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Perspective

Biliary Atresia: Symptoms and Therapy

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DESCRIPTION

Biliary atresia is a blockage inside the tubes (ducts) that deliver bile from the liver to the gallbladder. This congenital situation takes place while the bile ducts inside or outside of the liver doesnot expand or dilate normally. It isn't known why the biliary machine fails to broaden normally. In infants with biliary atresia, bile flow from the liver to the gallbladder is blocked. This can cause liver damage and cirrhosis of the liver that is lethal if now no longer treated. Biliary atresia is a rare disease of the liver and bile ducts that occurs in infants. Symptoms of this disease appear or develop about 2-8 weeks after birth. The cells in the liver produce fluid called bile. Bile helps digest fat. It also carries waste products from the liver to the intestines and removes them from the body. This tube and network of tubes is called the biliary system. When the bile system is functioning normally, it drains bile from the liver into the intestines.

SYMPTOMS

Newborns in this condition may look normal at birth. However, jaundice develops in the second or third week of life. Infants may gain weight normally during the first month, but then begin to lose weight, become frustrated, and have worsening jaundice.

- Other symptoms include:
- Dark urine
- Splenomegaly
- Floating stool
- Swollen stomach
- Light or clay-colored stool
- Slow or no weight gain
- Slow growth

DIAGNOSIS

- Physical examination is done to look for an enlarged liver. Other tests include:
- Abdominal ultrasonography.
- Blood test to check total and direct bilirubin levels.
- Hepatobiliary Imininodiacetic Acid (HIDA) scan (also called biliary scintigraphy), bile duct.
- Liver biopsy to determine if the gallbladder is functioning properly.
- Bile duct ultrasonography to determine the severity of cirrhosis or to rule out other causes of jaundice.

TREATMENT

A blood test is done to determine if there is an abnormality in liver function. You can also identify the cause of jaundice. X-rays of the abdomen look for enlargement of the liver and spleen. Abdominal ultrasonography can determine if there is a small gallbladder or no gallbladder at all. The gallbladder is an organ that stores bile. If this organ is missing or has not been present since birth, it often indicates biliary atresia. A liver biopsy shows whether the baby is more likely to have biliary atresia. A liver biopsy takes a small sample of the liver with a needle. Then observe this sample under a microscope. Liver biopsy is very reliable. If a biopsy shows that the baby is likely to have biliary atresia, additional surgery will be done to confirm the diagnosis and treat the condition. Diagnostic surgery checks to see if the child has biliary atresia. Through surgery, doctors can determine if part of the bile duct, from the liver to the intestines, is damaged. This can interfere with the normal flow of bile from the liver. During surgery, surgical cholangiography is performed to confirm the diagnosis of biliary atresia. Cholangiography is a procedure performed during surgery. In this procedure, the dye is injected through the gallbladder and through the bile ducts. Take an x-ray to see if the dye is flowing normally to the intestines and liver. In infants with biliary atresia, the ducts are

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blocked and the pigment usually does not drain from the gallbladder. Biliary atresia is excluded if the duct is normal or patented and the pigment flows normally. A larger liver biopsy (tissue sample) is then done to identify the cause of the liver disease. Biliary atresia is diagnosed when a biliary angiography shows that the bile duct is not patent. After that, the baby usually undergoes surgery called Kasai surgery.