Gradually, biliary cirrhosis develops, and patients will progress to decompensated liver disease with all the manifestations of ascites, esophageal variceal hemorrhage, and encephalopathy.

TREATMENT

Primary Sclerosing Cholangitis

There is no specific proven treatment for PSC. Some clinicians use UDCA at “PBC dosages” of 13–15 mg/kg per d with anecdotal improvement, although no study has shown convincing evidence of clinical benefit. A study of high-dose (28–30 mg/kg per d) UDCA found it to be harmful. Endoscopic dilatation of dominant strictures can be helpful, but the ultimate treatment is liver transplantation when decompensated cirrhosis develops. Episodes of cholangitis should be treated with antibiotics. A dreaded complication of PSC is the development of cholangiocarcinoma, which is a relative contraindication to liver transplantation.

■ CARDIAC CIRRHOSIS

Definition Patients with long-standing right-sided congestive heart failure may develop chronic liver injury and cardiac cirrhosis. This is an increasingly uncommon, if not rare, cause of chronic liver disease given the advances made in the care of patients with heart failure.

Etiology and Pathology In the case of long-term right-sided heart failure, there is an elevated venous pressure transmitted via the inferior vena cava and hepatic veins to the sinusoids of the liver, which become dilated and engorged with blood. The liver becomes enlarged and swollen, and with long-term passive congestion and relative ischemia due to poor circulation, centrilobular hepatocytes can become necrotic, leading to pericentral fibrosis. This fibrotic pattern can extend to the periphery of the lobule outward until a unique pattern of fibrosis causing cirrhosis can occur.

Clinical Features Patients typically have signs of congestive heart failure and will manifest an enlarged firm liver on physical