

# Biliary Atresia

## What is Biliary Atresia?

Biliary Atresia (BA) is an inflammatory process of unknown cause affecting the bile ducts. It results in blockage of the drainage system (biliary tree) from the liver to the intestines.

## How common is Biliary Atresia?

BA occurs in one in 8,000-20,000 live births. It is the leading indication for liver transplantation in children. BA most commonly occurs as an isolated problem. Most children who get BA are fullterm and normal size at birth. Common signs of BA are jaundice (yellowing of the skin and eyes), dark tea colored urine, and pale clay colored stool (non-pigmented stool).

For some (15-30%), BA occurs with a number of 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). Depending on the associated birth defects, other terms may be used such as: embryonic BA or BA Splenic Malformation Syndrome or heterotaxy.

### SPECIFIC INSTRUCTIONS:

## How is Biliary Atresia diagnosed?

It is important to make the diagnosis of BA early. This is because the long-term outcome depends on the age of initial surgical intervention (Kasai procedure). Therefore any newborn older than two weeks with jaundice should have blood tests to see if there is a liver problem. 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.

**HIDA Scan:** This test helps to determine whether 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 the drainage system is open and BA is not present. If no dye gets into the intestine, further studies are needed to confirm the diagnosis of BA

**Liver Biopsy:** The child is given an anesthetic and a tiny cut is made over the lower part of the ribs on the right-side. A small needle is then passed through that cut. A tiny piece of tissue is taken from the liver to look at under the microscope.

**Explorative Laparotomy:** 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 surgeon may also take a sample of tissue from the liver.



## 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 damaged drainage system is removed and the intestine is connected to the liver with the hope that bile flows again. 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.

# Biliary Atresia continued

## What should you expect after the hepatoporoenterostomy?

Children with BA in the first year of life take a number of medicines and special nutritionals. Antibiotics are given to prevent infection from going up into the liver (ascending cholangitis). Because nutrients might be difficult to absorb without a gallbladder and particularly if jaundice is present, special predigested formulas are used. Vitamin supplements for vitamins A, D, E and K are given as well. Steroids may also be given after surgery to help decrease inflammation. Ursodeoxycholic acid, a good bile acid, is given to help bile flow and help protect the liver. After infancy, many of these therapies are stopped if there is no jaundice and the child is growing well. For all children with BA, nutrition and growth is the most important part of long-term care by the medical specialist.

## Liver transplantation

For children who need liver transplantation, the cirrhotic 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 or family member 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.

*For further information visit the American Liver Foundation website at [www.liverfoundation.org](http://www.liverfoundation.org) and under "Liver diseases and related topics" view "Biliary Atresia".*

*For more information or to locate a pediatric gastroenterologist, please visit our website at: [www.naspgghan.org](http://www.naspgghan.org)*

**IMPORTANT REMINDER:** This information from the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) is intended only to provide general information and not as a definitive basis for diagnosis or treatment in any particular case. It is very important that you consult your doctor about your specific condition.

