GI Kids
Help & Hope for Children with Digestive Disorders
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Biliary Atresia and Liver Transplant

What is Biliary Atresia?

Biliary Atresia (BA) is an inflammatory process of unknown cause that affects the bile ducts (the tubes that carry digestive juices from the liver to the intestine. Bile ducts are part of the "plumbing system" (or the network of tubes and passageways) that carry bile from the liver to the small intestine to help us to digest fats in our food (see figure). Biliary atresia means that bile and other compounds made by the liver (such as bilirubin, a breakdown product of red blood cells) can't go into the intestine, and build up in the liver, causing liver damage.

What causes Biliary Atresia?

Nobody knows yet why BA happens. It is most likely related to genetic and environmental influences, but we don't know enough about this yet. Lots of research is happening to try to find more answers.

How common is Biliary Atresia?

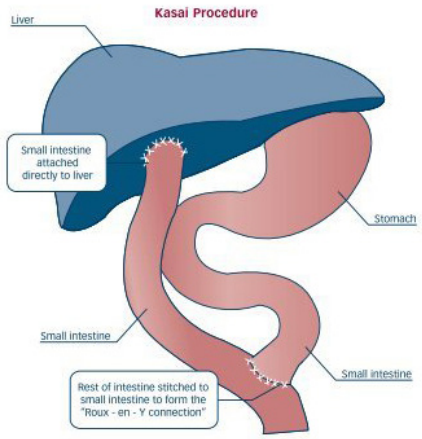
BA occurs in one in 8,000-20,000 live births. It is the most common reason for young children needing liver transplantation. BA most commonly occurs as an isolated problem, meaning that the infant just has this problem and no others. Most children who get BA are full term and normal size at birth.

In approximately 15% of children with BA, BA occurs in association with other major birth defects. These may include congenital heart disease, intestinal malrotation (abnormal position of the intestine), polysplenia (more than one spleen), no spleen, midline liver, or situs inversus (where the liver and spleen are in the wrong positions).

What are the symptoms and signs of Biliary Atresia?

Common signs of BA are:

- High "direct" or "conjugated" bilirubin on blood tests. Bilirubin is a breakdown product of red blood cells that causes jaundice.



- jaundice (yellowing of the skin and eyes)
- dark, tea-colored urine
- pale, clay colored stool (that does not contain brown, yellow or green color)

How is Biliary Atresia diagnosed?

It is important to make the diagnosis of BA early (ideally before 60 days of life). This is because the long-term outcome depends on the age when the first surgery to treat BA is performed. Therefore, any newborn older than two weeks with jaundice or abnormal bilirubin levels on blood tests should be evaluated. If so, other tests are likely to be done. These may include:

- **Abdominal Ultrasound:** A painless way to look at the liver, gallbladder and surrounding organs using sound waves. Ultrasound can assess liver size, and also look for other abnormalities that may cause jaundice. Ultrasound is not reliable in either definitively diagnosing or excluding biliary atresia.
- **Hepatobiliary (HIDA) Scan:** This test helps to see if the bile is able to flow from the liver to the small intestine. A small amount of radioactive dye is injected through a vein. If the dye is seen in the intestines, then that usually means the drainage

system is open and BA is not present. If no dye gets into the intestine, further studies are needed to evaluate for biliary atresia.

- **Liver Biopsy:** The child is given an anesthetic and a tiny incision is made over the lower part of the ribs on the right-side. A small needle is then passed through that incision. A tiny piece of tissue is taken from the liver to look at under the microscope. Doctors can look for features on the liver tissues that suggest BA.
- **Exploratory Laparotomy with intraoperative cholangiogram:** The diagnosis of BA is confirmed at surgery. The surgeon can directly inspect the biliary tree (drainage system) and inject dye to see if there is a blockage. The process of injecting dye into the biliary tree by the surgeon is called an intraoperative cholangiogram. The surgeon may also take a sample of tissue from the liver (wedge liver biopsy)

How is Biliary Atresia treated?

Surgery is the only treatment for BA. The operation is called a hepatoportoenterostomy or "Kasai" procedure. In this operation, the non functioning drainage system is removed. A piece of intestine is then directly connected to the liver, so that bile can flow directly from the intestine into the liver. This procedure is only successful approximately 50% of the time.

When this operation is successful the jaundice disappears and the bilirubin levels (brownish yellow substance found in bile) return to normal. As bilirubin leaves the body, it gives stool its normal brown color.

The operation has the best chance for success when done as early as possible. However, even with early surgery, many infants with BA still develop liver cirrhosis (permanent scarring of the liver). These children, at some point, are likely to require liver transplantation.

What should you expect after the hepatoportoenterostomy (Kasai procedure)?

Children with BA in the first year of life take a number of medicines and special nutrition. After infancy, many of these therapies are stopped if there is no jaundice and the child is growing well.

MEDICINES:

- Antibiotics are given to prevent infection from going up into the liver (ascending cholangitis).
- Ursodeoxycholic acid (also called Actigall and ursodiol), a medication that helps bile flow and help protect the liver.
- Vitamin supplements such as vitamins A, D, E and K are given as well.
- Steroids are sometimes given after surgery to help decrease inflammation.

NUTRITION AND GROWTH

- For all children with BA, nutrition and growth is the most important part of long-term care by the medical specialist.
- Because nutrients might be difficult to absorb without a gallbladder and particularly if jaundice is present, special pre-digested formulas are used.
- Regular follow up visits are important to make sure the child is growing appropriately.

LIVER TRANSPLANTATION

For children who need liver transplantation, the cirrhotic (scarred) liver is removed and a new liver from a donor is placed surgically. The donor can be a deceased donor or living related, meaning a parent, family member or friend can donate a piece of his or her own liver. Survival of children with BA after liver transplantation is excellent, with 90% surviving after 10 years of age. (see liver transplantation handout).

Useful Links and Resources

American Liver Foundation Biliary Atresia Page:
https://www.liverfoundation.org/for-patients/about-the-liver/diseases-of-the-liver/biliary-atresia/

National Digestive Diseases Information Clearinghouse Biliary Atresia Page:
https://www.niddk.nih.gov/health-information/liver-disease/biliary-atresia

Biliary Atresia Resources Available in the App Store

LearnAboutBA is a useful learning and teaching app for parents, patients and health-care providers.

Quick Facts

- Biliary Atresia is a birth defect causing blockage of the biliary tree – the plumbing that lets bile go from the liver to the intestines
- It occurs in 1 in every 8-20'000 live births
- The exact cause is not known but likely includes genetic and environmental factors
- There are blood tests, scans and surgical procedures that can help diagnose BA
- The treatment includes surgery called "hepatoportoenterostomy" which should be done as early as possible
- After the surgery, medications and sometimes special formulas are used to protect against infections and worsening liver problems, and to make sure the child grows well
- Often, liver transplantation is needed at some point as the liver can still become scarred and not work well enough

Locate a Pediatric Gastroenterologist

NASPGHAN
North American Society for Pediatric Gastroenterology, Hepatology and Nutrition

714 N Bethlehem Pike, Suite 300, Ambler, PA 19002 **Phone:** 215-641-9800 **Fax:** 215-641-1995 **naspghan.org**
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