Biliary Atresia (BA)

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

Biliary atresia (BA) is a liver disease that occurs in infants only. Bile is a liquid made in the liver and is required to digest food. Once bile is made, it is stored in the gallbladder and is squeezed out by the gallbladder into the small intestine after meals. In biliary atresia, the ducts where the bile flows from the liver to the gallbladder and the gallbladder to the small intestine become inflamed and scar down. This situation results in bile becoming backed up in the liver, causing the liver to get enlarged, inflamed, and scarred. If untreated, biliary atresia can cause liver failure and need for transplant. BA is the most frequent cause of liver-related death in children and the most frequent reason for liver transplantation.

The incidence of BA varies between 1:5,500 for Asians and 1:19,000 for Caucasians. The cause of BA unknown, but most experts believe inflammation and scarring caused by the immune system causes this problem.

Signs and Symptoms - "What symptoms will my child have?"

Early signs: It presents in early newborns with yellow skin (jaundice). The whites of the eyes will turn yellow (icterus). The stools become light grey when normally they should be yellow, green, or brown. For the most part, babies have no symptoms. They eat and drink well. It is normal for early newborn babies in the first two weeks after birth to have jaundice, especially if they are breast fed. However, jaundice beyond 14 days is not normal and should be investigated by a doctor.

Later signs and symptoms: Later signs include worsening of the yellow tint of the skin and eyes. Babies may have a hard time putting on weight. The urine color is dark brown. The belly gets swollen as the liver gets bigger and fluid (ascites) collect in the belly. If left untreated, biliary atresia can lead to failure of the liver, but this takes months to happen. Liver failure is life-threatening. Symptoms of liver failure include development of large blood vessels in the esophagus from back-pressure through the drainage system of the liver. These are called

varices and can bleed severely. The child would vomit blood. Large veins can be seen on the skin of the belly. The belly becomes very swollen.

Diagnosis - "What tests are done to find out what my child has?"

Blood studies: Blood will be collected to check the function of the liver. The bilirubin level is related to the degree of back-up of bile through the liver and indirectly the damage to the liver. In general, the worse the jaundice, the worse the bilirubin level. Other blood studies will be done to rule out other conditions that may also cause jaundice and liver damage such as infection or genetic problems.

Ultrasound: In this test, a probe is applied on the belly directly overlying the liver and gallbladder. The probe uses sound waves to look at the gallbladder, liver, and bile ducts. If the gallbladder is small or absent, it is suggestive of BA. The size of the spleen will suggest the degree of back pressure through the liver. Ultrasound may also show other problems that may cause jaundice such as gallstones.

HIDA scan: This test is done in the nuclear medicine department. A tracer is given to the infant through the vein. Pictures are taken to see if the tracer is excreted through the liver into the bile ducts and followed into the intestine. If there is no excretion, then further investigations for BA are indicated. Often, the baby is given a medicine (phenobarbital) for five days to increase the effectiveness of the HIDA scan.

While the previous studies can suggest that your baby may have BA, the main study that will prove it is a **cholangiogram**.

Cholangiogram: A cholangiogram is a study where dye is injected into the gallbladder to see the structure of the main bile duct draining the liver. If the bile duct is not present, then the baby has biliary atresia. This study can be done by a radiologist under the guidance of an ultrasound or by a surgeon in the operating room.

Liver biopsy: A liver biopsy may also be done by a radiologist or during surgery. While this test does not always confirm biliary atresia, it can be highly suggestive of the diagnosis. It also helps estimate the amount of liver damage that has occurred due to the disease.

Conditions that mimic this condition: Doctors will rule out other diseases that cause jaundice such as liver infection or inflammation (hepatitis). Bacteria and viruses can cause liver damage. Another condition that needs to be checked is alpha-1 antitrypsin disorder. It is a genetic disorder that affects the liver and lung. Another possibility is Alagille syndrome, a condition where there are fewer bile ducts than in the normal person.

Treatment - "What will be done to make my child better?"

Medicine: No specific medical therapy is available. Surgery is needed to allow bile to drain from the liver into the small intestine.

- Doctors may change your baby's formula to something that would be better tolerated by the liver.
- Your baby may need to take special vitamins (D, A, K, E) because these particular ones are not well absorbed when the liver is sick.

Surgery: How the surgery is started depends on how sure the surgeons are of the diagnosis of biliary atresia. If the diagnosis of biliary atresia before the surgery is not clear, then a dye is injected into the gallbladder to define the structure the bile ducts (cholangiogram) and a liver biopsy are performed. If no ducts are present, then the surgeon proceeds with the operation to drain the bile from the liver to the small intestine. This procedure is called the "**Kasai Procedure**". It is named for the surgeon who invented the surgery. In this procedure, the surgeon finds an area in the middle of the liver where there are small tubes that may be able to drain bile from the liver. A piece of small intestine is sewn on to this area of the liver for bile drainage.

- At the time of the procedure, if the cholangiogram shows that there is a normal bile duct from the liver to the intestine, a Kasai procedure will not be needed.
- When a cholangiogram is done in the course of this procedure, the gallbladder is usually removed.

