Biliary Atresia - Kasai Procedure (hepatoportoenterostomy)

A Guide for Families

What is biliary atresia?

Biliary atresia is a condition in which bile cannot drain from the liver. The cause of biliary atresia is unknown. This condition is also called extrahepatic biliary atresia.

The liver produces a liquid, called bile, which is needed for digestion. Bile drains from the liver and into the intestine via bile ducts (tubes) that are found inside (intrahepatic) and outside (extrahepatic) the liver. In biliary atresia, the extrahepatic bile ducts of the liver are abnormal: they are damaged by a progressive inflammatory process. With time, these bile ducts become so damaged that they become narrow and cannot drain bile from the liver into the intestine. This causes bile to become trapped in the liver, where it accumulates. This accumulation of bile damages the bile ducts inside the liver, ultimately causing injury and scarring of liver tissue. Biliary atresia results in jaundice (yellowing of the skin), cirrhosis and an enlarged liver.

Early surgical intervention to treat biliary atresia is thought to be important to prevent irreversible liver damage.

How is biliary atresia diagnosed?

Biliary atresia can be difficult to diagnose. A combination of diagnostic tests will be used to determine if your child has biliary atresia. These diagnostic tests include:

**Abdominal ultrasound:** An ultrasound provides an image of the liver and associated structures, identifying the anatomy and ruling out other causes of liver disease.

**HIDA (hepatobiliary iminodiacetic acid) scan:** This study provides an image of bile flow from the liver, through the gallbladder and extrahepatic biliary ducts into the intestine.

**Liver biopsy:** A tiny sample of liver tissue is removed with a needle and examined under a microscope by a pathologist, a physician specializing in tissue analysis. The biopsy may be helpful in determining whether surgery is necessary.
**Intraoperative cholangiogram:** This test is scheduled when the diagnosis of biliary atresia is likely. During this procedure, a contrast dye is injected into the gallbladder while the flow of dye is monitored. If there is no flow of contrast into the extrahepatic biliary ducts, a diagnosis of biliary atresia is made and a Kasai procedure is performed at that time.

**What is the treatment for biliary atresia?**

In order to restore bile flow, the extrahepatic bile ducts must be surgically reconstructed. This is done by an operation commonly referred to as the Kasai procedure. The Kasai procedure involves removing the blocked bile ducts and gallbladder and replacing them with a segment of your child’s own small intestine. This segment of intestine is sewn to the liver and functions as a new extrahepatic bile duct system.

The operation will be performed by a Pediatric Surgeon who has had special training in the management of surgically correctable problems in children. The surgeon may be able to operate through many small incisions (laparoscopic surgery) instead of one large (open surgery) incision. The surgeon will determine the safest method of operating and will discuss this with you before the procedure takes place. The operation will take approximately 4 hours to complete.

After the operation, your child will be cared for in the recovery area, and you can be with him or her while he or she is waking up. From the recovery room your child will be transferred to the pediatric floor and will stay in the hospital for several days. Once your child is able to eat well, has no fever, and is comfortable on pain medication by mouth, he or she will be discharged home.

**What are the possible outcomes for patients after a Kasai procedure?**

Normal restoration of bile flow and recovery of liver function occurs in approximately one third of children who undergo the Kasai procedure. These children may not require liver transplantation. The remaining two thirds of children who undergo the Kasai procedure will not have adequate bile flow and liver function, and will eventually require liver transplantation. Of this group, half will need transplantation soon after the Kasai procedure and half will need transplantation at a later time.

**How do I take care of my child at home after discharge from the hospital?**

**Pain:** Most children only need Motrin® for relief of pain once at home. These medications should be taken only if needed and are given by mouth every four to six hours. If your child is still uncomfortable, please call our office.

**Dressings:** Your child can bathe with the Steri-Strips™ on. These will fall off on their own in a few days to a few weeks. There are no stitches to be removed, they are under the skin and will dissolve with time.

**Swelling:** There will be some swelling at the incision. You will be able to feel a firm ridge under the incision that lasts several months. This is called a “healing ridge” and will go away with time.

