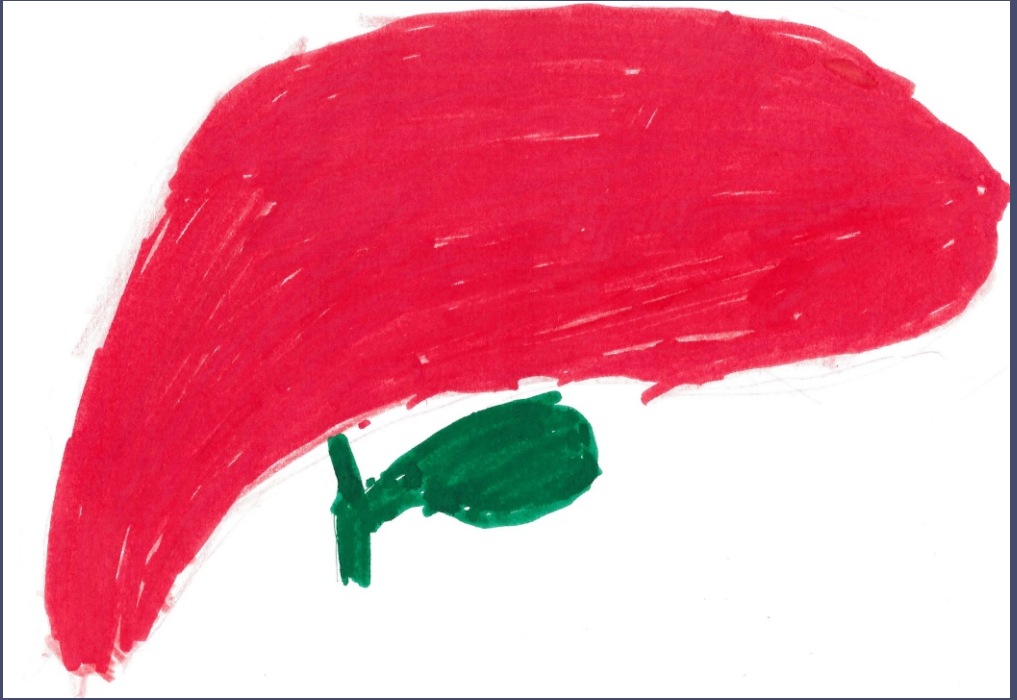




An Information for Patients and Parents

Biliary Atresia



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**Universitätsklinikum
Tübingen**

Dear patient, dear parents,

Biliary atresia - and now what? The diagnosis you have been given by your doctor certainly raises many questions, and you may feel uncertain and helpless.

Our goal is to provide you with the best possible support in the current situation. This brochure provides you with the most important information about biliary atresia. In addition, our team is always open to further questions. Please feel free to contact us!

Your pediatric gastroenterology/hepatology, endoscopy and nutrition team

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1. What is Biliary Atresia

Bile is an important digestive secretion and is produced in the liver. But what happens if the bile cannot be drained from there, or not sufficiently? This can have serious consequences for the liver and other organs.

Biliary Atresia (BA) is a rare liver disease, in which the bile ducts that conduct bile from the liver into the small intestine are not intact. Inflammation has caused them to become stuck together, resulting in bile congestion in the liver.

BA can be imagined as the final stage of a development: at the beginning, there are inflammations in and around the bile ducts, which gradually scar the small tubes. As a result, the bile can no longer be drained properly. The bile accumulating in the bile ducts and in the liver, has an irritating effect there and causes further inflammation. Over time, the bile ducts become blocked and lose their function. The flow of bile from the liver into the intestine comes to a standstill.

Only a few weeks after birth the first signs of biliary obstruction, which are jaundice and discoloured stools, can be observed.

So far the causes of the disease have not yet been adequately clarified. Most variants of BA are not hereditary. That means there is no familial clustering.

However Biliary Atresia is one of the rare liver diseases. Only one in countries, the disease is more common, for example in Taiwan the rate is 1:5,000.

Despite its rarity, it is the most common reason for a liver transplant in childhood: more than half of the liver transplant patients under the age of 2 are Biliary Atresia patients. Without medical treatment, the disease would lead to liver failure within the first two or three years and without liver replacement to death. about 18,000 children develops BA. In Asian countries, the disease is more common, for example in Taiwan the rate is 1:5,000.

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2. The healthy liver-biliary system

The healthy liver-biliary system

Functions of the liver

The liver is the most important metabolic organ in our body. Nutrients are absorbed in the small intestine, transported via the portal vein to the liver and processed or stored there (for example, fat-soluble vitamins and sugar in the form of glycogen). The liver also produces the proteins of the blood plasma, which are important for transport, immune defense and blood clotting (see glossary). End products of metabolism and also exogenous substances are converted by the liver so that they can be excreted in the stool. Consequently, it also serves as a detoxification organ.

