Primary Budd-Chiari syndrome: outcome of endovascular management for suprahepatic venous obstruction.

Lee BB, Villavicencio L, KimYW, DoYS, KohKC, LimHK, LimJH, AhnKW.

Department of Surgery, Uniformed Services University of the Health Sciences, Bethesda, MD, USA. bblee38@comcast.net

OBJECTIVE: Primary Budd-Chiari syndrome (BCS) is a rare form of hepatic venous outflow obstruction at the suprahepatic inferior vena cava (IVC), the hepatic veins, or both. We assessed our 4-year experience in the management of BCS to evaluate the results of our methods of care. METHODS: We conducted a retrospective review of a nonrandomized clinical trial conducted in three teaching hospitals. Among 28 primary BCS patients, 9 remained in medical treatment only, and 19 who failed to respond to medical treatment received additional endovascular (n = 17) or surgical therapy (n = 2). Nine underwent IVC balloon angioplasty alone, 6 had angioplasty plus stents, and 2 had transjugular intrahepatic portosystemic shunts (TIPS) for hepatic vein lesions. One patient had a mesoatrial bypass; another had liver transplantation. Immediate response to the therapy was assessed with angiography and ultrasonography based on anatomic and/or hemodynamic correction or reduction of the lesion. Subsequent assessment of portal hypertension status was made with periodic clinical and laboratory evaluation (eg, ultrasonography, liver biopsy). RESULTS: Twenty-six patients had had IVC stenosis or occlusion by focal or segmental lesion. Two patients had hepatic vein outlet obstruction. There was no evidence of coagulopathy as the pathogenesis; all were related to membranous obstruction of the vena cava. Excellent immediate response to the endovascular therapy and subsequent relief of portal hypertension were achieved in 14 patients. Four patients had restenosis or progression of the residual lesion within 2 years; three responded to repeated stenting. Primary patency was 76.5%, and primary assisted patency was 94.1%. Two patients with TIPS and two with surgical therapy maintained excellent results. The medical treatment remained effective only in a limited group of 6 (21.4%) of the 28 patients. CONCLUSIONS: In BCS, both endovascular and surgical interventions provide excellent results and potentially halt liver parenchymal deterioration caused by portal hypertension. Liver transplantation remains the ultimate solution for advanced liver failure.