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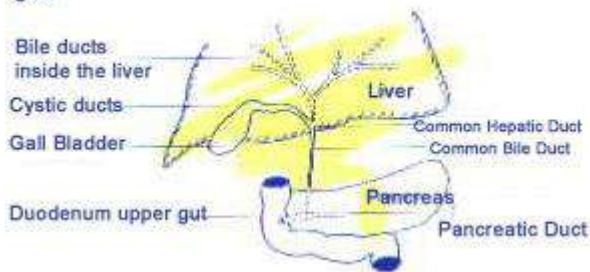
PATIENT INFORMATION ABOUT BILIARY ATRESIA

This information is designed to give a general overview of a rare condition termed as "Biliary Atresia" which affects infants.

Before reading this, it may be helpful to read about the liver and how it works from any medical physiology book. One important function of the liver is to make a substance called bile. Bile is a liquid, which passes into the gut through small tubes called bile ducts. Bile is necessary for the proper digestion of fat within the diet.

Diagram 1

Showing the normal liver, gall bladder, bile ducts and upper guts

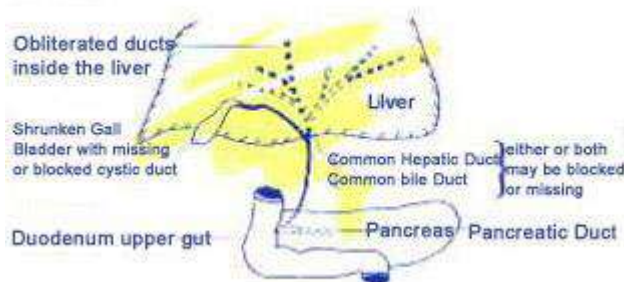


What is Biliary Atresia?

Biliary atresia is a condition in which inflammation develops within the bile ducts around the time of birth. The inflammation can occur in any of the bile ducts both inside and outside of the liver. This leads to bile duct damage and reduces the flow of bile which subsequently causes scarring (fibrosis or cirrhosis) of the liver. The bile ducts outside the liver are always irreversibly damaged preventing any bile flow into gut. The gall bladder is also affected.

Diagram 2

Showing the same structures in a child with biliary atresia



The darkened area represents the damaged bile ducts

What causes biliary atresia?

Unfortunately the cause of biliary atresia is not known. Research is being carried out but it has not yet provided any definitive answers. There are various theories attributing some viruses or autoimmune processes to cause the pathology. Biliary atresia, however is NOT a hereditary condition. Parents frequently feel guilty that they may in some way have contributed to their child having this problem. These feelings are groundless, as at present, there is no known way of preventing or anticipating the occurrence of biliary atresia.

What are the signs of biliary atresia?

In the first few weeks of life, babies with biliary atresia often seen well, apart from being jaundiced

(yellowing of the skin and whites of the eyes)

Jaundice in newborn babies is very common and mostly it clears within the first 2 weeks of life. This is caused due to the change over of the newborn baby's blood (hemoglobin) to the variety that is present in adults. However, if the cause of the jaundice is due to a Liver Disease, then, frequently the jaundice does not fade.

Other important signs are:

Yellow Coloured urine

Normally the urine of a newborn baby is colourless. If the baby's urine is persistently yellow or brown this can be an indication of Liver Disease.

Pale Stools

Normally the stools of a baby are green or yellow. If the stools are grey, white, fawn or pale then this can be an indication of Liver Disease.

Bleeding

Prolonged bleeding from the umbilicus or bleeding elsewhere (e.g. a nosebleed) may be an indication of Liver Disease.

How is biliary atresia diagnosed?

The signs that have been described are non-specific and can indicate other disorders, therefore there will be a need to number of investigations in order to make a diagnosis. The tests are to be carried out in hospital and include blood tests, scans and liver biopsy. Your baby will be admitted to hospital for a period of about a week for the tests.

Once other causes of Liver Disease have been excluded and investigations indicate biliary atresia your baby will be scheduled for surgery. The doctors and nurses will explain to you the type of surgery planned. The diagnosis and type of biliary atresia will only be confirmed at the time of operation.

How can biliary atresia be treated?

The initial treatment for biliary atresia is a surgical operation called Kasai procedure after the Japanese Surgeon who pioneered the operation. The aim of the Kasai procedure is to allow bile to drain from the liver into the gut. This is a major operation involving removal of the affected bile ducts that are outside the liver, and making an alternative system of drainage of bile from the liver into the intestines by making a "bile duct like tube" by restructuring the small intestines.

