

# Biliary Atresia

BIRTH DEFECT RESEARCH FOR CHILDREN



## What Is Biliary Atresia?

Biliary atresia is a rare liver disease which destroys the bile ducts necessary to carry bile from the liver to the intestines. This disease involves the bile ducts outside the liver (extrahepatic) and may progress to severely affect the bile ducts inside the liver (intrahepatic). Bile collects within the liver and other tissues where it acts as a slow poison. Without treatment, cirrhosis and eventual liver failure occur. Biliary means pertaining to the bile, bile ducts, and/or gall bladder. Atresia refers to a congenital absence of or closure of a normal body orifice. Children who have biliary atresia do not have a complete functional biliary system.



# Biliary Atresia



## How Common Is Biliary Atresia?

Biliary Atresia occurs in 1 out of 10,000 live births and slightly more often in females than males (1.4:1). It is the most common cause for pediatric liver transplantation.

## What Causes Biliary Atresia?

The cause of biliary atresia is uncertain, although several different theories have been proposed. Researches are studying both Reovirus 3 and Cytomegalovirus as possible viral agents in the development of biliary atresia. Instances of time and space clustering have also suggested that infectious or environmental toxic factors may cause the disease.

## How To Help A Child With Biliary Atresia

**Diagnosis:** Any infant with signs of jaundice beyond 2 weeks of life, combined with acholic (pale or clay-colored) stools, dark urine, enlarged liver, and/or enlarged spleen should have liver function tests (blood work). If these tests are elevated, further evaluation, including an ultrasound and liver biopsy, may be needed. Early diagnosis (by 8-12 weeks of age) is important and will result in better prognosis.

**Treatment:** The Kasai Procedure is an operation that is performed to reestablish bile flow from the liver to the intestine. A section of the small intestine is used to attach the liver directly to the small intestine. If the Kasai Procedure is unsuccessful, liver transplantation is the only option.

## Complications:

Several complications can occur with Biliary Atresia. These complications include:

**cholangitis** - a bacterial infection from the bowel which causes an inflammation of the bile ducts

**portal hypertension** - high blood pressure in the portal vein

**varices** - occurs when veins expand due to increased blood pressure and can rupture

**ascites** - an abnormal build-up of fluid in the abdomen

**failure to thrive** - poor growth

**ammonia intoxication** - altered mental status  
cirrhosis

Fact Sheet by:

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