Biliary Atresia



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Cholestasis
Coal-stop-sign
Cholestasis is a common presenting symptom of liver disease in infants. Chief cause among diseases presenting as neonatal cholestasis is biliary atresia, the most common cause of chronic liver disease in children, but little is known about the pathogenesis of this disease.

Unknown Etiology
Question-mark
The cause of biliary atresia is unknown, although several mechanisms have been implicated. Genetical and viral causes might play a role.

Symptoms

Persistent Jaundice
Jaundice-janitor
Jaundice may be seen only in the sclerae and is the first sign of biliary atresia. The onset of jaundice occurs any time from birth up to eight weeks of age, and it is highly unlikely to appear later.

Acholic Stools
Pale Stool
Some infants have acholic stools. Acholic stools often go unrecognized because the stools are pale but not white, and the stool color can vary on a daily basis.

Hepatomegaly
Liver-balloon
Progressive disease and jaundice going unnoticed can lead to an enlarged liver and spleen.

Dark Urine
Dark Urine
Because of bilirubin excretion into the urine, most infants may have dark urine. Parents or caregivers may not recognize that dark yellow urine or a yellow-stained diaper is abnormal in an infant.

Diagnosis

Lab Values
Laboratory Test-tubes
Infants with symptoms of biliary atresia can reveal elevations in direct and/or conjugated bilirubin and mild or moderate elevations in serum aminotransferases, with a disproportionately increased gamma-glutamyl transpeptidase (GGTP).

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Ultrasound

Ultrasound-machine

The suspicion of biliary atresia is strengthened by the results of a variety of tests, typically an ultrasound.

Treatment

Kasai Procedure

Karate Procedure

After confirmation of the diagnosis, a Kasai procedure (hepatoportoenterostomy [HPE]) should be performed promptly.
This operation is undertaken in an attempt to restore bile flow from the liver to the proximal small bowel. For this procedure, a Roux-en-Y loop of the bowel is created by the surgeon and directly anastomosed to the hilum of the liver, following the excision of the biliary remnant and portal fibrous plate.

Liver Transplantation

Liver Train-plant

The majority of individuals with biliary atresia eventually require liver transplantation. Biliary atresia is the most common indication for liver transplantation in infants and children.

Complications

Cirrhosis

C-roses-liver

If undetected, early biliary liver cirrhosis (from about the 10th week of life) children can die in the first 2 years.

Portal Hypertension

Portal Hiker-BP

The chronic hepatobiliary inflammation characteristic of biliary atresia leads to progressive biliary cirrhosis. Biliary cirrhosis causes portal hypertension, which can lead to esophageal varices and ascites.

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