**Bathing:** Your child should bathe regularly once at home.

**Activity:** There are no activity restrictions.

**Diet:** Special formula may be required for your child. In addition, fat soluble vitamins A, D, E & K, which are important to your child’s health, may be prescribed.

**Bowel Movements:** Your child’s stool will regain color with return of bile flow.
Do I need to see the surgeon again after the operation?

One or two weeks after you arrive home from the operation a nurse from our office will call you to see how your child is doing. Please call our office if you have any questions or concerns after the surgery.

Your child will need an appointment with the Gastroenterologist and our surgeon two or three weeks after discharge from the hospital.

When do I call your office?

Call our office at (415) 476-2538 for the following:

• Fever can be a sign of cholangitis, an infection in the bile ducts and liver. Infection of the liver can lead to decreased bile drainage and liver damage. Although this occurrence is rare, prompt treatment is necessary. If you suspect such and infection, call your child’s primary provider, or bring your child to the nearest emergency room.

• Signs of infection at the incision site that you should report to your pediatrician or pediatric surgeon are: redness, swelling, pain, tenderness and drainage from the incision.

• If your child is experiencing increased pain or vomiting, please call our office for advice.

• Any concerns you have about your child’s recovery.

Pediatric Liver Disease Research Study at UCSF

Dr. Philip Rosenthal from the Department of Pediatrics and Dr. Hanmin Lee from the Department of Pediatric Surgery at the University of California, San Francisco and colleagues at eight other universities are conducting a study to learn more about children born with liver disease. These diseases include neonatal hepatitis and biliary atresia. Because little is known about what causes these diseases or their long-term effects, you may be asked to participate in this study if your infant has a liver problem that involves blockage of bile flow. You may also be approached about participating in a separate clinical trial for biliary atresia. Participation in this study includes, but is not limited to, sharing your child’s medical information, family medical history, and collection of blood, urine, and liver tissue samples. Your surgeon can discuss the details of this research study with you.

National Resources for Patients and Families

The American Liver Foundation
800-GO-Liver (800-465-4837) toll-free
http://www.liverfoundation.org

The American Liver Foundation recognizes that parents of children with biliary atresia need help in coping with the immense strain of this chronic illness. This organization coordinates mutual help groups through its chapters to provide emotional support for families and keeping people aware of the latest research developments. Information on local chapters can be found on this foundation’s website.
The Children’s Liver Association for Support Services is an all-volunteer, nonprofit organization dedicated to serving the emotional, educational, and financial needs of families coping with childhood liver disease and transplantation.

**GLOSSARY OF TERMS:**

**Bile:** a liquid produced by the liver which flows through the bile ducts into the intestine. Once in the intestine, bile mixes with food to help digest and absorb fats.

**Bile Ducts:** A system of interconnected, small tubes that carry bile out of the liver and into the intestine and the gallbladder. In biliary atresia, these ducts become so inflamed that they can close off, blocking the flow of bile. Bile ducts are found inside and outside of the liver.

**Intrahepatic Ducts:** Smaller bile ducts that form a tree-like structure within the liver. Their function is to carry bile from where it is produced in the liver to the larger ducts that lead outside of the liver.

**Extrahepatic Ducts:** Larger bile ducts that are located on the outside of the liver and drain bile from the intrahepatic ducts. The extrahepatic ducts carry bile to the gallbladder for storage and to the intestine for digestion and absorption of fats.

**Gallbladder:** Small sac-like structure that stores bile until it is needed for digestion. The gallbladder is connected to the liver and intestine through the extrahepatic bile ducts.

**Cirrhosis:** Liver damage caused by the accumulation of bile in the liver. This accumulation of bile can result from a variety of conditions, one of which is biliary atresia. Early surgical intervention is performed for biliary atresia in an effort to prevent cirrhosis from developing.

**Kasai Procedure:** A surgical technique developed in 1959. This procedure is used for treating biliary atresia. It involves removing the blocked bile ducts and replacing them with a portion of the child's own intestine. The intestine is sewn directly to the liver, acting as a new duct allowing bile to flow directly into the intestine.