Babies with prolonged jaundice and suspected of having a Liver Disease such as biliary atresia, will often need special feeds. The hospital Dietitian will advise. There are a number of special milks available for babies suffering from Liver Disease. These milks have fats, which are more easily digested by babies with Liver Disease. Extra calories may also be added to the feeds in the form of powders to help your baby maintain an adequate weight gain. Most babies with biliary atresia are unable to gain weight satisfactorily on breast milk alone. In these cases the Dietitian will advise how to combine breast and bottle-feeding.

The doctor may prescribe extra vitamins as babies with prolonged jaundice also have problems absorbing certain vitamins.



What happens before the operation?

When any operation is required some preparation is necessary. Blood tests will be done so that your baby's blood group can be identified in case there is a need to give blood to your baby before, during or after surgery. Your baby may require special medicines to prepare the gut for surgery.

Your baby may also require some feeds to be replaced by special non-milk feeds and/or some fluids to be given into the vein by means of drip. The nurses on the ward will tell you what you should give and when

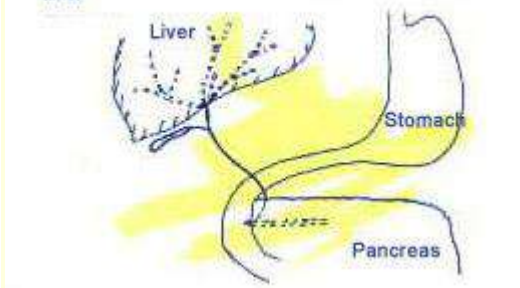
What happens during the operation?

The aim of the operation is to make a drainage channel to allow bile to drain from the liver. The exact way in which this is done will depend on the type of biliary atresia your baby has. During the operation the Surgeon will examine the liver, look for abnormalities and may take a liver biopsy (i.e. remove a tiny piece for further laboratory tests).

The operation usually takes usually takes a whole morning or afternoon. The surgeon takes as long as is needed to carry out what he / she needs to do.

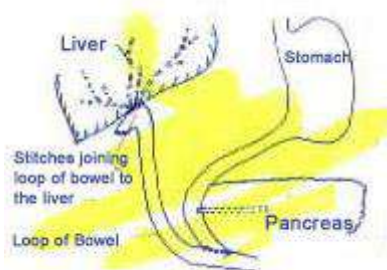
There are 2 types of operations.

A. The Surgeon finds no patent (open) duct outside the liver



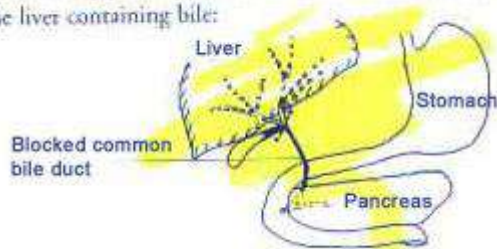
A. The surgeon finds no patent (open) ducts outside the liver.

This is the most common finding noted in 85% of cases of biliary atresia. The ducts are blocked (or missing altogether) right up to the liver surface. Under this circumstance, the blocked ducts are removed completely up to the liver surface. The surgeon cuts the bowel and also cuts into the under surface of the liver to find patent (open) bile ducts, i.e. microscopic bile ductules, which can drain bile effectively. The loop of bowel is then joined to this area of the liver.

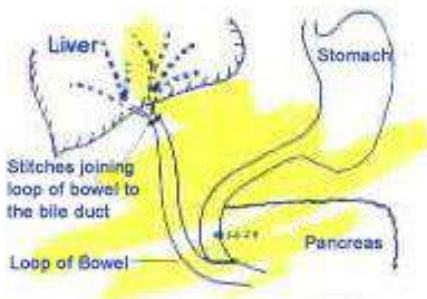


B. The surgeon finds patent bile ducts immediately below the liver containing bile.

B. The Surgeon finds patent bile ducts immediately below the liver containing bile:



In this type, the surgeon can join the loop of the bowel to the remaining bile ducts.



The surgeon will perform the operation, which is best for your baby. Experience has shown no noticeable difference in long-term success rates between the two operations.

What happens immediately after the Kasai operation?

Your baby will return to the ward, usually with Oxygen hood box over the upper part of your baby's body and head. This enables the nurse to control the amount of oxygen and water vapour in the air your baby breathes. This allows your baby to breathe more easily and to prevent chest infections. Sometimes the babies have to be kept in Pediatric Intensive Care Unit for a few days before shifting them to the ward.

There will be a tube running up your baby's nose and down into the stomach, this is called a nasogastric tube. This usually drains into a clear bag or small pot and keeps the stomach empty which helps to prevent vomiting.

There will also be an intravenous infusion or "drip" into a vein usually in the back of the baby's hand. This is used to give:

Fluids, which are given in order to maintain the correct sugar, salt and water level in the body. Daily blood tests will be carried out in order to check these levels so that they can be adjusted accordingly. All fluid required by your baby will be given through the drip until feeding is re-established.

