leads to repeated bouts of ascending cholangitis and inflammatory parenchymal destruction. It predisposes to cholangiocarcinoma.

- **Neonatal cholestasis** is not a specific entity; it is variously associated with cholangiopathies such as primarily *biliary atresia* and a variety of inherited or acquired disorders causing conjugated hyperbilirubinemia in the neonate, collectively referred to as *neonatal hepatitis*.
- **Primary biliary cirrhosis** is an autoimmune disease with progressive, inflammatory, often granulomatous, destruction of small to medium sized intrahepatic bile ducts.
  - Primary biliary cirrhosis occurs most often in middle aged women, is associated with autoantibodies (particularly AMA), and with other autoimmune diseases such as Sjögren syndrome and Hashimoto thyroiditis.
- **Primary sclerosing cholangitis** is an autoimmune disease with progressive inflammatory and sclerosing destruction of bile ducts of all sizes, intrahepatic and extrahepatic; diagnosis is made not by biopsy, but by radiologic imaging of the biliary tree. It occurs most often in younger men and has strong associations with inflammatory bowel disease, particularly ulcerative colitis.

Structural Anomalies of the Biliary Tree

Choledochal Cysts

Choledochal cysts are congenital dilations of the common bile duct. They present most often in children before age 10 as jaundice and/or recurrent abdominal pain, symptoms that are typical of biliary colic. Approximately 20% of cases become symptomatic only in adulthood. In some cases choledochal cysts occur in conjunction with cystic dilation of the intrahepatic biliary tree (Caroli disease, discussed later). The female-to-male ratio is 3:1 to 4:1. These uncommon cysts may take the form of segmental or cylindric dilation of the common bile duct, diverticula of the extrahepatic ducts, or choledochoceles, which are cystic lesions that protrude into the duodenal lumen. Choledochal cysts predispose to stone formation, stenosis and stricture, pancreatitis, and obstructive biliary complications within the liver. In older patients the risk of bile duct carcinoma is elevated.

Fibropolycystic Disease

Fibropolycystic disease of the liver is a heterogeneous group of lesions in which the primary abnormalities are congenital malformations of the biliary tree. Lesions may be found incidentally during radiographic studies, surgery, or at autopsy. The most severe forms of fibropolycystic disease may become manifest as hepatosplenomegaly or portal hypertension in the absence of hepatic dysfunction, starting in late childhood or adolescence. Three sets of pathologic findings may be seen, sometimes overlapping with each other:

- *Von Meyenburg complexes* are small bile duct hamartomas (Fig. 18-40). Occasional von Meyenburg complexes are common in otherwise normal individuals. When they are diffuse they signal the underlying, more clinically important fibropolycystic disease.
- *Single or multiple, intrahepatic or extrahepatic biliary cysts.* When present in isolation these may be symptomatic due to ascending cholangitis and are referred to as *Caroli disease*. When biliary cysts occur along with congenital hepatic fibrosis, the term *Caroli syndrome* is used (Fig. 18-41). Ducts may be cystically dilated, but true cysts are also present. These may be intrahepatic cysts or choledochal cysts, as already described.
- In *congenital hepatic fibrosis,* portal tracts are enlarged by irregular, broad bands of collagenous tissue, forming septa that divide the liver into irregular islands. Variable numbers of abnormally shaped bile ducts are embedded in the fibrous tissue, although they remain in continuity with the biliary tree (Fig. 18-42). Individuals

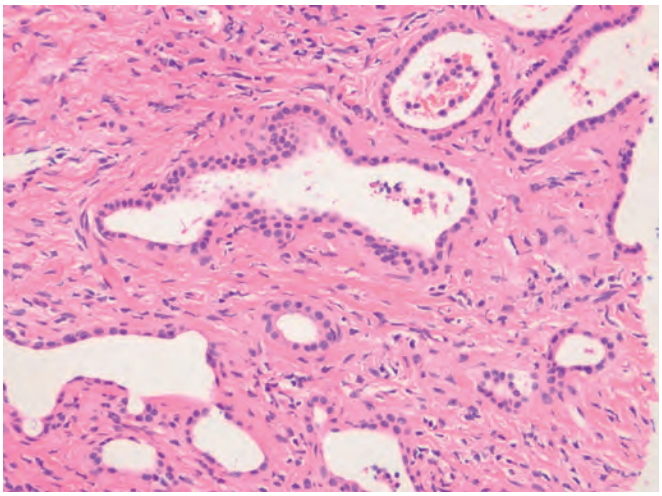

Figure 18-40 Von Meyenburg complex. This bile duct hamartoma is always associated with portal tracts. Note the dilated and irregularly shaped bile ducts.

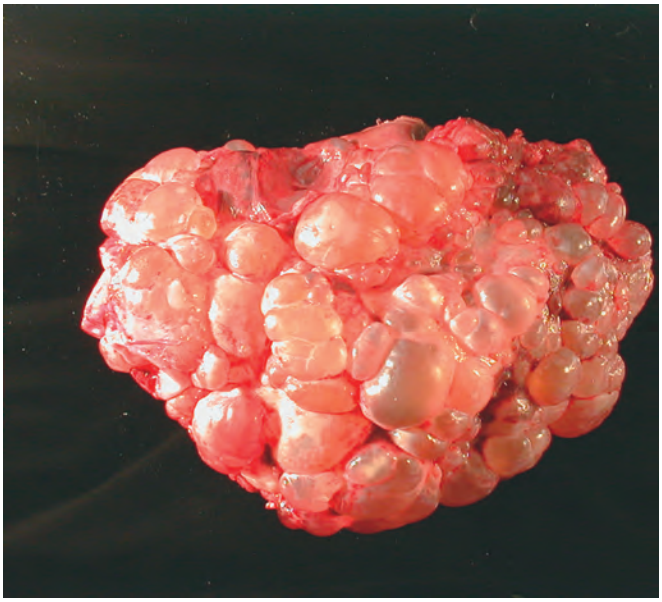

Figure 18-41 Congenital hepatic fibrosis with multiple biliary cysts.