CLINICAL STUDY

Anatomic Recanalization of Hepatic Vein and Inferior Vena Cava versus Direct Intrahepatic Portosystemic Shunt Creation in Budd-Chiari Syndrome: Overall Outcome and Midterm Transplant-Free Survival

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ABSTRACT

Purpose: To assess overall outcome and midterm transplant-free survival of patients with Budd-Chiari syndrome (BCS) undergoing radiologic interventions including anatomic recanalization of the hepatic vein (HV) and inferior vena cava (IVC) and direct intrahepatic portosystemic shunt (DIPS) creation, both as combined and as independent groups.

Materials and Methods: From November 2010 to October 2014, 136 patients with BCS were treated with HV/IVC recanalization (group 1) or DIPS creation (group 2). Both groups were periodically analyzed for stent patency on Doppler ultrasound, clinical outcome, biochemical parameters, and survival until death, liver transplantation, or last clinical evaluation.

Results: Actuarial transplant-free survival for the entire cohort was 94% at 1 year and 5 years with no significant difference in overall survival. There was significant biochemical improvement in group 1 with decrease in mean serum bilirubin level (1.8 mg/dL to 1.4 mg/dL, $P < .011$), mean serum aspartate aminotransferase (48.6 IU/L to 33.2 IU/L, $P < .05$), and mean serum alanine aminotransferase (38.7 IU/L to 28.5 IU/L) and increase in mean serum albumin level (3.2 g/dL to 3.45 g/dL, $P < .001$) after 3 and 24 months. There were 4 deaths in each group at 1-year follow-up; all 4 patients had acute fulminant BCS at presentation.

Conclusions: Radiologic interventions for BCS lead to remarkable improvement of liver function and a good overall outcome and midterm transplant-free survival. Patients receiving anatomic recanalization show improved liver synthetic functions compared with patients treated with DIPS.

ABBREVIATIONS

BCS = Budd-Chiari syndrome, DIPS = direct intrahepatic portosystemic shunt, HV = hepatic vein, IVC = inferior vena cava, METAVIR = meta-analysis of histological data in viral hepatitis

Budd-Chiari syndrome (BCS) is a rare disorder caused by obstruction of the hepatic venous outflow tract at any level between the small hepatic veins (HVs) and the right atrium (1). Clinical presentation of hepatic venous congestion ranges from asymptomatic to fulminant liver failure, most commonly as subacute liver disease with manifestation of portal hypertension, such as ascites or upper gastrointestinal bleeding (2,3). Prompt, appropriate treatment improves prognosis. Medical management primarily consists of anticoagulation; surgical options include portosystemic shunt creation and orthotopic liver transplantation (4,5). However, parenchymal damage may be mitigated by re-establishing hepatic venous outflow, which has been achieved by anatomic recanalization of the HV and inferior vena cava (IVC) and direct intrahepatic portosystemic shunt (DIPS) creation, the mainstay radiologic endovascular interventions for BCS (6). Liver transplantation may be offered to patients who do not benefit from radiologic interventions or patients with acute fulminant hepatic failure. There is wide variation in the treatment algorithm in BCS

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owing to lack of controlled clinical trials (3). Two previous studies used prognostic indices and scores for objective assessment of disease severity and efficacy of treatment (7,8). The aim of the present study was to evaluate the overall outcome and midterm transplant-free survival in patients with BCS treated by either anatomic recanalization of HV/IVC or DIPS creation and to determine factors that predict favorable outcome in these patients.

MATERIALS AND METHODS

This study was approved by the institutional review board.