The liver is also the largest gland in the body. It produces bile, which is either released directly into the small intestine via the bile ducts or stored thickened in the gall bladder. There are intrahepatic (inside the liver) and extrahepatic (outside the liver) bile ducts. At the end, the small branches unite to form a large bile duct and flow together into the small intestine via the so-called papilla.

Components of bile

Bilirubin is responsible for the greenish-yellow colour of bile. It is produced when red blood cells are broken down. In the blood, bilirubin is transported bound to albumin. This form is also called indirect bilirubin because it cannot be transported directly in the blood. Before bilirubin can be excreted in the bile, it must be conjugated in the liver. In other words, its chemical structure is changed so that it becomes more water-soluble and can be transported directly. In contrast to direct and conjugated bilirubin, indirect bilirubin is also called unconjugated bilirubin. For the diagnosis of biliary atresia, it is important to know whether the direct bilirubin is elevated (see Chapter 4 and Glossary).

The healthy liver-biliary system

Apart from bilirubin, bile contains a large amount of bile acids, which are produced in the liver from the body fat cholesterol. They are responsible for binding the fat and with it fat-soluble vitamins (A, D, E, and K) in the small intestine to be better absorbed into the bloodstream.

In order to recycle core components of bile acids, a large part of these molecules is reabsorbed into the blood in the last section of the small intestine (ileum) and transported back to the liver - the so-called enterohepatic circulation (see Figure 1). A small part of the bile is not "recycled" but released into the stool and excreted together with unusable metabolic products. This includes the aforementioned bilirubin, which also contributes significantly to colour the stool.

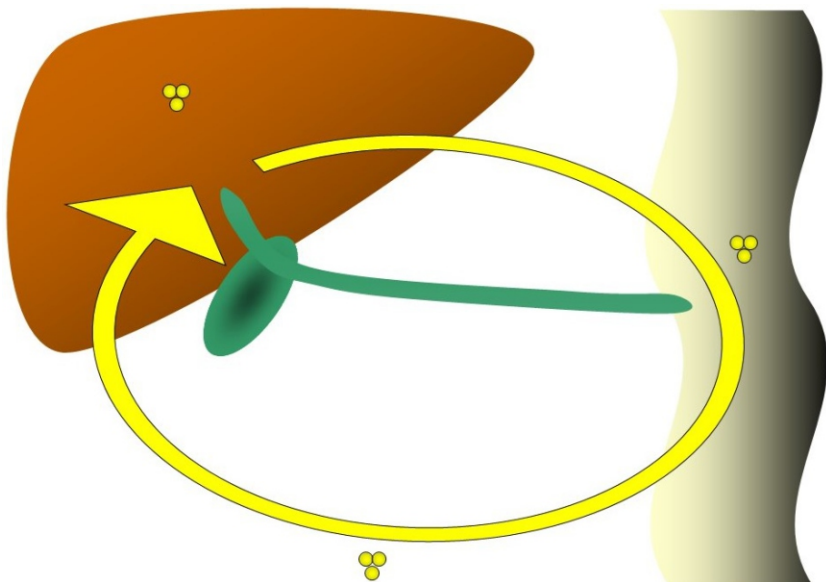


Figure1: Enterohepatic circulation

3. Causes of Biliary Atresia

The causes of Biliary Atresia and the development of bile duct obstruction are not yet fully understood. Biliary atresia should be seen as one manifestation of several different causes. It is assumed that in many cases an infection around birth or shortly afterwards is the triggering event. In combination with other factors, this leads to BA.

It is known from a similar disease in sheep that toxins also destroy the bile ducts and thus lead to scarring and cholestasis. The poison biliatresone, for example, which originates from the plant world, destroys in higher concentrations the extrahepatic bile ducts outside the liver. However, this effect has only been seen in sheep and it is not certain whether the results can also be transferred to humans.

The most common form of BA occurs in an isolated fashion, which means that it is not associated with other malformations. Only the bile ducts and the liver are affected. The pregnancy is usually unremarkable and most of the newborns with this form of BA are born mature, so that either in this case a specific cause cannot be identified. According to current data, various incidents before birth together contribute to the development of this biliary atresia: Among others, viruses, such as the cytomegalo-virus, and the subsequent activation of the immune system following the virus exposure seem to play a role.

In other forms of BA, there are malformations of other organs in addition to the bile duct changes, which are then impaired in their function. In BASM (biliary atresia spleen malformation syndrome) you can find polysplenia (many small spleens) or asplenia (no spleen), altered blood vessels in the abdomen and heart defects in addition to the biliary obstruction. Moreover, positional anomalies can often be seen, i.e. an altered rotation or position of the intestine, the heart or even a *situs inversus* (mirror image arrangement of the body organs). This strongly indicates disorders in prenatal organ development. BASM occurs in 10-20% of children with biliary atresia.

