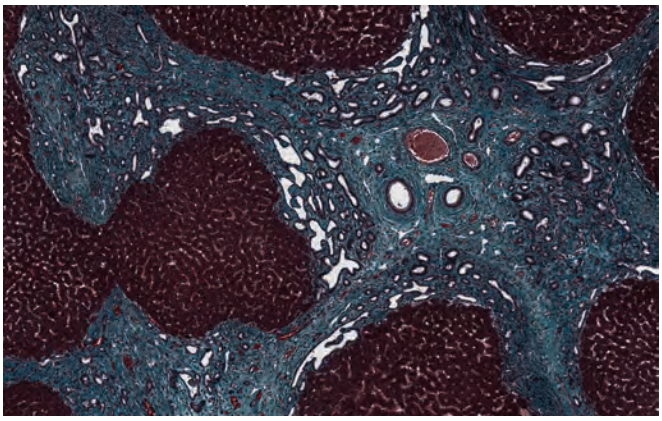

Figure 18-42 Congenital hepatic fibrosis. Broad bands of stroma are seen coursing through this liver, which is involved by a variant of fibropolycystic disease. Note the dilated remnants of ductal plates along the margins of the stroma. The intervening parenchyma is comprised of relatively normal parenchyma (Masson Trichrome stain).

with congenital hepatic fibrosis are not truly cirrhotic, despite the serpiginous scarring separating the hepatic parenchyma, but they may still face complications of portal hypertension, particularly bleeding varices.

All of these lesions are related to abnormal development of the biliary tree representing *ductal plate malformations* associated with persistence of the periportal ductal plates from fetal development. The caliber of involved portal tracts determines the different size, morphology, and distributions of lesions. Fibropolycystic liver disease often occurs with autosomal recessive polycystic renal disease. The involved gene encodes a protein called polycystin which is expressed in fetal kidney as well as liver (Chapter 20). Persons with fibropolycystic liver disease have an increased risk for cholangiocarcinoma.

Circulatory Disorders

Given the enormous flow of blood through the liver, it is not surprising that circulatory disturbances have considerable impact on the liver. In most instances, however, clinically significant liver function abnormalities do not develop, but hepatic morphology may be strikingly affected. These disorders can be grouped according to whether blood flow into, through, or from the liver is impaired (Fig. 18-43).

Impaired Blood Flow into the Liver

Hepatic Artery Compromise

Liver infarcts are rare, thanks to the double blood supply to the liver. Nonetheless, thrombosis or compression of an intrahepatic branch of the hepatic artery by embolism (Fig. 18-44), neoplasia, polyarteritis nodosa (Chapter 11), or sepsis may result in a localized infarct that is either pale and anemic or hemorrhagic if there is suffusion with portal blood. Interruption of the main hepatic artery does not always produce ischemic necrosis of the organ, particularly if the liver is otherwise normal. Retrograde arterial flow through accessory vessels, when coupled with the portal venous supply, is usually sufficient to sustain the liver parenchyma. The one exception is hepatic artery thrombosis in a transplanted liver, which generally leads to infarction of the major ducts of the biliary tree, since their blood supply is entirely arterial.

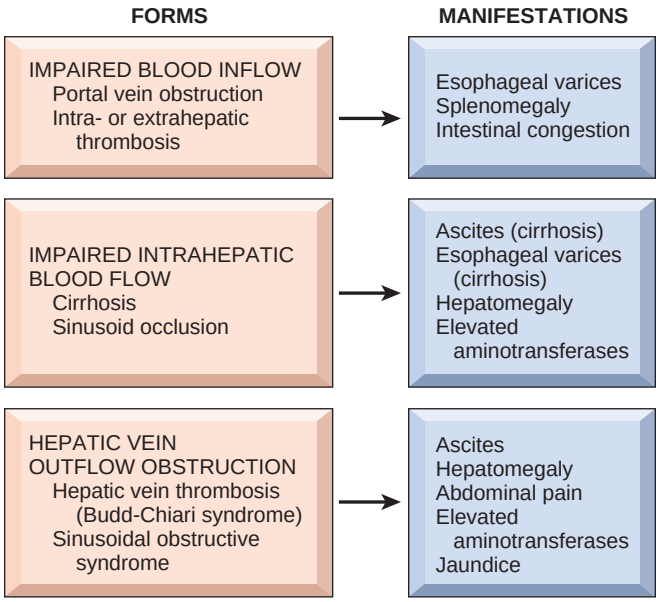

Figure 18-43 Forms and clinical manifestations of hepatic circulatory disorders.

Portal Vein Obstruction and Thrombosis

Blockage of the extrahepatic portal vein may be insidious and well tolerated or may be a catastrophic and potentially lethal event; most cases fall somewhere in between. Occlusive disease of the portal vein or its major radicles typically produces abdominal pain and, in most instances, other manifestations of portal hypertension, principally esophageal varices that are prone to rupture. Ascites is not common (because the block is presinusoidal), but when present is often massive and intractable.

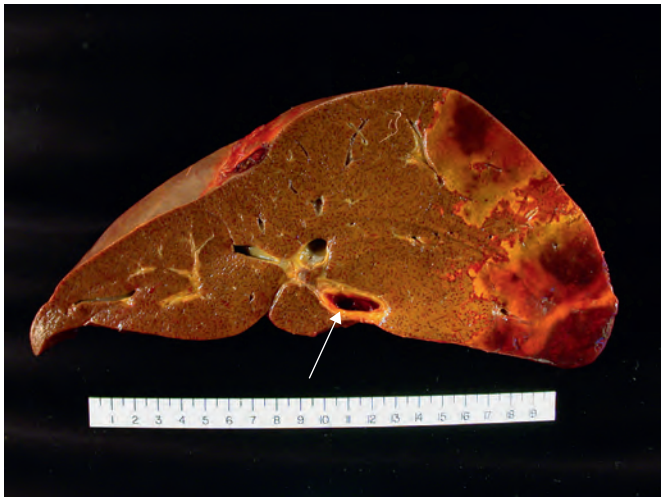

Figure 18-44 Liver infarct. A thrombus is lodged in a peripheral branch of the hepatic artery (arrow) and compresses the adjacent portal vein; the distal hepatic tissue is pale, with a hemorrhagic margin.