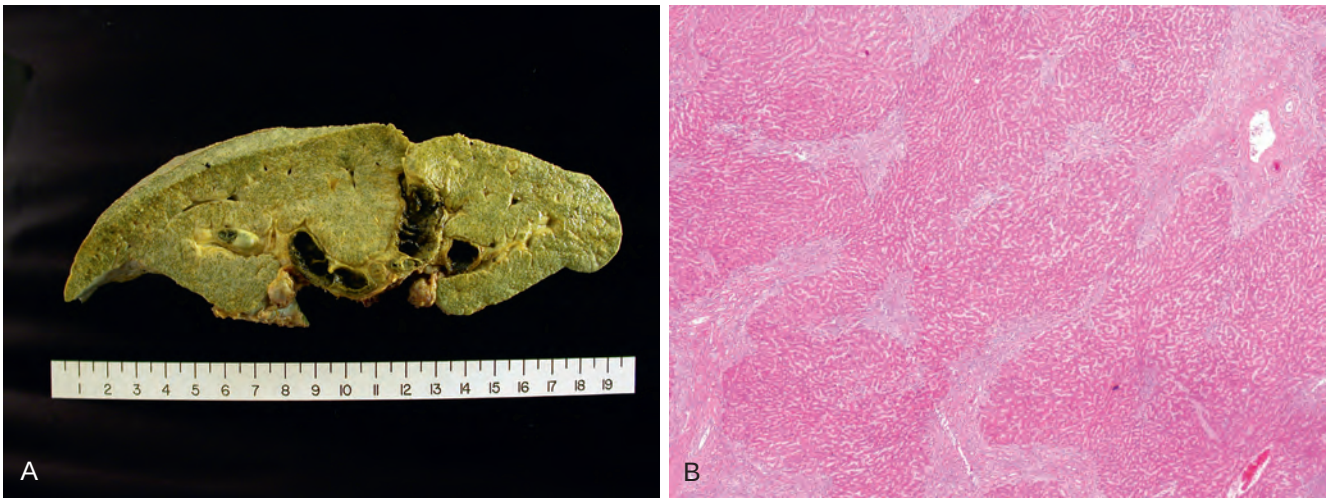

Figure 18-31 Biliary cirrhosis. **A,** Sagittal section through the liver demonstrates the nodularity (most prominent at the right) and bile staining of end-stage biliary cirrhosis. **B,** Unlike other forms of cirrhosis, nodules of liver cells in biliary cirrhosis are often not round but irregular, like jigsaw puzzle shapes.

pyogenic cholangitis focusing on its most common clinical findings and *oriental cholangitis* based on its ethnic predilection, but the underlying disease is one of stone formation, thus the currently accepted name, *hepatolithiasis.* There are regional differences regarding the composition of stones, but the consequences are largely the same.

MORPHOLOGY

Hepatolithiasis has **pigmented calcium bilirubinate stones** in distended intrahepatic bile ducts (Fig. 18-33). The ducts show chronic inflammation, mural fibrosis, and peribiliary gland hyperplasia, all in the absence of extrahepatic duct obstruction. Biliary dysplasia may be seen and may evolve to invasive cholangiocarcinoma.

Clinical Features. Individuals may present with repeated episodes of cholangitis due to secondary infections of the involved ducts, marked by fever and abdominal pain. Due to repeated rounds of inflammation, parenchymal collapse and scarring, the disease sometimes presents with a mass-like lesion mistaken for malignancy and is thus sometimes diagnosed at resection. This disease seems to increase the risk of cholangiocarcinoma by unknown mechanisms.

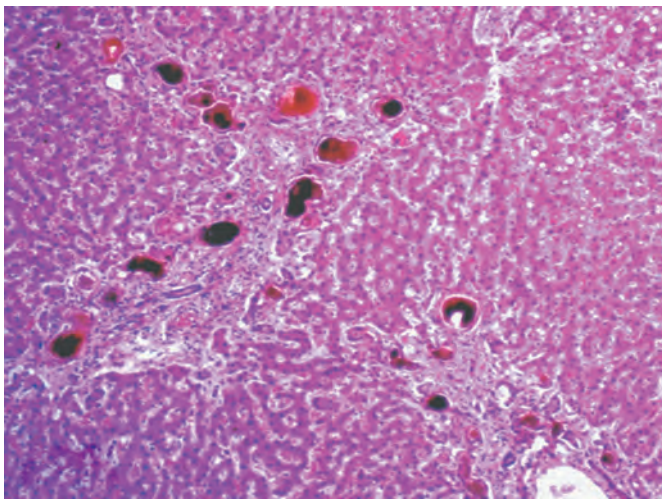

Figure 18-32 Ductular cholestasis of sepsis. Large, dark, bile concretions within markedly dilated canals of Hering and ductules at the portal-parenchymal interface. (Courtesy Dr. Jay Lefkowitch, Columbia University College of Physicians and Surgeons, New York.)

Neonatal Cholestasis

Prolonged conjugated hyperbilirubinemia in the neonate, termed neonatal cholestasis affects approximately 1 in 2500 live births. Since physiologic jaundice of the new born abates by two weeks, infants who have jaundice beyond 14-21 days after birth should be evaluated for neonatal cholestasis. The major causes are (1) cholangiopathies, primarily *biliary atresia* (discussed later), and (2) a variety of disorders causing conjugated hyperbilirubinemia in the neonate, collectively referred to as *neonatal hepatitis.*

Neonatal hepatitis is not a specific entity, nor are the disorders necessarily inflammatory. Instead, the finding of

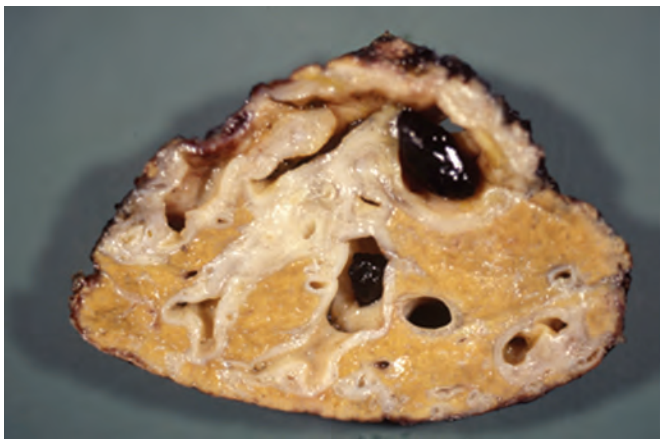

Figure 18-33 Hepatolithiasis. A resected, atrophic right hepatic lobe with characteristic findings including markedly dilated and distorted bile ducts containing large pigment stones and broad areas of collapsed liver parenchyma. (Courtesy Dr. Wilson M.S. Tsui, Caritas Medical Centre, Hong Kong.)