4. The Course of the disease

The first symptoms of a BA can be recognized within a few weeks after birth. The first sign of biliary obstruction is yellowing of the skin, also called jaundice or icterus. Shortly after birth, BA is often mistaken for the common newborn jaundice, as in which jaundice also occurs due to an accumulation of bilirubin. This physiological, i.e. not pathological, neonatal jaundice usually disappears within two weeks after birth. However, jaundice caused by BA otherwise is longlasting and is accompanied by a discoloured pale stool (see figureFigure 2). At the same time the normally colourless urine of the newborn is persistently yellow or dark in a child with BA. The cause of both the loss of pigment in the stool and the dark colouring of the urine is, that bilirubin can no longer be excreted through the bile and therefore does not colour the stool. It is then excreted with the urine via the kidneys.

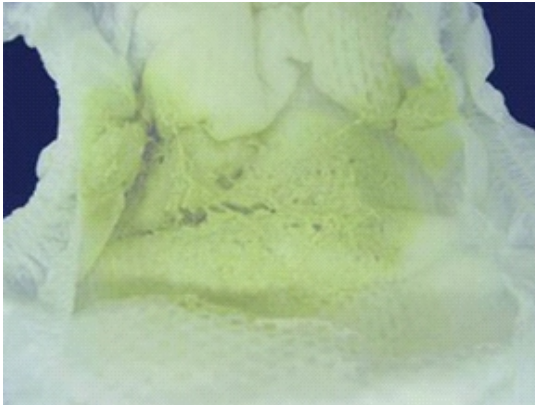


Figure2: Discoloured stool of a child with Biliary Atresia

In the further course, sometimes a strong itching occurs, which is probably related to the increased bile acid concentration in the blood in cholestasis.

Because fat and the fat-soluble vitamins A, D, E and K cannot be efficiently absorbed in the intestine due to the reduced or absent bile, a lack of vital fatty acids and a vitamin deficiency occur as the disease progresses. This may lead to significant failure to thrive. The vitamin deficiencies result in further complications (see also Table 1). For example, bleeding in the brain can occur due to the vitamin K deficiency, so that in the case of a suspected liver dysfunction, the coagulation must always be checked and, if necessary, vitamin K must be given intravenously, i.e. as an infusion directly into the blood.

Table 1:
Fat-soluble vitamins and consequences of vitamin deficiency triggered by bile stasis in BA

Vitamin A	Impaired vision (night blindness), susceptibility to infections
Vitamin E	Coordination disorders, disorders in the function and development of the nervous system
Vitamin D	Rickets (softening of bones)
Vitamin K	Blood clotting disorders (bleeding gums, nosebleeds, rarely severe bleeding, e.g. in the brain)

Bile stasis also directly damages the liver cells: The bile acids cause inflammation in the liver and its surroundings. The consequence is a progressive remodeling of the liver tissue with an increase in connective tissue - a so-called fibrosis develops. The liver initially becomes larger due to the inflammation and fibrosis. The fibrotic conversion of specific liver cells (the so called "stellate cells") into connective tissue cells, leads to a loss of function of the liver.

For example, serum albumin, a protein that is built in the liver and circulates in the blood, can no longer be produced in sufficient quantities. This in turn disturbs the distribution of water in the blood and other compartments of the body. Together with other causes this leads to accumulation of water from blood to abdominal cavity. This build-up of water is called ascites and can put in extreme cases a great strain on the cardiovascular system, the kidneys and breathing and can diminish quality of life.

If BA remains undetected or the therapy is unsuccessful, fibrosis (reversible in early stages) progresses and nodular scarring develops. This stage is called cirrhosis and is in most cases non-reversible. The cirrhotic liver leads to a congestion of blood streaming into the organ from the spleen and intestine. In consequence, the size of the spleen increases (splenomegaly). In addition, so-called varices (comparable to varicose veins) are formed in the food pipe (oesophagus), stomach and, more rarely, in the intestine. This complication of BA can become visible through blood vomiting and black stools, as a sudden large loss of blood can occur with a variceal haemorrhage.

In this emergency situation, it is essential to call the ambulance service, seek help in the nearest emergency room and, if applicable, contact the liver centre treating the patient.

As cirrhosis progresses, the liver increasingly fails and must therefore be replaced by a liver transplant.

5. Diagnosis of Biliary Atresia

Early diagnosis is of crucial importance for successful treatment of BA.

Unlike harmless neonatal jaundice, in which indirect bilirubin is elevated, direct bilirubin is elevated in jaundice caused by biliary atresia (see also Glossary). A simple blood test can distinguish between both forms. For this reason, experts recommend that if a newborn is still jaundiced for longer than 2 weeks, with or without discoloured stools, not only total bilirubin but both forms, direct and indirect, should be determined in the laboratory. One small blood sample is needed for this test.

