

## **Etiology and Pathophysiology**

The exact cause of BA is unknown, although immune- or infection-mediated mechanisms may be responsible for the progressive process that results in complete obliteration of the bile ducts. BA is not seen in fetuses or stillborn or newborn infants. This suggests that BA is acquired late in gestation or in the perinatal period and is manifested a few weeks after birth. The majority of cases of BA (85%) have a complete obliteration of the extrahepatic biliary tree at or above the porta hepatis ([Hassan and Balistreri, 2016](#)).

Many infants with BA are full term and appear healthy at birth. If jaundice persists beyond 2 weeks old, especially if the direct (conjugated) serum bilirubin is elevated, the nurse should suspect BA. The urine may be dark, and the stools often become progressively acholic or gray, indicating absence of bile pigment. Hepatomegaly is present early in the course of the disease, and the liver is firm on palpation.

## **Diagnostic Evaluation**

Early diagnosis is critical to the child with EHBA; the outcome in children surgically treated before 2 months old is much better than in patients with delayed treatment. The diagnosis of BA is suspected on the basis of the history, physical findings, and laboratory studies. Laboratory tests include a CBC, electrolytes, bilirubin levels, and liver function studies. Additional laboratory analyses, including  $\alpha_1$ -antitrypsin level, TORCH titers and other intrauterine infections (see [Maternal Infections, Chapter 8](#)), hepatitis serology, and urine CMV may be indicated to rule out other conditions that cause persistent cholestasis and jaundice. Abdominal ultrasonography allows inspection of the liver and biliary system. The patency of the extrahepatic biliary system will be demonstrated by a nuclear scintiscan using technetium-99m iminodiacetic acid ( $^{99m}\text{Tc}$  IDA) or hepatobiliary iminodiacetic acid (HIDA) scan. If there is no evidence of radioactive material excreted into the duodenum, BA is the most probable diagnosis. Because the nuclear scan may take up to 5 days for the results, a percutaneous liver biopsy is probably the most useful method of diagnosing BA.