

progressive inflammatory process that causes both intrahepatic and extrahepatic bile duct fibrosis, resulting in eventual ductal obstruction. The incidence of BA is approximately 1 in 10,000 to 15,000 live births ([Hassan and Balistreri, 2016](#)). Associated malformations include polysplenia, intestinal atresia, and malrotation of the intestine. BA, if untreated, usually leads to cirrhosis, liver failure, and death ([Box 22-6](#)).

Box 22-6

Clinical Manifestations of Extrahepatic Biliary Atresia

Jaundice

- Earliest manifestation and most striking feature of disorder
- First observed in sclera
- Usually not apparent until 2 to 3 weeks old after resolution of neonatal jaundice

Dark yellow urine

Stools lighter than expected or white or tan

Hepatomegaly and abdominal distention common

Splenomegaly occurs later

Poor fat metabolism results in:

- Poor weight gain
- Failure to thrive

Pruritus