Selective Management of Hepatic Venous Outflow Obstruction

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To describe the outcome of selective management of hepatic venous outflow obstruction (HVOO), based on its presentation and liver function, we reviewed the records of 49 consecutive patients managed at our institution between 1984 and 1993. Twenty-six patients were managed surgically, 12 nonsurgically, and 11 were not treated. Portosystemic shunts (PSS) were performed in 18 patients (patency 83%). Two patients (11%) died postoperatively, 11 (61%) did well (mean follow-up 6.4 years), three (17%) required subsequent orthotopic liver transplantation, and two (11%) died of late liver failure. PSS remained patent if the preoperative pressure gradient between the portal vein and the infrahepatic inferior vena cava was greater than 10 mm Hg and across the intrahepatic inferior vena cava 18 mm Hg or less. All six orthotopic liver transplantations (three as primary treatment and three after failed PSS) were successful (mean follow-up 4.8 years). Five patients underwent other procedures. Nine (75%) of the 12 nonsurgically treated patients did well (mean follow-up 3.8 years). The most important predictor of successful outcome after PSS or medical management was the degree of liver function. All 11 untreated patients died either of end-stage liver failure (n = 7; 63%) or of severe comorbid disease (n = 4; 37%). In patients with preserved liver function, medical management of HVOO can be successful early in the course of the disease; a late presentation necessitates PSS. Orthotopic liver transplantation should be employed in patients with liver failure and may decrease the high mortality rate of HVOO. (J GASTROINTEST SURG 1997;1:377-385.)

Hepatic venous outflow obstruction (HVOO), historically referred to as the Budd-Chiari syndrome, is a complex clinicopathologic disease of variable etiology, characterized by thrombosis of the hepatic veins[1] and associated with progressive hepatocellular dysfunction. It is rare in the general population; patients with HVOO comprise less than 5% of those operated on for portal hypertension[2] and approximately 1% of those undergoing liver transplantation.[3] Regardless of the mechanism of HVOO, increased postsinusoidal resistance to hepatic blood flow causes centrilobular venous congestion, intrahepatic hypertension, and hepatocellular injury. Clinical evidence suggests that severity of hepatic dysfunction is probably related to the rapidity of onset and extent of HVOO.[4] Abrupt, complete HVOO leads to extensive hepatocellular necrosis and fulminant hepatic failure. In contrast, insidious, incomplete HVOO may be associated with portal decompression through hepatovenous collateral vessels and reduced hepatocellular injury. This pathophysiologic concept of HVOO suggests a broad clinical spectrum of disease.

We have been impressed by the variability in clinical presentation and the unpredictability of progression of HVOO. Consequently the management of patients with HVOO has been difficult. Numerous treatment options ranging from diuretic alone to orthotopic liver transplantation have proved effective, but few reports have evaluated an unselected patient cohort to assess the disease spectrum and outcome.[5-7] Although HVOO is being diagnosed with increasing frequency,[8] its low incidence makes a prospective study unrealistic. Thus our aims were to review the spectrum of clinical presentation and treatment of HVOO, to characterize factors associated with disease severity and chronicity, to determine the long-term clinical outcome at a referral center, and to propose an algorithm for clinical management.

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