Pediatric hepatologists are discussing this blood test for bilirubin differentiation to be integrated into the newborn screening in order to be able to start the treatment of BA as early as possible.

In addition to jaundice, the color of stool and urine can also provide initial clues. Parents can compare their child's stool with a so-called stool chart (see Figure 3) and see whether the stool is discoloured in a worrisome way. Currently apps for the smartphone that can be used to check the stool colour are in development.



Figure 3: Stool chart (Wildhaber BE, Hepatology 2011)

Diagnosis of Biliary Atresia

If blood results suggest biliary atresia, ultrasound is used to visualize the gallbladder and bile ducts. Physicians can see if the gallbladder is present and/or abnormal in shape. A more precise visualisation of the bile ducts and thus a more reliable diagnosis is possible with an endoscopic examination called ERCP. The bile ducts are examined using a type of tube (= catheter), at the end of which there is a small camera and a light source (see also Figure 4). Instruments can also be inserted through the catheter, which can be used, for example, to remove small amounts of tissue and perform interventions. X-ray contrast agents enter the bile ducts through the tube. Subsequently, an X-ray can visualize the small bile ducts. If the bile ducts can be displayed properly by ERCP, BA is ruled out at the time of examination.

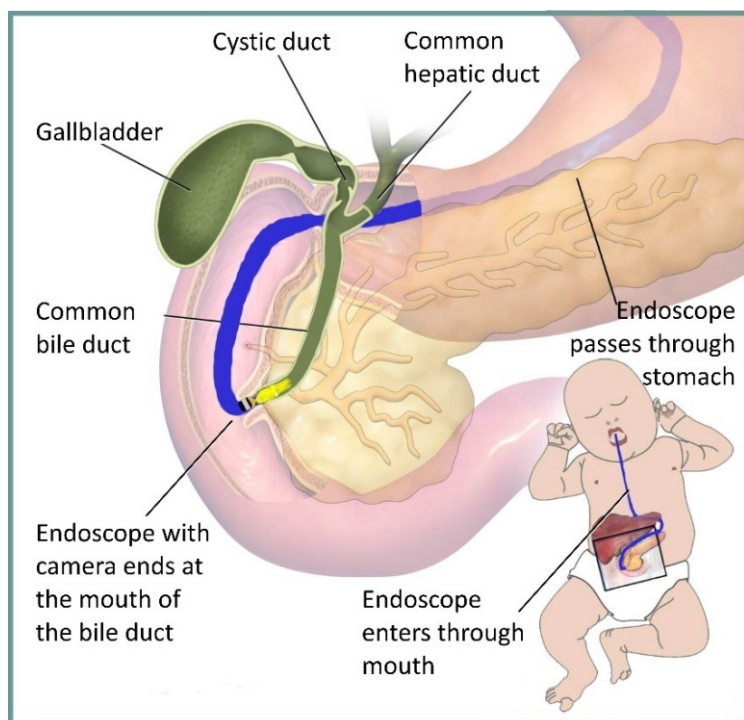


Figure 4:
Performance of an ERCP

A liver biopsy is used to examine the liver tissue for typical changes in the bile ducts and their surroundings. This can confirm the suspicion of atresia and rule out other causes for the symptoms. It also allows doctors to determine the extent to which biliary atresia has already damaged the liver. In this short biopsy procedure, which is performed under sedation and sufficient pain control, the physician removes a very small cylinder of tissue from the liver through a skin prick with a hollow needle.

Specialized pathologists then examine this sample under the microscope in a laboratory. Complications during biopsy removal, such as bleeding or injury to other organs, are rare. If the liver biopsy is unremarkable at an early stage of the disease, it may be worthwhile to repeat it at a later time.

If this procedure is necessary for your child, you will receive detailed information in advance from your attending physicians.

6. Therapy of Biliary Atresia

The primary therapeutic option used to treat biliary atresia in most cases is portoenterostomy according to Kasai (named after the Japanese doctor Morio Kasai, who developed it). Liver transplantation is an option for treatment in those children who do not recover after the Kasai procedure and require organ replacement. Accompanying BA symptoms are treated with medication and the child's nutrition is also supplemented to achieve an adequate level of thriving.

The Kasai operation

The Kasai operation should take place as early as possible which is crucial for its success. It should therefore be performed preferably in the first 8 weeks of life. To perform the Kasai procedure, surgeons first carefully remove the damaged ducts outside of the liver. They use a small segment of the patient's own intestine to replace the ducts at the spot where bile is expected to drain. This segment not only connects to the liver, but also connects to the rest of the intestine. The Y-shaped passageway formed by the Kasai operation allows bile to flow from the liver into the intestine (see Figure 5).

