3 to 6 months. Long-term anticoagulation may be used in cases of chronic thrombosis, especially when associated with hypercoagulable states.

Concern exists that anticoagulation may precipitate hemorrhage from varices that arise as a consequence of portal hypertension; however, studies have not shown an increased risk for variceal bleeding in anticoagulated patients with chronic PVT. In fact, recent studies suggest a role for prophylactic anticoagulation (Enoxaparin) for prevention of PVT and hepatic decompensation in cirrhosis. If variceal hemorrhage occurs; it is best managed with endoscopic obliteration. Prophylaxis with β-blockers to prevent variceal bleeding may decrease the portal pressure, potentially propagating thrombus, and therefore is not usually recommended. If endoscopic treatment fails, surgical management with portosystemic shunting may be attempted, but this approach is often difficult because of the absence of suitable patent vessels.

Budd-Chiari Syndrome

Definition and Etiology

Occlusion of the major hepatic veins or the inferior vena cava, especially in the intrahepatic and suprahepatic segments, causes Budd-Chiari syndrome. Most cases are associated with hematologic disease (e.g., polycythemia vera, paroxysmal nocturnal hemoglobinuria, essential thrombocytosis, other myeloproliferative disorders), pregnancy, oral contraceptive use, tumors (especially HCC), or other causes of a hypercoagulable state (e.g., factor V Leiden mutation, protein C and S deficiency). Abdominal trauma and congenital webs of the vena cava are also related to Budd-Chiari syndrome. About 20% of cases are idiopathic, but many of these patients prove to have early, subclinical myeloproliferative disease or genetic mutations associated with a hypercoagulable state.

Clinical Presentation

Budd-Chiari syndrome can manifest acutely, possibly in association with acute liver failure, or it can manifest as a subacute or chronic illness. Acute disease produces right upper quadrant abdominal pain, hepatomegaly, ascites, and jaundice, whereas the subacute or chronic form produces primarily portal hypertension. Elevation of serum bilirubin and transaminase levels may be mild, but liver function is often poor, with profound hypoalbuminemia and coagulopathy.

Diagnosis

The diagnosis can be established noninvasively with Doppler ultrasonography, which shows decreased or absent hepatic vein blood flow, and computed tomography, which shows delayed or absent contrast filling of the hepatic veins and hypertrophy of the caudate lobe. Magnetic resonance angiography may also demonstrate these findings. Hepatic venography is especially useful if the results noninvasive imaging are inconclusive. Venography often shows an inability to catheterize and visualize the hepatic veins; the characteristic spiderweb pattern of collateral vessels may also be demonstrated, and the inferior vena cava may appear compressed owing to hepatomegaly or an enlarged caudate lobe. On liver biopsy, centrilobular congestion, hemorrhage, and necrosis (nutmeg liver) are seen, with cirrhosis developing in patients with chronic obstruction.

Treatment

Treatment should be individualized and is dependent on the mode and severity of presentation and the potential cause of the disease. Supportive therapy to relieve ascites and edema (e.g., dietary sodium restriction, diuretics) and chronic anticoagulation may be considered for patients with chronic Budd-Chiari syndrome in whom methods to decompress congestion are not feasible. Thrombolysis followed by anticoagulation is most useful in patients with acute forms of the disease. In selected patients (such as those with venous webs or strictures or single-vessel thrombosis), angioplasty with or without stent placement may be used. Decompressive modalities are most useful before the development of cirrhosis and include transjugular intrahepatic portacaval and side-to-side portacaval shunts. In patients with cirrhosis, liver transplantation followed by continued anticoagulation is often considered the best option.

Veno-Occlusive Disease

Definition and Etiology

Hepatic veno-occlusive disease, also called sinusoidal obstruction syndrome, often occurs after cytoreductive therapy and before bone marrow transplantation but may also follow exposure to other drugs or herbal preparations (e.g., azathioprine, pyrrolizidine alkaloids). Endothelial cell injury leads to obstruction at the level of the hepatic venules and the sinusoids.

Clinical Presentation

The disease is characterized by jaundice, painful hepatomegaly, and fluid retention. Clinical manifestations can be rapidly progressive and lead to multiorgan dysfunction and death in 20% to 25% of patients.

Diagnosis

The diagnosis is clinically suspected when weight gain, epigastric or right upper quadrant abdominal pain, and jaundice develop within the first 3 to 4 weeks after bone marrow transplantation. Laboratory abnormalities include hyperbilirubinemia, elevated transaminases, and, in severe cases, profound synthetic dysfunction. Doppler abdominal ultrasonography may reveal ascites, reversal of portal vein flow, and an elevated hepatic artery resistance index. Liver biopsy is diagnostic and is usually obtained with use of the transjugular approach. The advantages of this approach compared with the percutaneous route include the ability to measure the hepatic venous pressure gradient (which is typically elevated in veno-occlusive disease) and a lower incidence of bleeding.

Treatment

Mild forms of the disease may favorably respond to supportive therapy alone. In moderate to severe disease, treatment has been attempted with tissue plasminogen activator and heparin, antithrombin III, prostaglandin E_1, and glutamine plus vitamin E, although the efficacies of these treatments have not been clearly established. Recently, defibrotide (a mixture of porcine-derived