pressure in the portal vein. This is the main vein carrying blood from the gut to the liver. When the liver is damaged, it becomes stiff. This stiffness makes it harder for blood to flow through the liver. This causes the high pressure in the portal vein.

Portal hypertension can make the spleen get bigger (splenomegaly). It can also cause swollen blood vessels in the food pipe (oesophageal varices). The swollen blood vessels have thin walls and may bleed. This may cause your child to vomit blood or pass black tarry stools. Both symptoms need immediate medical attention. It is uncommon for this to happen before a child is 2 years old.

If there are no symptoms or complications from portal hypertension, then treatment may not be needed. But some children will need treatments such as banding or sclerotherapy to the oesophageal varices.

Fluid in the tummy (ascites)

Ascites is the term for a build-up of fluid in the tummy. A small amount of ascites can be present for up to 6 weeks after the Kasai procedure but should then disappear. If it comes back again and your child's tummy becomes noticeably bigger, or they have unusual weight gain, contact your medical team.

Treatment for ascites may be started by your GP or local paediatrician and may include medicines or changing your child's diet. In more serious cases, doctors may need to admit your child to hospital for treatment.

Use this QR code to view our leaflet on portal hypertension or visit **liveruk.org/portal-hypertension-children**

Hepatopulmonary syndrome

This is an uncommon complication that affects the ability of the lungs to exchange oxygen. It happens in around 5–7% of children with biliary atresia. Symptoms include shortness of breath and low blood oxygen levels.

Portopulmonary hypertension

This is a rare complication and happens in less than 3% of children and young people with biliary atresia. It usually starts in adolescence and is more common in girls. Symptoms include shortness of breath and tiredness.

Liver cancer

Liver cancer can develop in anyone with a badly scarred liver, but it is generally very rare in babies and children. It has been reported in less than 2% of babies and children after a Kasai procedure. Every person with biliary atresia will have routine screening for liver cancer using blood tests and ultrasound scans. If picked up early a liver transplantation will be considered.

What will happen in the future?

Every child with biliary atresia is different, so we don't always know what will happen in the future.

As your child grows, they will be closely monitored with regular clinic appointments, blood tests and scans. How often they need to go to hospital will vary for each child.

"Our family life has been challenging, particularly for my two older children. My husband took extended leave from work to care for us, and my parents, family and in-laws provided essential support with our other children. There were difficult moments, but with video calls and hospital visits, we managed to stay connected. Now, our children are happy, and our family is slowly getting back to normal" - Parent

If your child reaches adulthood with their own liver, it is likely that they will develop complications at some point. Existing complications can also become more difficult to manage. It is highly likely that they will need a liver transplant at some point in their adult life.

Remember - Liver UK is here to support your child through their transition to adult services and throughout their adult life.

If your child reaches adulthood after receiving a liver transplant as a child, they will need to take medicines and have regular check-ups to keep their new liver healthy.

It is important to remember that most children with biliary atresia reach adulthood and have a good quality of life. This may be with their own liver following a Kasai procedure or following a liver transplant. They move (transition) to adult liver services and keep having regular check-ups, blood tests and scans throughout their lives.

“I know a lot of families will be heartbroken to hear that their child may need a liver transplant or that a Kasai procedure may fail, as it did for us. But this does not mean it is the end of their child’s life. We welcomed the idea of a transplant because we knew it would change his life for the better. Families should know that children can go on to live very healthy, normal lives, grow up, and even have families of their own” - Parent

Liver UK is in touch with a large number of families who have children with biliary atresia. These children live happy, fulfilling lives. Biliary atresia doesn’t have to hold a child back from living a relatively normal life.

Liver Transplant

Some babies and children with biliary atresia will need a liver transplant early in life. Others will keep their own liver for a lot longer following a Kasai procedure and may only need a transplant later in life.

A liver transplant is an operation to replace a damaged liver with a healthy one from a donor. Children may receive a whole, split or reduced liver from a deceased donor, or part of a liver from a living donor. It is also possible that a parent or family member could give part of their own liver. Ask your child's medical team for more information on this.

A successful liver transplant can save a child's life and will greatly improve their quality of life. But a liver transplant is a major operation and it will only be done after the benefits and risks have been carefully weighed up. After a successful transplant, children need life-long medicines and regular follow-up appointments. If liver transplantation is an option for your child, this will be discussed with you by your child's medical team.

The outlook for children who need a liver transplant in the UK is very encouraging. More than 9 out of 10 children are alive five years after their transplant, and over 8 out of 10 are still doing well twenty years later.

Use this QR code to view our leaflet on liver transplant leaflet or visit **liveruk.org/liver-transplant-children**

Louis was four days old when doctors diagnosed him with biliary atresia and he had the Kasai procedure eight days later. Since then he has joined a transplant football team, run the London Marathon and is training to be a teacher. Now, as he waits on the liver transplant list, Louis is pushing himself to stay as fit and healthy as possible.

Use this QR code to read Louis' full story or visit **liveruk.org/biliary-atresia**

Useful resources

Useful websites

Liver UK
www.liveruk.org.uk
Information and support for everyone living with a liver condition.

Genetic Alliance UK
www.geneticalliance.org.uk
A national charity providing support for people with genetic, rare and undiagnosed conditions.

Rare Disease UK
www.raredisease.org.uk
A national campaign for people living with rare conditions.

How Liver UK can help

A diagnosis of liver disease can be worrying, and you may have a lot of questions. The Liver UK support team are here for you and for your family and friends.

Whether you have questions or just need someone to listen, we can help.

Find out more about our support options: **liveruk.org/support**

Call our helpline **0800 652 7330**

How we produce information

Feedback and information services

Your feedback is important to us and will help us improve our information. To provide feedback, email **patient-info@liveruk.org**

For more information on how our information was developed, visit our website: **www.liveruk.org**

Trusted Information Creator
Patient Information Forum

We have gained PIF TICK accreditation for our information production process.

Thanks

All our information resources are reviewed by medical experts and people living with liver disease. We would like to thank:

Professor Mark Davenport, Consultant Paediatric Surgeon, King's College Hospital.

Professor Deirdre Kelly, Professor of Paediatric Hepatology

All our parent and family reviewers

Disclaimer

This resource provides general information but does not replace personal medical advice. It is important to contact your medical team if you have any worries or concerns.

Donate to Liver UK

We are entirely funded by donations.

To make a gift in support of our work today, please scan the QR code or visit

www.liveruk.org/donate

Liver UK is the UK's leading charity dedicated to supporting adults, children and young people affected by liver disease or liver cancer. Our mission is to transform liver health by raising awareness, promoting prevention, and improving care and support for everyone affected.

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