Therapy of Biliary Atresia

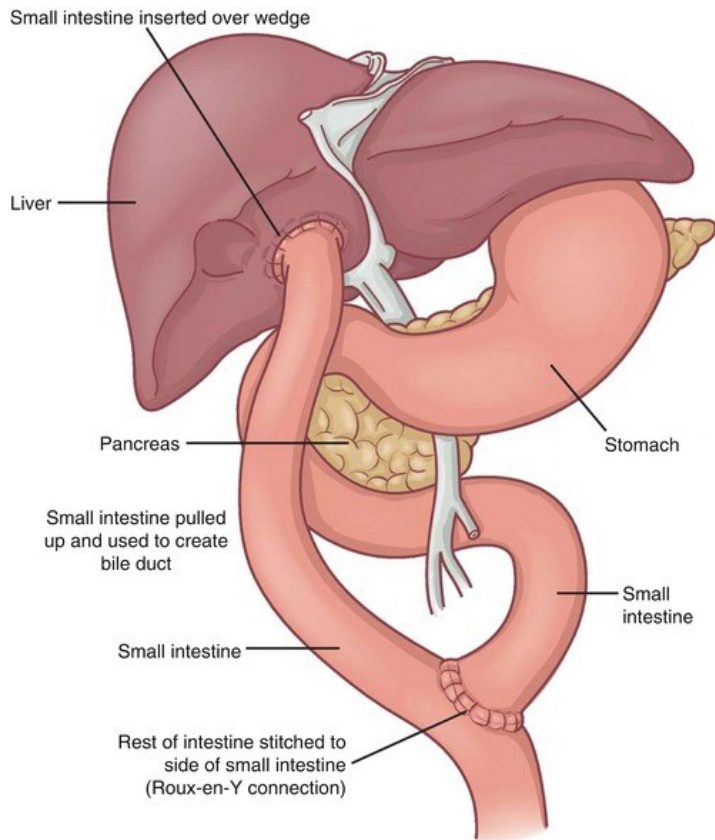


Figure5: Kasai surgery (McCance and Huether, Pathophysiology, 2010)

However, complete and life-long recovery of the liver is achieved only in a minority of cases. About one third of patients benefit from this operation long term, over years, and are able to grow up to adolescents without complications. A second third will benefit for a few months or years and another third will require liver transplantation soon after the failing Kasai procedure.

After the operation, the child's circulation, breathing and excretion are closely monitored and painkillers and antibiotics are administered. An abdominal drainage system is used to drain the wound fluid. In some cases, complications can arise after Kasai surgery. For example, excessive fluid-build up in the abdomen (Ascites) can be an unwanted consequence of the Kasai operation.

Whether the operation was successful can be determined by the child's bilirubin level and stool. The first stools are still dark from the blood of the surgical suture. When the liver starts to regenerate, and bile flow re-commences, pigment ideally will be visible in the stool after a few days. When the stool turns more and more yellowish, greenish or brownish, and the urine also becomes more colourless, it is a prognostically positive sign. If the stool remains colourless, it may mean that the liver is not regenerating sufficiently.

Liver Transplantation

If the Kasai surgery fails, the patients may require a liver transplantation. In early failure this may be required in infants, if a partial regeneration took place the patients in need for organ replacement may be older. The majority of BA patients will need a donor organ sooner or later. The transplant procedure is well established in experienced centres. Benefits and risks require detailed discussion between the experts and the family. In experienced European centres, 1-year survival after liver transplantation for BA is above 90% for most patients irrespective of age.

When is the replacement of the liver no longer preventable?

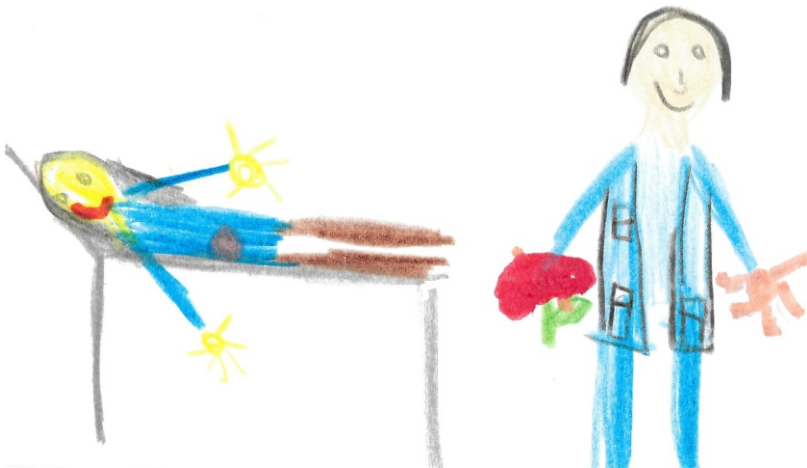
Two to six months after the Kasai procedure, the bilirubin level should be near to normal in those likely to recover long-term. Progressive liver cirrhosis with complications, such as the already mentioned liver congestion (portal hypertension), and liver failure may also indicate the need for planning a liver transplantation.

