A PARENT’S GUIDE TO BILIARY ATRESIA

WHAT IS BILIARY ATRESIA?

Biliary atresia is a liver disease of infants characterized by progressive blockage of the ducts that drain fluid or bile from the liver to the intestine. The bile ducts are the tubes between the liver and the intestine. The cause is unknown and it happens in the U.S in about 1 in 15,000 live births. Successful treatment requires early diagnosis and relief of the biliary blockage through surgery.

HOW DO I KNOW IF MY BABY HAS BILIARY ATRESIA?

Babies with biliary atresia may develop stools that are pale in color or gray/white (called acholic). Jaundice (yellow skin or eyes) in infants that continue past the first 2 weeks of life needs to be evaluated as it could be a sign of biliary atresia. Babies with biliary atresia do not usually appear ill or malnourished. These babies may have a large liver and/or spleen. A pediatric gastroenterologist and pediatric surgeon will be consulted to help care for your baby and do some tests to determine if the baby has biliary atresia. Depending on their age, the baby may need to be admitted to the hospital to speed up the evaluation.

HOW IS BILIARY ATRESIA DIAGNOSED?

A complete blood work analysis will be done. An ultrasound of the liver and gallbladder will be performed. Infants with biliary atresia will usually have a shrunken or absent gallbladder and abnormal bile ducts. If the workup is concerning for biliary atresia, the next step is to perform a liver biopsy and cholangiogram. A cholangiogram is a study where a dye is injected into a duct or gallbladder. This provides a picture of the bile ducts. This procedure may be performed under general anesthesia by a pediatric radiologist or a surgeon.
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ONCE BILIARY ATRESIA IS DIAGNOSED OR HIGHLY SUSPECTED, WHAT'S NEXT?

The surgical team will need to prepare the baby for surgery. Your baby may be placed on a clear liquid diet the day before surgery. Lab tests will be done to determine if your baby’s clotting ability is normal and or if blood for transfusion is necessary. If your baby’s clotting studies are abnormal, vitamin K or blood products may be necessary to correct the clotting abnormalities. In addition, oral antibiotics and a laxative medicine may be used to clean out the intestine. This will help reduce the bacterial count in your baby’s intestine. Your infant will also require IV fluids and will be made NPO (nothing per mouth) a few hours before surgery. You will visit with the pediatric surgeon who will explain the procedure, risk, benefits and alternatives of the surgery. You will also visit with the pediatric anesthesiologist who will address all your questions about anesthesia, need for additional monitoring lines, and if needed, an epidural for pain control. Often babies will have the cholangiogram test mentioned above and surgery to repair biliary atresia (assuming the cholangiogram is positive for biliary atresia) at the same time.

WHAT EXACTLY IS A KASAI PORTOENTEROSTOMY?

The initial goal of the surgery is to confirm the diagnosis of biliary atresia. A large upper abdominal incision is needed. This is necessary to allow the proper exposure for the surgeon to perform the operation safely. A liver biopsy and cholangiogram may be done to confirm the diagnosis. If the cholangiogram does show biliary atresia then the surgery called KASAI PORTOENTEROSTOMY will be performed immediately. The next step is to remove the gallbladder and blocked bile ducts to expose the area of the liver where any remaining ducts may be. The operation is completed by connecting a segment of intestine to the liver. A drain will then be placed to drain this area until it has healed. The drain will collect extra fluid from this area and will be on the right side of the abdomen.
WHAT HAPPENS AFTER THE SURGERY?

This is the time to support your child until the intestine and new biliary connection recovers and starts to function. This usually takes anywhere from 5 to 7 days. Nutrition after the surgery is important and may require supplementation by a feeding tube into the stomach or nutrition by vein. Pain will be controlled by means of IV medicine, oral medicine, and in some cases an epidural catheter placed by the anesthesia doctors. Blood tests will be closely monitored. Your baby will remain on antibiotics to help prevent an infection. Your child will likely have an NG tube (nasogastric tube) to help suction the stomach and intestinal juices and a foley catheter which will be used to monitor the urine. Once your baby is showing signs that the bowel is starting to function by passing gas and stool, the baby will be allowed to feed. The next step is to remove the tubes and epidural catheters as your baby no longer needs them. Once your baby is eating well and having normal bowel movements, then you will be able to go home.

WHAT IS THE MAJOR POSTOPERATIVE COMPLICATION AFTER A KASAI PROCEDURE?

Cholangitis is an infection of the bile ducts as a result of bacterial contamination from the intestine. It can occur at any time following the surgery and requires immediate medical attention. Cholangitis usually responds to antibiotics. Babies with cholangitis can present with fever, jaundice, white stools, feeding problems, belly pain, or bloating.

WHAT ARE THE LONG TERM RESULTS OF THE KASAI PROCEDURE?

A successful Kasai protects the liver from further damage and resolution of jaundice and return of normal color to the stools. For many babies the liver continues to deteriorate and they will require a liver transplant. The goal of the Kasai is to protect the liver as long as possible, but most patients at some age will need to have a liver transplant. About a third of patients will have a great response to surgery and clear their jaundice. Some will get temporarily better but then gradually progress to biliary cirrhosis ultimately requiring a liver transplant. The other third of patients will have minimal to no bile drainage following the surgery and will rapidly progress to biliary cirrhosis requiring a liver transplant.

WHAT SHOULD I EXPECT ONCE MY BABY IS DISCHARGED FROM THE HOSPITAL?

Your baby will be followed closely by your gastroenterologist and surgeon and seen in the outpatient office regularly. He/she will be on vitamins and oral antibiotics. Ursodiol will be prescribed to help increase bile flow. Nutrition is extremely important and weight will be closely monitored. Periodic blood work will also be necessary to evaluate the status of the liver.

ONCE I’M HOME, WHEN SHOULD I WORRY?

If your baby is eating and having normal colored stools and feeling well with no fevers, then they are recovering just like we would expect after this operation. If your baby develops fevers, jaundice, acholic (white) stools, vomiting, abdominal distention or pain, then he/she should be seen by your gastroenterologist or surgeon. You should call their office right away if it is during the day and if it is at night, you should bring your baby to the Dell Children’s Emergency Department for evaluation.

We understand that having a baby with biliary atresia is very stressful but we have taken care of a lot of babies with this problem and can help you. WE ARE HERE TO SUPPORT YOU! The Pediatric Surgery Service can be reached 24/7 at (512) 708-1234. The Pediatric Gastroenterology Service can also be reached at (512) 628-1810 or through MedLink after hours.