

Starship Children's Health

Paediatric Liver Disease

Parent Guide

Biliary Atresia



The
Kids
Foundation
key immune deficiency support


Starship
Children's Health

How do I contact the Clinical Nurse Specialist?

The Clinical Nurse Specialist team for Liver & Gastroenterology are often your first point of contact when you have questions about your child relating to their illness.

Our role is national so regardless of where you are living within New Zealand we may need to provide support or guidance from time to time. We work closely with the paediatric services (nursing and medical) in your home town. Most of our families have children with ongoing medical needs due to poor or deteriorating liver disease. Some have also had a liver transplant. We should be automatically notified of any child who has biliary atresia but this does not always occur.

When contacting the team we do ask that you consider if your question is urgent or non-urgent.

Because your calls are important to us, we have launched an on-call phone so that the CNS team can be contacted quickly as needed.

If Urgent contact:

CNS team during business hours 8- 4.30pm weekdays

By Phone: 021 837 870

PLEASE DO NOT TEXT MESSAGE US AS WE HAVE NO WAY OF KNOWING YOU HAVE TRIED TO CONTACT US IF THE MESSAGE IS DELAYED OR NOT DELIVERED.

After hours and weekends: take your child to your GP clinic or nearest emergency department for assessment

If outside Auckland please first contact your local team for advice.

If Non urgent:

We can be emailed: StarshipLiverCNS@adhb.govt.nz

This email will be received by each CNS and replied to as appropriate.

If you require new PRESCRIPTIONS please contact your GP or our team support:

For questions relating to CLINIC VISITS please contact our clinic scheduler (see phone number on previous page)

What is Biliary Atresia?

Biliary atresia is a medical condition. Irritation and swelling called inflammation develops within the bile ducts around the time of birth. The inflammation can happen in any of the bile ducts inside or outside the liver. This causes bile duct damage. When ducts are damaged the bile cannot flow out of the liver. Bile is trapped in the liver and causes scarring of the liver. This scarring is called fibrosis. The bile ducts outside the liver are always permanently damaged preventing any bile flow into the gut. The gall bladder is also affected.

What causes Biliary Atresia?

The cause of biliary atresia is not known. Research is being carried out but it has not yet provided any answers.

Parents often worry or feel guilty. They sometimes believe that they may have caused their child to have this problem. These feelings are very normal but parents are not at fault! At present there is no known way of preventing or expecting the occurrence of biliary atresia.

What are the signs of Biliary Atresia?

In the first few weeks of life babies with biliary atresia often seem well, apart from yellowing of the skin and whites of the eyes. This colouring is called jaundice. Jaundice in babies is very common and mostly clears within the first two weeks of life. This is often called breast milk jaundice. Babies with this condition will be helped by placing the babies under lights or sunlight or helps the body to clear the jaundice. This type of jaundice does not re-occur. If the cause of jaundice is liver disease then frequently the jaundice does not fade.

Other important signs are

Dark yellow coloured urine - normally the urine of a newborn baby is colourless. If a baby's urine is persistently yellow this can be a sign of liver disease.

Pale stools - normally the stools of a baby are green or yellow. If the stools of a baby are grey, white, fawn or pale then this can be an indication of liver disease

Bleeding - bleeding which lasts longer than normal may be a sign of liver disease. Common places babies bleed from is the mouth or nose.

What is Biliary Atresia Splenic Malformation Syndrome - BASM?

In a small number of babies with biliary atresia other problems are found.

These problems might include:

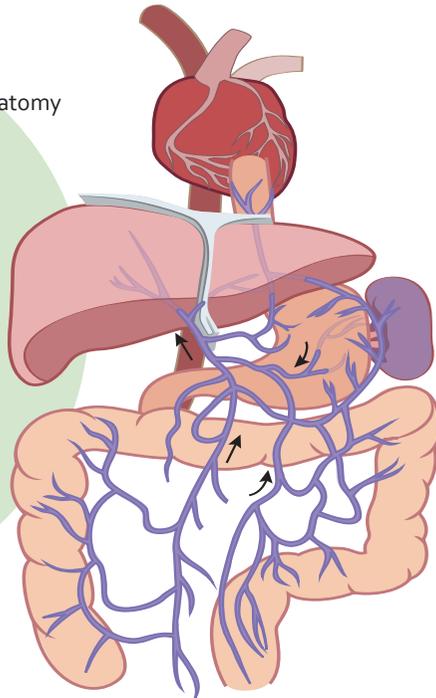
A spleen which is in many pieces (called polysplenia) or is missing (called asplenia). The spleen is an organ which sits under the lower ribs on the left side. It sits next to the stomach and helps the fight against infection. It also removes blood cells, especially platelets at the end of their life. It is normally a solid single organ.