Therapy of Biliary Atresia

New methods of liver transplantation make it possible for more patients of different ages to have a chance of recovery. In the split technique, the donated liver is divided. The smaller left lobe is usually given to children, the right lobe to an adult.

In living donation, the partial liver graft is donated by one parent or close relative. Mostly it is a part of the left liver lobe and the graft grows and adapts in the new body after transplantation. In order to avoid possible risks, the donor is carefully examined beforehand to make sure that he or she is healthy and suitable and that risks are minimized for both donor and recipient.

If a child needs a liver transplantation and none of the relatives are eligible, he or she is placed on a waiting list for organs in the Eurotransplant organization. The score used for organ allocation ranking is based on the age at entry on the list, the diagnosis and certain laboratory values that indicate the risk of liver failure. When the suitable donor organ is found, the family is informed by their liver centre. This can happen during day or night hours.



The operation takes about 6 to 10 hours. Before the new organ is put in, the old one is removed. After the operation, the child first moves to the intensive care unit for monitoring. Immediately after the operation, immunosuppression is started. These drugs suppress the body's own defense system to prevent rejection of the new liver. In general a lifelong medication is necessary so that the body does not reject the liver transplant.

If a transplantation is necessary for your child, we will provide you with comprehensive information about the procedure.

Medication

Apart from the Kasai operation, the symptoms of BA can be alleviated with the help of medication.

The aim of the medication is to keep the bile acid concentration in the blood within tolerable limits, above all to reduce the annoying itching. Ursodeoxycholic acid or (rarely) sometimes rifampin are often used to improve bile transport and metabolism. Ascites is usually treated with diuretic medications such as spironolactone. Difficult to treat cases may require hospitalization and intravenous (directly into the bloodstream) administration of albumin (protein).

New drugs that affect the enterohepatic circulation are currently being studied for their efficacy in BA.

Table2: Drug therapy for Biliary Atresia

Therapeutic Target	Medication
Optimisation of bile flow	Ursodeoxycholic acid
Therapy of itching by intervention of bile acid metabolism	Rifampin
Therapy of ascites	Spironolactone, Furosemide

Nutritional Support

Nutrition plays an important role in the therapy of BA. Due to the impaired absorption of fats in particular but also proteins and carbohydrates, failure to thrive, i.e. delayed weight and length development, can occur in the course of the disease. At the same time, children with BA have an increased energy metabolism, which is about 120 to 150 % of the average requirement. Medium-chain fats (MCTs) can be used to meet this increased demand. In contrast to long-chain fatty acids they can be absorbed in the gut without bile acids and then be used for energy production.

There are special infant formula diets that contain these MCT fats in particular. They can be prescribed by the attending doctor and counteract the development of failure to thrive at an early stage. In order to achieve a higher calorie intake in breastfed infants, MCT oil can be used in addition to breast milk. MCT oils can also be used to supplement baby foods after breast feeding and afterwards the normal diet.

In addition to sufficient energy supply, the supply of vitamins must also be taken into account, as not only fat but also fat-soluble vitamins (A, D, E, K) are poorly absorbed due to the reduced release of bile acids. To prevent a deficiency, these vitamins are administered by means of tablets, drops or, less frequently, as injections. In blood tests the level of vitamins can be checked. Controls of the weight and size are carried out to check that the babies are thriving and, if both the diet and the supplements need to be adjusted. Rarely, long-term parenteral nutrition is required.

The nutritionists at the liver center will support you from the beginning to ensure that your child receives the best possible care. This is also important because good care increases the chance of a successful liver transplant and the child recovers more quickly from the operation.

7. What can we as parents do?

What can we
as parents do?

First of all, parents often notice that something might be wrong if they notice the persistent jaundice or discoloured stool of their baby. Or the paediatrician might observe the jaundice at the first routine examinations. If you did not do already, ask your paediatrician to determine the different bilirubin levels in the blood (direct and indirect form) at an early stage. This is very helpful for early detection of a liver disease and possibly BA. If the stool of your child seems suspiciously colourless, you can check it with the help of a stool colour chart (see Figure 3) and discuss your concern with your paediatrician.

You can relieve the itching, that sometimes occurs later in the course of the disease, with moisturising creams. Skin-cooling interventions, such as a cooler bath or a cooler room temperature at home, can also help. Dress your child mainly in cotton clothing, avoid buying woolen or polyester clothes.

If your child is confirmed to have BA, the Kasai surgery is coming up in the near future. Before the operation, you will receive all the relevant information from the doctors of the gastroenterology/ hepatology and paediatric surgery team. If anything is unclear to you, do not hesitate to ask us your questions. The paediatric surgery and paediatric gastroenterology and hepatology team will also be on hand to give you advice and support after the operation.