Pain relieving drugs are often given through the drip or as suppositories.

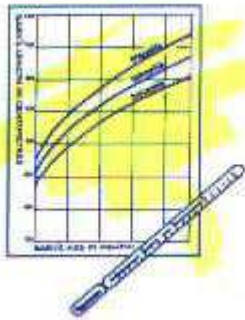
Antibiotics will be given immediately after the operation and for at least 48 hours. If your baby does not develop a high temperature and once oral feeds are started these will be often be given orally rather than through the drip.

A dressing will cover the wound on your baby's abdomen. A thin, soft rubber drain may have been inserted below the stitch line during the operation and this stays in for a few days to allow excess fluid to drain away- a bag may be put over this to measure the amount of fluid lost. The dressing will be changed as necessary and the stitch(es) removed, if necessary, when the wound has healed – usually about 2 weeks after the operation.

By the following day it may be possible for you to hold your baby, the nurse will help you. The Doctors will listen to the tummy with a stethoscope for sound of the bowel starting to work normally again after the operation. Once this happens, usually about the third day the baby's feeds will be restarted with frequent small amounts of sugar water (dextrose) which are slowly increased. When these are tolerated, diluted milk feeds will be given and the strength of these will be slowly increased until full strength feeds are resumed. If your baby was being breast fed before diagnosis at least a partial return to this method may be possible. It is important that the milk supply has been maintained by "expressing" the milk while your baby is unable to feed. The ward staff will help and advise you this.

When will my baby be able to leave hospital and what happens next?

Providing that there are no complications following surgery, your baby will be home within two weeks of the operation. Before returning home the wound must be healed and your baby must have a normal temperature and have started to put on weight. You will be given an outpatient appointment. Before your baby leaves hospital, you will be taught all about the special medicines and feeds. It is quite common for the hospital staff to advise your General Practitioner (GP) and local hospital about your child's condition and any special aspects of care, which are needed. They can offer you useful support when you are at home.



When you get home you must try and treat your baby as normally as possible. Obviously you will be worried at times and this is quite normal. Babies with biliary atresia get common illness just like any normal baby. However, if your baby has a high temperature for more than 24 hours or becomes jaundiced, or if there is an obvious change in colour of the urine and stools, or your are worried, it is important for your to see your doctor as soon as possible.

Unless there is some problem other than biliary atresia it is very important that your baby is immunized against Diphtheria, Whooping Cough, Tetanus, polio, Hib and MMR in the normal way. It does not matter if the immunisations are a little late starting because of the operation and hospital stay.

If you or your local doctor have any queries regarding your child, please contact the Consultant or Ward Sister at the hospital treating your child

What medicines may my baby need and why are they given?

The medicines needed will depend upon your child's condition and progress but some commonly used are:

Antibiotics – After the operation a course of antibiotics is initially given by "drip" and completed by mouth. They are used to reduce the risk of infection spreading from the gut into the liver. Further courses will be prescribed as required.

Vitamins – Additional vitamins are necessary because poor bile flow can reduce absorption of vitamins. Some may need to be given by injection.

Phenobarbitone – This may be given to increase the flow of bile. As it may make your baby sleepy at first it is usually given in one dose a day, in the evening.

Cholestyramine – This comes in the form of a powder in individual sachets. Each sachet is mixed with water and given in liquid form. This may be given to improve liver function and remove substances which may otherwise cause the skin to itch. Cholestyramine can affect the absorption of some drugs, especially vitamins. Therefore, with babies it is best to give some at each feed time except when vitamins are given. As your baby becomes older and manages larger volumes the daily dose of Cholestyramine can be given all at once or split into two. For a toddler or an older child needing Cholestyramine, it is best given in one or two doses away from the main meal times and vitamins. Always leave two hours between giving Cholestyramine and vitamins.

Sprironolactone – This may need to be given to help your baby get rid of excess fluid which otherwise collects in the abdomen and is called ascites. It is not needed in all cases.

URSO – This drug may be given at the onset of feeding after Kasai. It is thought to help promote the flow of bile.

Ranitidine – It is common to find stomach irritation after any type of abdominal operation and this drug may be given to help alleviate this problem.

If your baby/child needs medicine to reduce a temperature, remember aspirin, and drugs containing aspirin, must not be given; it is safer to sponge your baby with lukewarm water but if this does not work use paracetamol. Ibuprofen should not be given.

How is it possible to tell if the operation has been successful?

At the time of operation the surgeon will not be able to tell if bile drainage will occur satisfactorily. The first stools after the operation are usually very dark, they then will become paler. If bile drainage does occur the stools gradually change to a yellow / green / brown colour and later the urine becomes paler.

The colour of the stools will be monitored whilst your baby is in hospital. It is not unusual for there to be some variation in the colour of the stools. It is impossible to say how long it may take to observe these changes as every baby is different.