Preoperative care: The doctors may give vitamin K for three days before surgery. When there is not enough vitamin K in the body, there is a higher chance of problems with bleeding.

Antibiotics (medicine to fight infection) will be given through the vein just before the operation to decrease risks of wound infection after surgery.

Informed consent: A consent form is a legal document that states the tests, treatments or procedures that your child may need and the doctor or practitioner that will perform them. Before surgery, your doctor should tell you what the operation is, the goal of the surgery and other possible treatment options that are available. Your doctor should explain the risks and benefits of the surgery. You give your permission when you sign the consent form. You can have someone sign this form for you if you are not able to sign it. You have the right to understand your child's medical care in words you know. Before you sign the consent form, make sure all of your questions are answered. It is important to know that during surgery, there are things that can happen that your doctor may have not predicted before going in. He or she will explain these to you after the surgery.

Postoperative care: Where the infant will be cared for after the surgery depends on many factors, including the general health of the baby, amount of blood loss in the operating room,

and how long the surgery took. The surgeon and the anesthesiologist will decide whether the baby needs to be in an intensive care unit or the regular ward. Sometimes, a ventilator (breathing machine) may be needed for a day or two to support breathing. The infant will receive antibiotics to prevent infection. Pain medicine will be given through the baby's IV until they are able to start eating again, at which time pain medicines can be given by mouth.

The baby will be fed once they pass stool and gas. The doctors may be interested to know the color of the stool. If the stool is yellow, green, or brown, it is an indication that bile is being drained successfully from the liver into the small intestine.

Risks:

- **Bleeding**: If there is blood loss during surgery and/or the baby started with a low blood count (anemia), the infant may need a blood transfusion.
- **Leak or hole in the intestine**: During the surgery, the surgeons will sew the intestine to the liver or to another segment of small intestine. If the areas where the intestines are sutured do not heal properly, a hole results with leakage of intestinal contents into the belly. This is a serious condition that is signaled by fever, vomiting and belly pain in the first few days after surgery. An operation will be needed.
- **Intestinal blockage** (obstruction): Whenever anybody undergoes surgery in the abdomen, there is a chance of intestinal blockage. The most common cause is internal scarring that can kink the intestines. Intestinal obstruction can happen at any time, from a few days after surgery to years after the surgery is complete. In some cases, surgery may be needed to release the blockage.
- **Infection** in liver (cholangitis) and /or blood: The infant will have fever, jaundice, light stools, and abnormal blood studies. This is treated with antibiotics, usually given through the vein. This can be a life-threatening infection and therefore, immediate medical attention is needed.

Benefits:

The Kasai procedure is the treatment for biliary atresia. If the surgery is successful, the infant may grow to adulthood and the liver function may remain stable, saving the liver from further damage.

Sometimes, even after the Kasai procedure, the damage to the liver is bad enough that the liver eventually fails, and a liver transplant is needed. In these cases, the Kasai procedure allows the child to grow for several years. Obtaining a donor liver for transplant becomes easier because the child is bigger.

It is important to know that the earlier the diagnosis of BA is made and the sooner the Kasai procedure is performed, then the damage to the liver is minimal. The results are better if the baby has the surgery within six weeks of birth. If the diagnosis of BA is delayed and the Kasai

procedure is done later than three months of life, there is likely damage and scarring to the liver that the baby cannot recover from. These patients will have a higher risk of eventually needing a liver transplant.

Home Care - "What do I need to do once my child goes home?"

Diet: The infant may be given a special formula that is better tolerated by the liver.

Activity: There are no activity limitations.

Wound care: No special wound care at time of discharge.

Medicines: Clarify with physicians prior to discharge. The most common medicines are vitamins, pain medication, and antibiotic to decrease risk of infection of the bile duct and liver.

What to call the doctor for: Fever, vomiting, any redness or drainage from the surgical wound, worsening jaundice.

Follow-up care: Regular doctor visits are needed to make sure the infant is growing and gaining weight. Bloodwork is done to make sure that Kasai procedure is allowing good bile drainage from the liver and if the liver's function remains satisfactory.

Long Term Outcomes - "Are there future conditions to worry about?"

In general, one-third of infants undergoing the Kasai procedure avoid transplant, one-third have liver failure in the first year, and one-third progress to liver failure later in childhood. The long-term survival of BA infants with their native liver, without liver transplant, depends largely on the condition of the liver at the time of the Kasai procedure. A successful surgery restores flow of bile from the liver to the small intestine and lessens further damage to the liver.

- Infants with a shorter duration of jaundice have a better response to the Kasai procedure. This is because the longer the liver does not drain bile, the more injured and scarred it becomes, and the Kasai procedure cannot reverse the damage already done.
- After a Kasai procedure, babies can have an infection of the bile ducts, spreading up into the liver (cholangitis). This presents with fevers, chills, yellow skin, yellow eyes, and light-colored stools. This is a very serious infection and requires immediate medical attention. If the baby has repeated episodes of cholangitis, it can scar the liver more and may lead to overall worsening of the liver function.

Updated 2019
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