Patients

BCS was defined as hepatic venous outflow obstruction at any level between the small HVs and the junction between the IVC and the right atrium and confirmed by imaging (Doppler ultrasound, computed tomography or magnetic resonance imaging scan, or hepatic venography). Review of the hospital electronic database identified 180 patients with BCS presenting from November 2010 to October 2014. Of 180 patients, 136 patients required radiologic interventions and were included in this study; the remaining patients received other treatments and hence were excluded. Liver biopsy was performed in 60 patients before radiologic intervention; in 5 of these patients, imaging findings were not classic for BCS, and biopsy was done in the remaining 55 patients to grade the fibrosis per institutional protocol. The patients were divided into 2 groups: group 1 underwent HV/IVC recanalization, and group 2 required DIPS creation. Of 136 patients, 50 patients also underwent transient elastography before and after radiologic intervention. Biochemical, radiologic, and biopsy details were collected from patient data in a case record form before and after radiologic intervention and at the end of follow-up, which was death, liver transplantation, or last follow-up.

Transient Elastography

Transient elastography was performed using a FibroScan ultrasound device (Echosens, Paris, France) before and after radiologic intervention in patients to observe the effect of intervention on hepatic decongestion. The measurements were obtained from right lobe of liver using a 3.5-MHz transducer. Mean value of measurement was obtained using 10 successful values and was calculated in kilopascals (kPa). Transient elastography was not done in patients with severe ascites; however, paracentesis was performed in patients with mild ascites before transient elastography.

Patient Selection for Radiologic Interventions

Routine abdominal ultrasound examination and Doppler examination were performed to assess the status of native HVs. A short segment narrowing was defined as narrowing of HV up to 2 cm. These HVs were considered to have potential for flow restoration by balloon dilation with or without stent placement. In patients with long segment narrowing or complete nonvisualization of all HVs including the accessory HVs, DIPS creation was performed. All procedures were done under fluoroscopy with ultrasound guidance.

Technique

Recanalization of HV/IVC. Transjugular access was obtained, and a 10-F, 40-cm-long sheath (Cook, Inc, Bloomington, Indiana) was placed in the IVC and venography was performed. If HV stump was identified, an MPA catheter (Cook, Inc) and sheath assembly was used to negotiate and cross the occluded segment of HV using a straight-tip 0.035-inch hydrophilic guide wire (Radifocus; Terumo Corporation, Tokyo, Japan). The hydrophilic guide wire was advanced distal to stenosis. Later it was exchanged with a 260-cm, 0.035-inch Amplatz Extra Stiff guide wire (Cook, Inc), and the stenotic segment was dilated with a 6- to 12-mm balloon catheter (Mustang; Boston Scientific, Marlborough, Massachusetts); the balloon catheter was oversized by 30%–50% of the normal venous lumen. In case of residual stenosis, stent placement was done using a self-expanding metallic stent (E-Luminexx; Bard Peripheral Vascular, Tempe, Arizona). If there was difficulty in negotiating the site of occlusion through the transjugular route, a percutaneous transhepatic route was taken. The HV was punctured and accessed under ultrasound guidance, and the occluded segment was crossed using a straight-tip 0.035-inch hydrophilic guide wire. In resistant cases, the reverse end of the 0.035-inch hydrophilic guide wire was also tried. After crossing the occluded segment, the floppy end of the guide wire was placed in the superior vena cava. The wire was then snared and pulled out through the jugular vein to enable balloon dilation or stent insertion through the jugular route (Fig 1a, b).

In 4 patients, catheter-directed thrombolytic therapy with urokinase (50,000–100,000 IU) was used for acute HV thrombosis through a transjugular approach using a 5-F multi-side-port infusion catheter (Cook, Inc) followed by balloon angioplasty. IVC web/short segment stenosis was attempted via either jugular or femoral venous access or combined jugular and femoral venous access in resistant cases. Cavography was performed to evaluate for the site and degree of occlusion. In cases of incomplete occlusions, a straight-tip hydrophilic guide wire was negotiated across the stenosis followed by sequential balloon dilation up to 20–24 mm (Atlas; Bard Peripheral Vascular). A stent was placed only if there was immediate recoil or the pressure gradient across the stenosis was $>$ 8 mm Hg. If the occlusion was complete, the reverse end of a 0.035-inch hydrophilic guide wire was used to cross it. Failure to cross the narrowing made it necessary to use a combined transfemoral