An abnormal arrangement of the intestine (gut), known as intestinal malrotation.

Some abnormalities of the veins in the abdomen
Heart defects.

If your baby has some or all of these problems they have BASM. These children have less chance of clearing their jaundice even with the right treatment. Babies with BASM also need extra tests such as a heart scan (ECHO) to check for problems. Babies will usually have extra immunisations. They will also have to take special medicines called antibiotics. These are needed as the spleen does not work as well and babies have a higher risk of infection.

Normal anatomy



Important telephone numbers

Starship (Auckland City Hospital)	09 307 4949
Liver Nurse Specialist	Cate Fraser-Irwin & Karyn Sanson Oncall phone - held by the nurse responsible for outpatients 021837870
Gastroenterology Consultants	Care of Dani (Team Support) Extn 5471 Out of business hours please contact On-call Consultant via Auckland City Hospital switchboard 09 307 4949
Team Support	09 3074949 Extn 5471
Clinic bookings	Deborah Robb 09 307 4949 Extn 2150
Ward 26B Medical Specialties	09 307 4949 Extn 25760
Social Worker	09 307 4949 0212416268
Pharmacist	09 307 4949 Extn Pager No 935059 / 937471

When to contact the Starship team?

Return of pale stools and/ or dark urine.

Swollen abdomen

Excessive bruising

Nose bleeds lasting more than 5 mins

Blood in vomit or bowel motions



Immunisations:

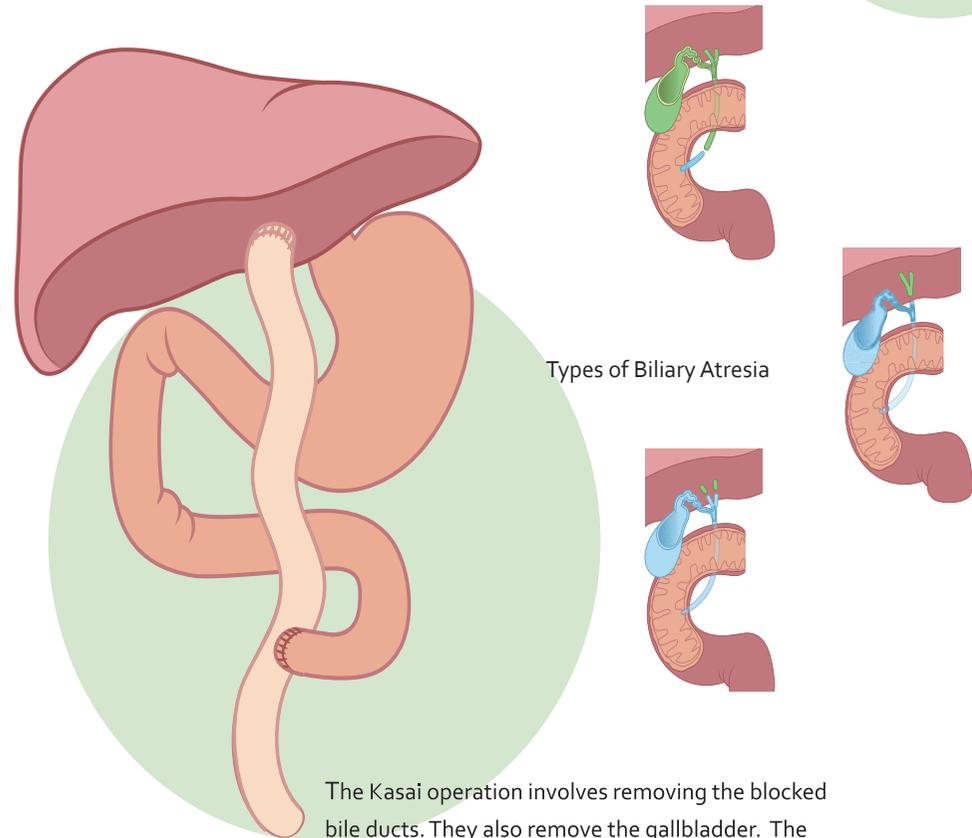
Children with liver disease need to be immunised.

