Biliary Atresia

What is Biliary Atresia?

Biliary Atresia (BA) is an inflammatory process of unknown cause from which the bile ducts (tubes that carry bile from the liver to the small intestine) inside and outside the liver are irreversibly blocked.

How Common is Biliary Atresia?

Biliary atresia occurs in one in 10,000 – 15,000 live births. It is the leading cause of chronic liver disease in children and it is the most common reason why children require liver transplantation. BAs is the second most common cause of obstructive jaundice (when the bile duct is blocked) in the first 2 months of life, and is responsible for 25-30% of cases in this age group.

There are two forms of biliary atresia. The more common “classical” form which accounts for 80-90% of cases presents in full term infants after about two weeks of age. Jaundice (yellow skin and eyes), dark tea colored urine, pale clay colored stool (non-pigmented stools), a large liver and spleen are the most common symptoms. The less common “embryonic” form seen in up to 20% of cases usually appears soon after birth with jaundice, dark urine and pale stool in association with other abnormalities that include congenital heart disease, intestinal malrotation (abnormal position of the intestine), polysplenia (more that one spleen) or situs inversus (where the liver and spleen are in the wrong positions).

How is Biliary Atresia Diagnosed?

It is important to make the diagnosis of BA early, preferably before two months of age because the long term outcome depends upon the age of treatment. Therefore any newborn older than two weeks with jaundice should have a blood test to assess whether the jaundice is “obstructive”. If these tests demonstrate obstructive jaundice other blood test will be done to see how well the liver is working. These tests may include:

- **Abdominal ultrasound**: Ultrasound imaging, or sonography, obtains images from inside the body using high frequency sound waves. This test looks at the liver and bile ducts and the sound waves’ echoes are displayed as real-time images allowing the physician to see blood flow.

- **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 bile duct is open and biliary atresia is not present. If no dye gets into the intestine, further studies are needed to help confirm the diagnosis of biliary atresia.

- **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 tiny needle is then passed through that cut and a tiny tissue sample is taken from the liver.

- **Explorative Laparotomy**: The diagnosis of biliary atresia is confirmed at surgery. The surgeon can directly inspect the bile duct and inject dye into the ducts to see whether the ducts are blocked. The surgeon may also take a sample of tissue from the liver.

How is Biliary Atresia Treated?

Surgery is the only treatment for biliary atresia. The operation is called a portoenterostomy or Kasai procedure. In this operation the blocked bile duct is removed and a loop of small intestine is connected to the liver with the hope that the bile will flow again. When this operation is successful the jaundice disappears and the bilirubin levels (brownish yellow substance found in bile when the liver breaks down bilirubin) return to normal. Bilirubin leaves the body as feces and gives stool its normal brown color. The operation has the best chance for success if done before 2 months of age, with up to an 80% chance of the jaundice clearing.

Even with early surgery, many infants with biliary atresia develop increasing scarring of the liver and eventual cirrhosis. These children will require liver transplants once the scarred liver’s function fails. Recent evidence suggests that fewer than half of the children with biliary atresia after portoenterostomy survive into adulthood without requiring a liver transplant.

What Should You Expect After the Portoenterostomy?

The child will be given antibiotics to prevent infection of the bile ducts. He or she will also be given fat soluble vitamins (ADEK) and Ursodeoxycholic acid (a good bile acid that improves the flow of bile and may protect the liver cells).

Nutrition is important and special formulas (predigested formula) are a necessary part of the child’s diet. Steroids (anti-inflammatory medications) are sometimes used.

Liver transplantation

For children who come to liver transplantation, the cirrhotic liver is removed, and a donor liver organ is surgically placed. The donor can be a deceased donor (i.e. has been declared “brain dead”) or living related, meaning a parent or family member can donate a piece of his or her own liver. Survival for children with biliary atresia after liver transplant is very good, with 90% surviving after 10 years of age.

For further information visit the American Liver Foundation website at www.liverfoundation.org and under “Liver Health Information view ‘Pediatric Liver Disease’

For more information or to locate a pediatric gastroenterologist in your area please visit our website at: www.naspghan.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.

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