After 10-14 days after the operation, you can usually leave the hospital with your baby. When your child is leaving the hospital, you will be informed about all the important points to be considered at home. You will be given a plan for medication at home. Breastfeeding after the operation is possible, but sometimes the doctor will also prescribe the special high-calorie baby food already mentioned. Talk to the dietician for tips on how to feed your child with a combination of breast feeding and formula diet. It is also advisable to take vitamins if the jaundice persists. These will be prescribed for you, so it is better not to take freely available multivitamin preparations.

What can we as parents do?

It is best to weigh your child weekly or have your child weighed by the paediatrician to monitor weight gain. A successful course is marked by decreasing bilirubin levels in the blood and thus also by an improving jaundice in the first 2-3 months after the operation. The jaundice may even disappear completely.

In any case, make use of the regular follow-up controls, because even if the jaundice disappears, complications can arise that damage the liver. During the follow-up visits, blood and liver function tests are carried out. The liver and spleen are also controlled by ultrasound at regular intervals.

In the near future, watch out for symptoms that may indicate deterioration of the liver. A colourless stool together with a general and unexplained malaise or fever should prompt you to contact your paediatrician first to help check for causes.

If the clothes suddenly tighten over the abdomen or the child gains weight rapidly, water retention could be the cause.

To be there for your child, you should also take care of yourself. Don't forget to eat, sleep, exercise and relax regularly. Perhaps you can take turns with your partner in caring for your child. It can help to talk to other affected families or to your friends and family about your situation. This will give you strength to be a good support for your child.



8. Who can give us help?

Who can
give us help?

You can get further information about the care and support of your child at home from the paediatric nurses and the nutritionists of our paediatric gastroenterology and hepatology team. If you need psychological support or social legal advice, the team of the psychosocial service of our childrens hospital is there for you.

If you are looking for further information online, you need to be sensitive to the right sources. In the following, we have compiled some reliable sources for you. Since biliary atresia is a rare disease, you will often not find specific information on atresia, but general information on liver diseases, transplantation and self-help.

European wide websites:

- European Rare Liver Disease Network: www.rare-liver.eu/
- European Society For Paediatric Gastroenterology, Hepatology and Nutrition: www.espgan.org
- Portal for rare diseases: www.orphanet.net

British websites:

- Children's Liver Disease Foundation (CLDF):
<https://childliverdisease.org/>

German websites:

- Verein Leberkrankes Kind e.V.: www.leberkrankes-kind.de
- Kinderhilfe Organtransplantation: www.kiohilfe.de
- Deutsche Leberstiftung: www.deutsche-leberstiftung.de
- Deutsche Leberhilfe e.V.: www.leberhilfe.org
- Lebertransplantierte Deutschland e.V.:
www.lebertransplantation.eu
- Self-help for rare diseases: www.orpha-selbsthilfe.de
- National Contact and Information Centre for the Initiation and Support of Self-Help Group

9. Glossar

Albumin

Albumin is a plasma protein, with a proportion of 60% the most important one, and serves primarily to maintain the so-called colloid osmotic pressure in the blood plasma. Thus it is also responsible for the correct distribution of electrolytes such as potassium, magnesium etc. in the body. If less albumin is produced in the liver, for example in the case of cirrhosis, the pressure is reduced and water flows into the space between the body cells. This leads to the formation of oedema (see also ascites).

Ascites

In cirrhosis, liver tissue may have been replaced by scar tissue which limits the function of the liver. The proteins that fulfill important transport tasks in the blood and ensure an osmotic balance are no longer produced sufficiently. As a result, water flows from the blood into the intercellular spaces of the abdomen. The abdomen may appear bloated from the outside.

BASM = Biliary Atresia Splenic Malformation Syndrome

In BASM syndrome, atresia of the bile ducts occurs in combination with modified spleens. There are either many small spleens that do not function properly (polysplenia) or there is no spleen at all (asplenia).

Bilirubin

Bilirubin is produced as a breakdown product of the red blood cells (erythrocytes) and is excreted from the body through the bile. In liver disease, bilirubin is responsible for the yellowing of the skin and mucous membranes by accumulating in the blood and being distributed to all parts of the body.

- unconjugated / indirect bilirubin

Bilirubin is insoluble in water and must be attached to a protein (Albumin) for transportation. (=indirect or unconjugated form). If this bilirubin species is elevated, increased break-down of red blood cells may be the cause.

- conjugated / direct

In the liver bilirubin is conjugated, that means an enzyme attaches glucuronic acid, making the bilirubin more water-soluble. Thus, it can be transported "directly" with the bile even in aqueous milieu. If the concentration of direct bilirubin in the blood is increased, it is an indication of impaired bilirubin transport in the liver or in the bile ducts.

Blood clotting

In order to prevent excessive blood loss when the blood vessels become injured the so-called haemostasis starts. Blood platelets (= thrombocytes) from the vessel wall attach themselves to the wound and fibrin, with the participation of the clotting factors, ensures further closure. If the liver becomes cirrhotic and the absorption of vitamin K decreases, fewer clotting factors are produced and there is an increased tendency for bleeding events, which can be dangerous under certain circumstances.

