About Biliary Atresia

What is biliary atresia?

Biliary atresia is a rare disease of the liver and bile ducts that occurs in infants. Symptoms of the disease appear or develop about two to eight weeks after birth. Cells within the liver produce liquid called bile. Bile helps to digest fat. It also carries waste products from the liver to the intestines for excretion.

This network of channels and ducts is called the biliary system. When the biliary system is working the way it should, it lets the bile drain from the liver into the intestines. When a baby has biliary atresia, bile flow from the liver to the intestine is blocked. This causes the bile to be trapped inside the liver, quickly causing jaundice, damage and scarring of the liver cells (cirrhosis), and finally liver failure.

What causes biliary atresia?

The cause of biliary atresia is not known. For some children, biliary atresia may occur because the bile ducts did not form properly during pregnancy. For other children with biliary atresia, the bile ducts may be damaged by the body's immune system in response to a viral infection acquired after birth.

What is known for certain is that biliary atresia affects only newborns; it is not hereditary; it is not contagious; and it is not preventable.

Who is at risk for biliary atresia?

Although it is relatively rare, biliary atresia occurs once in every 15,000 births and is an irreversible problem that is fatal without treatment. Biliary atresia is the most common liver disease in children that requires transplantation.

It occurs slightly more often in females than in males (1.4:1), and affects children of all races. In the United States, approximately 300 new cases are diagnosed each year. Biliary atresia is not caused by anything the mother did during pregnancy.

Symptoms of biliary atresia

Babies with biliary atresia usually appear healthy when they are born. Symptoms usually begin to appear between two and eight weeks after birth, and include:

- Jaundice (a yellow appearance of the skin and whites of the eyes) that does not improve within one to two weeks
- Dark yellow or brown urine due to excessive bilirubin in the bloodstream that passes to the kidneys
- Pale or clay-colored (acholic) stools, an indication that very little or no bile (which gives bowel movements their normal color) is reaching the intestine
- Enlarged liver that feels harder than normal, enlarged spleen
- Poor weight gain

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