Biliary Atresia

What is biliary atresia?
Biliary atresia is a rare disease of the liver and bile ducts that occurs in infants. Normally, cells within the liver produce bile (a liquid that helps digest fat and carries waste products to the intestines). The biliary system (a network of channels that carry bile) lets the bile drain from the liver into the intestines. When a baby has biliary atresia, bile flow is blocked, which causes the bile to be trapped inside the liver. This buildup causes liver damage, including cirrhosis (scarring of the liver cells) and liver failure.

What are the symptoms of biliary atresia?
Babies with biliary atresia usually appear healthy when they are born. Symptoms of biliary atresia usually develop between 2 – 8 weeks of age and may include:

- Jaundice (yellow color to the skin and eyes) due to an increased level of bilirubin (substance that gives bile a yellowish color)
- Dark urine due to an increased level of bilirubin in the urine
- Acholic (clay-colored) stools due to an absence of bilirubin in the intestines
- Weight loss
- Irritability

What causes biliary atresia?
The cause of biliary atresia is not known. Possible explanations include abnormal formation of the bile ducts during pregnancy, viral infections, or by damage to the bile ducts caused by the immune system after the baby is born. Biliary atresia is most often an isolated condition (no other birth defects are associated). However, approximately 10 – 15% of infants with biliary atresia have other birth defects, including cardiac (heart) defects or problems with the spleen, blood vessels, or intestines.

How is biliary atresia treated?
Biliary atresia is usually corrected with surgery. The Kasai procedure creates a new, open duct so bile can drain from the liver. Another available treatment option is a liver transplant; however, a transplant is only done if no other treatments are available. Your child’s doctor(s) will discuss appropriate treatment options with you.

For more information
Cincinnati Children’s Hospital Medical Center - http://www.cincinnatichildrens.org/svc/alpha/l/liver/diseases/biliary.htm

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