We advise that your child receive all routine immunisations and these may be given earlier than the normal times. The team at Starship will discuss these with you as they are due. If you are worried about this it is important that you discuss it with the caring

What is the treatment for Biliary Atresia?

There are two types of bile ducts. Bile ducts inside the liver are called intrahepatic ducts. Outside the liver they are called extrahepatic ducts. In biliary atresia these ducts are not connected.

Initial treatment for biliary atresia is an operation called the 'Kasai operation'. The Kasai operation allows bile to drain from the liver into the bowel.



The Kasai operation involves removing the blocked bile ducts. They also remove the gallbladder. The surgeon brings a piece of bowel up and sews it to the liver. This piece of bowel acts like a new extra-hepatic duct system.

How do I know if the operation worked?

Almost 95% of patients with biliary atresia survive.

Some will clear their jaundice, have normal liver function and are unlikely to experience other problems.

Some children will continue to have liver function which is not quite normal. These children can may develop treatable problems such as portal hypertension or worsening liver function which may lead to liver transplant.

At the time of the operation the surgeon will not be able to tell if bile will start draining well.

Around 50 – 60% of Kasai procedures are successful.

The first stools after the operation are usually very dark, and then they will become paler.

If bile is draining well the stools will slowly change to a yellow/green or brown colour. Later you will see the urine becomes paler.

Stool colours change a little from nappy to nappy. This is normal for a child after kasai.

If bile drainage is working well the jaundice slowly fades. Blood tests show that the chemical bilirubin, which causes jaundice, drops towards a normal level. The operation may not drain bile from all sections of the liver so it is not unusual for liver function tests to remain higher than normal, even if the jaundice has gone

The difference maximising nutrition makes



Medications for itchy skin

If your child starts to become very itchy, please let the CNS or your doctor know. We will prescribe medicines to assist. Itch caused by liver disease is very difficult to treat.

Paracetamol (Panadol / Pamol)

Children with liver disease are allowed Paracetamol to help pain and fever. The dose needs to be lower than normal dose and worked out on the weight of the baby not age. Please ask the doctor what dose your baby should have.

PLEASE DO NOT GIVE: ibuprofen (e.g. Nurofen, I-profen) or medicines which contain Aspirin

If you want to give your child natural or alternative medicines – please discuss this with your child's doctor.



What medications will my baby be on?

Before discharge you will spend time with our pharmacist. You will be given a yellow medication card which we need you to bring to each clinic appointment.

Vitamins A, D, E
(Vitadol C, Micel E or Vit ABDeK)

When bile is not flowing freely it is more difficult for your baby to take in the fats from their food. There are some vitamins which need to be attached to the fat to be taken in.

These vitamins are important for such things as healthy skin, sight, bone development etc.

The liver plays a part in storing and managing these vitamins
When the liver becomes damaged the liver is unable to complete these important roles so your child is given extra vitamins to support the normal function of the liver

We measure the level of Vitamins A, D and E every 3 months. We need to make sure that your child is receiving the right dose for them.

Vitamin K
Vitamin K helps your baby's blood to clot –Vitamin K is the most important medication your child will be taking.
Vitamin K comes in glass tubes called vials. The medicine needs to be drawn up from the vial with a special syringe. Please ask your pharmacist to give you the 2mg/ml preparation so that you are given the right supplies.

Antibiotics
Children who have had a Kasai are given medicines called antibiotics. Antibiotics work to kill infections in our bodies caused by bugs called bacteria. These antibiotics may be given for up to a year or longer.
Antibiotics are given to your baby to help stop infections starting and this is called prophylaxis's. This dose will be lower than if we are treating an infection which has already started.