If bile drainage is established the jaundice gradually fades and blood tests show that the chemical, bilirubin that causes jaundice, drops towards a normal level. Since the operation may not drain bile from all sections of the liver it is not unusual for liver function tests to remain abnormal even if the jaundice completely clears.

Satisfactory bile drainage occurs in approximately 80% of children who undergo the Kasai operation before the age of eight weeks. The incidence of successful bile drainage is lower if the baby is over 8 weeks old. However, even some of the children who lose their jaundice may develop some of the complications due to liver damage. So all children continue to be seen for regular checkups.

Professor Kasai who first performed this operation in Japan in the 1950's has found that 90% of patients who lose their jaundice are alive and well at almost 15 years of age. Since the operation is a relatively new one, the longer-term prognosis is, as yet, less certain.

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

In these cases bile produced in the liver builds up causing damage to the cells of the liver eventually resulting in severe scarring (cirrhosis). If this continuous liver function deteriorates, complications occur and general health suffers. In the past sadly these children often died in infancy, but liver transplantation has altered this outlook and is now an accepted form of treatment. It is only considered when other treatments are ineffective.

Liver transplantation became available for children in the 1980's, the results are now encouraging and are steadily improving. There is an overall 80% one-year survival rate for liver transplantation. Liver transplantation is now possible in children of any age.

If liver transplantation is indicated for your child this will be discussed with you by the team caring for you and your child. Diet and medication will be intensified in order to ensure that your baby or child is as well as possible leading up to transplant, and progress will be monitored. The aim of liver transplantation is to restore your child to good health.

What other problems might occur?

A number of problems may occur of which you should be aware:

Cholangitis

This is an infection of the bile ducts in the liver resulting in inflammation.

This may cause a fever or jaundice or both. It requires urgent treatment with intravenous antibiotics which can only be given at your local hospital or the Unit treating your child. If the treatment is given locally, please ask the doctors to contact the Liver Unit treating your child, if you attend one.

If cholangitis is left untreated it can cause further damage to liver. That is why prompt investigation of any high temperature (over 37.0 C) or a non-specific illness lasting for than 24 hours is important.

Ascites

This is pronounced Ay – sigh – tees and is an abnormal collection of fluid in the abdomen. This can be present for up to 6 weeks after the operation but should then disappear. If it recurs at any time this is usually a sign that the disease has affected the ability of the liver to make a protein called albumin.

One of the blood tests performed frequently after the operation and each time blood is taken in outpatients is to measure the albumin level. When the albumin level is too low, water from child's blood "leaks" out of the blood vessels and collects in the abdomen (in severe cases it may also collect in other parts of body).

The first sign of this problem is often when clothes do not fit so well over the tummy. This usually occurs gradually but may be started or worse by infection.

The doctor may measure your child's abdominal girth at each hospital visit and/or may ask you to do this at home. If your child's tummy becomes noticeably bigger while at home you should see your GP to exclude other causes such as wind or constipation for which he/she can advise you. Treatment for ascites may also be started by your GP or your local pediatrician in consultation with the specialist unit and does not necessarily mean admission to hospital.

Treatment is initially by medicine and/or altering the diet to maintain calorie intake but reduce fluid and salt intake. In more serious cases it may be necessary to admit your child to hospital and give albumin intravenously through a "drip". You should never attempt to alter your child's diet and, in particular, their fluid intake without medical advice.

Portal hypertension.

Portal hypertension means high blood pressure in the portal vein, the main vein carrying blood from the gut to the liver and is different from high blood pressure. This may occur due to scarring in the liver, which causes backpressure to the portal vein. This may cause veins like varicose veins to develop in the lining of the gut, stomach or gullet.

These veins become fragile and bleed. It is rare for this to occur before the child is two years old. Bleeding can cause black stools, pallor or vomiting of blood. If bleeding does occur, then contact your local hospital immediately. Treatment is available.

NEVER GIVE YOUR CHILD ASPIRIN OR IBUPROFEN OR ANY MEDICINES THAT CONTAINS ASPIRIN AS THIS MAY START THE ONSET OF BLEEDING.

Itching (Pruritus)

Some children with reduced bile flow experience itching of the skin. This can develop at any time and can make a child quite irritable and miserable. If you think your child may be affected consult your GP or specialist unit additional medicines may be required.

What will happen in the future?

Your baby / child's growth and development will be followed up by periodic visits to the hospital. The frequency will vary with each child. Blood tests will be performed to measure the bilirubin level and the liver function. Periodically an ultrasound may be performed to assess the size of the liver and other structures in the abdomen (e.g. the spleen).

It may be that at some point in the future a short admission to hospital may have to be arranged, no matter how well your child is doing to generally assess progress.