Biliary Atresia

What is it?

Biliary atresia (pronounced bil-e-air-e ah-tre-zhah) is a rare condition of the **liver** and **bile ducts**. **Bile** is a liquid produced by the liver that helps digest fats and get rid of wastes. **Bile ducts** are channels that move bile from the liver to the small intestine. The network of channels and bile ducts is called the **biliary system**. When the biliary system works normally, it lets the bile drain from the liver into the intestines. But when a baby has **biliary atresia**, bile flow from the liver to the gallbladder is blocked. This means bile is trapped inside the liver. Liver damage, including cirrhosis and liver failure, occurs. **Cirrhosis** is the scarring of the liver cells.

Babies with biliary atresia are usually healthy when they are born. Symptoms usually start between 2 and 8 weeks of age. Symptoms may include:

- **Jaundice**, a yellow color to the skin and eyes due to an increased level of bilirubin. **Bilirubin** is a substance in bile that gives it a yellowish color.
- **Dark urine** due to an increased level of bilirubin in the urine.
- **Clay-colored stools** due to an absence of bilirubin in the intestines.
- **Swollen or firm abdomen** from an enlarged liver.
- **Weight loss**.
- **Irritability**.

Often biliary atresia is a single (isolated) birth defect. A small number of babies may have other problems. These problems may involve the heart, spleen, blood vessels, and intestines.

How common is it?

Biliary atresia is a rare condition. About 1 in 12,000 babies will be born with it each year. It tends to affect girls more than boys.

What causes it?

The exact cause of biliary atresia is not known. In some cases the bile ducts did not form normally during pregnancy. In other cases it may be due to damage to the bile ducts from the immune system caused by an infection after the baby is born. More research is needed to understand all causes of biliary atresia.

How is it diagnosed?

Jaundice is the first sign of a liver condition. However, biliary atresia is not the only liver condition that causes jaundice. Several tests are usually done to diagnose biliary atresia. These tests may include blood tests, X-rays, abdominal ultrasound, liver biopsy, and diagnostic surgery. Surgery is the only way to confirm that a baby has biliary atresia.

How is it treated?

Biliary atresia is usually corrected with surgery. The **Kasai procedure** is a surgery that creates a new, open duct so bile can drain from the liver. After surgery, long-term antibiotic therapy is given to reduce the risk of infection. Other medications may be given to promote bile flow. The surgery is successful about 60% - 85% of the time. This means bile will drain from the liver and the jaundice level will disappear. The surgery is not a cure, but it does allow babies to grow and enjoy fairly good health for several years. Another available treatment option is a **liver transplant**. A transplant, however, is done only if the surgery does not work. Your child's doctors will discuss appropriate treatment options with you.

Indiana State Department of Health

BDPR
Indiana Birth Defects and Problems Registry

For more information:

National Institute of Diabetes and Digestive and Kidney Diseases
https://www.niddk.nih.gov/health-information/liver-disease/biliary-atresia

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