What treatment will my baby need after Kasai?

Children who have biliary atresia will often need special milk formula.
A person called a dietitian is an expert in helping babies grow. She will work with you to make sure your baby gets the right amount of the right feed to help them grow.

At the start we will try to keep your baby breast feeding if this is what you want. Unfortunately this is often not the right thing for your baby and it does not work. Children with biliary atresia are often very hungry. They may want to feed all hours of the day and night. This can be almost constant. Even when this is happening they will not manage to gain weight. It is impossible to keep up with the demands of your baby when they are feeding this often.

If your baby's liver is not draining properly they will not be able to take in and use the fats in breast milk. Normal baby formula won't work either.

Your baby will be given a special baby formula called Pepti Junior. Pepti junior has fats which are more easily taken in by babies with biliary atresia. Sometimes we need to add extra calories to the formula. This might be a liquid or a powder. These extras help your baby to grow without having to feed as often or drink large bottles. Sometimes we can do a mixture of breast and bottle feeding. The formula will be prescribed by your doctor and you will pick it up from the chemist just like other medicines.

Sometimes babies with biliary atresia need to be fed by a naso-gastric tube. This is a tube inserted through the nose into the stomach. It means you can feed the baby directly into the stomach. This may be because they refuse to drink the formula. It may also be because they are not able to take enough feed from the bottle on their own.

It is very important that babies with biliary atresia have the full amount the dietitian tells you they need every day. In the first months of life we need to see 20-30g weight gain every day.

Your baby will be seen often by the dietitian.

What will happen when I go home?

We realise that going home can be a little scary. Many parents worry that they won't notice if their baby is becoming sick. This is unlikely, as you are the ones who know your baby best!

A child with liver disease will need life long follow up to ensure that any changes in his/her liver disease is detected early.

Children with Liver Disease have some specific care requirements this information is available to you in this pamphlet.

You will receive a letter when you are discharged from the hospital. This includes a follow up plan. If you are not given this before you leave it will be posted within days – contact the CNS team if this does not happen.

This letter is also sent to your local General Practitioner (GP) and paediatrician.

We recommend that you take your child to your GP soon after getting home so they can get to know your baby better.

Your GP is still important in the care of your child. In most cases a specialist appointment is arranged within 1 – 2 weeks following discharge.

A referral will be made to either a community nurse or plunket nurse – they will weigh your child, and provide you with support.

Routine Follow up includes:

- Outpatient clinic with a team doctor
- Blood tests
- Nutritional assessment, by specialist dietitians.
- Developmental assessment and support is available when needed.
- Immunisation coordination.
- Ongoing emotional and practical support. This is through the social worker, and/ or team psychologist and Kids Foundation support worker.



How do I know if my baby is becoming unwell?

The following signs could mean that your child is becoming unwell, and they should be seen by your GP or a member of the hospital team.

Jaundice or Pale Stools

If jaundice gets darker or stools become paler this is a sign that things are not right. The easiest place to see jaundice is in the whites of your baby's eyes. Skin colour may vary due to different lighting.

High Temperature - 38 C or higher

Like any other growing child, your child will come into contact with childhood illnesses. Many of these cause fever. He or she will need to be seen by your GP for this. If your GP can't see the cause for the temperature such as ear, chest or throat infection then they need to contact the Starship team. It is possible that your child may have an infection of the bile ducts called cholangitis.

In this case your child will be admitted to hospital, blood tests will be taken.

After bloods are taken your baby will be started on intravenous antibiotics for up to 2 weeks. If cholangitis is left untreated it can cause further damage to the liver. Cholangitis is most likely in the first year or two after the Kasai procedure.

Poor Weight Gain and Growth:

It is important for children with liver disease to have good nutrition. We know that good nutrition can increase the chances of liver recovery after Kasai. We will measure your baby's length and weight at each clinic visit. Feeds may be changed or adjusted following discussion with the dietitian and Dr responsible for your child's care.

Itching

Some children with slow bile flow become very itchy. This can develop at any time and can make your child quite unsettled. Please let us know if this is happening.

There are a number of things we can do which may help.