Cholangitis

Inflammation of the bile ducts. Bacterial cholangitis can occur as a complication after Kasai surgery. It may be indicated by new-onset jaundice and/or fever.

Enterohepatic circulation

"Recycling" of bile acids. Bile is released into the duodenum (first part of the small intestine) where it helps digest fat. In the ileum (last part of the small intestine) 95% is reabsorbed into the blood. This is the only way to provide the large amount of bile acids needed for fat digestion.

ERCP = endoscopic retrograde cholangiopancreatography

This is a combined endoscopic and X-ray procedure in which the bile ducts become visible by using contrast agents.

Extrahepatic

outside the liver

Fat-soluble vitamins

..are the vitamins A, D, E and K. "Fat-soluble" means that they have similar chemical properties to dietary fats. That is why they are absorbed together with them and with the help of bile from the intestine into the blood.

Bile

The main components of bile are bile acids, phospholipids (such as lecithin), bilirubin, electrolytes and water. It is formed in the hepatocytes (liver cells) and either released directly into the intestine or stored in the gallbladder. In the intestine, it is needed for the digestion and absorption of fat.

Gallbladder

..is an organ for the storage of bile. When the liver produces more bile than is needed for digestion, bile backs up to the gallbladder, where it is thickened for better storage

Bile ducts

The ducts in which bile flows from the liver into the intestine are called bile ducts. There are intrahepatic and extrahepatic bile ducts. Outside the liver, the bile ducts join to form a common bile duct, which together with the duct of the pancreas flows out at the papilla into the small intestine.

jaundice (= Icterus)

Jaundice occurs when the bilirubin concentration in the blood is significantly increased. In this case, it leaks out of the blood vessels and spreads to the tissues. At first the white conjunctiva of the eyes changes colour and becomes yellowish. At even higher concentrations, the skin also turns its colour. The causes of jaundice are manifold, a reduced breakdown of Bilirubin due to disturbances in the liver or a disturbed release into the bile can be responsible.

- physiological

Physiological jaundice is not caused by a disorder in the liver-biliary system. After birth, a lot of the fetal blood is still being broken down. That is why there is an accumulation of (unconjugated) bilirubin that turns the skin yellow. After 2-3 weeks after birth, this bilirubin build-up is metabolised or treated with the help of light therapy, in which the bilirubin becomes more water-soluble and can be excreted by the kidneys..

hepatic

related to the liver

("hepar" = Latin for liver)

Icterus

See Jaundice

Intrahepatic

within the liver

Itching (= Pruritus)

Itching is a very unpleasant feeling that affects the skin. Primarily, no changes in the skin can be observed, as would be the case with a skin disease. Only secondarily, as a result of the constant scratching, irritations or even bleeding occur. The cause of itching in liver or bile disorders is not yet fully understood. Presumably, the increased bile acid concentration in the blood is responsible. The pruritus can be generalised, that means over the whole body, or localized, limited to individual parts of the body. The suffering is extremely stressful and is often underestimated. It is often worse in the evening and at night.

Liver Fibrosis

The liver is remodeled by the storage of connective tissue. Inflammatory processes are also involved. Attempts to regenerate the damaged tissue can lead to scarring of the liver and a progressive loss of function.

Spleen

Located under the left costal arch, next to the stomach, this organ is involved in inflammatory processes and removes dead blood cells. In BASM, there are many small spleens (polysplenia), which cannot protect the infant adequately from infection, or no spleen at all (asplenia).

Portal Vein

..is also called Vena portae and collects blood rich of nutrients from the digestive organs and leads it to the liver.

Situs inversus

... belongs to the positional anomalies and is characterised by a mirror-image arrangement of the internal organs, for example the liver is located on the left whereas the spleen is located on the right side. Whether complications occur depends on whether the situs inversus occurs in isolation or is accompanied by other disorders, for example heart defects.

Varices

... are varicose veins, i.e. veins in the area of the oesophagus or stomach whose walls are dilated and thus thinner. The cause is the high blood pressure in the portal vein caused by cirrhosis of the liver. When the varices rupture, bleeding events can occur in the upper part of the digestive tract. Vomiting of blood is the consequence.

10. References

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11. Figures

Figure 1: Enterohepatic circulation Page 5

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<https://creativecommons.org/licenses/by-sa/2.5/deed.de>]

Figure 2: Discoloured stool of a child with Biliary Atresia Page 9

Figure 3: Stool chart Page 13

[Wildhaber, B.E.: Screening for biliary atresia: Swiss stool color card.
in: Hepatology, 2011 July; 54 (1):368-368]

Figure 4: Performance of an ERCP Page 14

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Figure 5: Kasai surgery Page 17

[McCance and Huether, Pathophysiology, 2010]



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