Budd-Chiari Syndrome: Hepatic Venous Web Outflow Obstruction Treated by Percutaneous Placement of Hepatic Vein Stent

Alireza Bozorgmanesh, M.D.,¹ D. Arul Selvam, M.D.,¹ and James G. Caridi, M.D.¹

ABSTRACT

Budd-Chiari syndrome (BCS) denotes a heterogeneous group of diseases characterized by hepatic venous outflow obstruction at the level of the hepatic veins or inferior vena cava resulting in portal hypertension. Traditional approach to treatment of BCS involves systemic thrombolysis and surgical portosystemic shunt or transjugular intra-hepatic portosystemic shunt in progressive cases of BCS or as a bridge to transplantation. Recently, an increasing number of successful reports of BCS therapy have involved endovascular techniques, including angioplasty and stent placement. The present report illustrates successful percutaneous recanalization of complete hepatic vein occlusion by angioplasty and stent implantation in a patient with membranous obstruction.

KEYWORDS: Budd-Chiari syndrome, hepatic vein obstruction, hepatic vein stent, inferior vena cava stenosis, endovascular treatment

Objectives: Upon completion of this article, the reader should become familiar with Budd-Chiari syndrome and current approach to its diagnosis and treatment, including hepatic vein and inferior vena cava stent placement.

Accreditation: Tufts University School of Medicine (TUSM) is accredited by the Accreditation Council for Continuing Medical Education to provide continuing medical education for physicians.

Credit: TUSM designates this educational activity for a maximum of 1 *AMA PRA Category 1 Credit*TM. Physicians should only claim credit commensurate with the extent of their participation in the activity.

Budd-Chiari syndrome (BCS) results from hepatic venous outflow obstruction at the level of the hepatic veins or inferior vena cava. Although traditional management of BCS involves thrombolysis or surgical portosystemic shunt creation, an increasing number of successful reports of BCS therapy have involved endovascular techniques, including angioplasty and stent placement. We describe successful percutaneous recanalization of complete hepatic vein occlusion by angioplasty and stent implantation in a patient with

membranous obstruction, and subsequently discuss clinical aspects of BCS.

CASE REPORT

A previously healthy 7-year-old boy was referred to our institution with a 2-month history of abdominal distension and decreased energy. The patient had no history of abdominal pain, jaundice, bruising, or acholic stools. Laboratory examination at another hospital revealed

¹Department of Radiology, University of Florida College of Medicine, Gainesville, Florida.

Address for correspondence and reprint requests: James G. Caridi, M.D., Director of Vascular and Interventional Radiology Fellowship Program, Department of Radiology, University of Florida College of Medicine, PO Box 100374, Gainesville, FL 32610.

Case-Based Interventional Radiology; Guest Editors, Ron C.

Gaba, M.D., Derek L. West, M.D., James T. Bui, M.D., Charles A. Owens, M.D.

Semin Intervent Radiol 2007;24:100–105. Copyright © 2007 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662.
DOI 10.1055/s-2007-971200. ISSN 0739-9529.

Figure 1 (A) and (B) Axial computed tomography images demonstrate narrowing of inferior vena cava secondary to enlarged caudate lobe (arrow). There is patchy liver parenchymal enhancement, or so-called nutmeg liver. Ascites are also present.

abnormal liver enzymes. Ultrasound examination had revealed some ascites with normal hepatopetal portal blood flow. Magnetic resonance (MR) imaging examination also performed at the outside hospital demonstrated poor visualization of the hepatic veins. However, the ultrasound and MR images were not available for our review. Clinical examination confirmed abdominal distention with dilated cutaneous veins, hepatomegaly, and ascites. Repeat laboratory evaluation revealed mildly elevated coagulation studies and bilirubin levels with near normal serum albumin and transaminases. Computed tomography (CT) examination performed at our institution revealed hepatomegaly with heterogeneous enhancement, so-called nutmeg liver (Fig. 1). The caudate lobe appeared hypertrophied, narrowing the inferior vena cava (IVC) (Fig. 1). Moderate ascites was also noted (Fig. 1). The patient was diagnosed with BCS and referred to interventional radiology for venography and possible intervention.

Venography was performed utilizing a transjugular approach, and it revealed a paucity of hepatic veins (Fig. 2). Probing was attempted but unsuccessful. Percutaneous transhepatic access confirmed patent hepatic veins with central occlusion due to web with intrahepatic collateral vessel formation (Fig. 3). A Glidewire (Terumo Medical, Somerset, NJ) was then manipulated through the hepatic vein occlusion and advanced successfully to the IVC and the right jugular vein using a snare technique (Fig. 4). Balloon angioplasty of the occlusion with 8- and 10-mm balloons reestablished flow (Figs. 5 and 6). However, the collateral vessels persisted (Fig. 6). Hence, a bare 8 mm $\times$ 3.8 cm hepatic vein stent was successfully placed with good results (Fig. 7). The patient was ambulating the following day, and he had marked reduction in abdominal girth. No postprocedure anticoagulation was given. Subsequent examinations during the

following 2 years revealed a patent hepatic vein stent without recurrent symptoms (Fig. 8).

DISCUSSION

Budd-Chiari Syndrome

BCS denotes a heterogeneous group of diseases characterized by hepatic venous outflow obstruction at the level of hepatic veins or inferior vena cava resulting in portal hypertension. The syndrome can be fulminant, acute,

Figure 2 Digital subtraction angiography demonstrates small invagination in expected position of main hepatic vein, as well as lack of hepatic veins, indicative of Budd-Chiari syndrome.

Figure 3 Digital subtraction angiography following percutaneous transhepatic access shows patent hepatic veins with central occlusion and intrahepatic collateral vessel formation.

chronic, or asymptomatic. It occurs in 1 out of 100,000 individuals and is more common in females.

Hepatic Venous Anatomy and Pathophysiology

Hepatic veins serve as the only conduits of blood from the liver to the IVC and right heart. The right,

middle, and left hepatic veins connect the liver to the retrohepatic IVC in the bare area of liver below the diaphragm. The right hepatic vein drains directly into the IVC; left and middle hepatic veins often unite to drain (80%). Small accessory hepatic veins from the pericaval liver segments drain directly into the IVC. The caudate lobe of the liver is the only hepatic segment with direct venous drainage to the IVC. This explains the degree of hypertrophy and the perfusion changes noted in the caudate lobe on imaging studies of BCS. Hepatic vein outflow obstruction leads to increased hepatic venous pressure, resulting in collateral venous circulation. The sites of collateral veins in BCS are different from portosystemic collaterals from portal hypertension. The collateral veins of deep and central tributaries of the systemic veins, such as ascending lumbar veins, vertebral venous plexus, azygous, and hemiazygous veins, are commonly seen in BCS. In addition, the increasing hepatic venous pressure leads to increased sinusoidal pressure and finally to portal vein thrombosis or portal hypertension. This cascade of events results in hepatocyte dysfunction and leads to liver failure.

Causes of Budd-Chiari Syndrome

The etiology of BCS may be separated into primary and secondary causes. Primary BCS is due to intrinsic intraluminal thrombosis or webs. BCS may be secondarily caused by extraluminal compression (by abscess

A

B

Figure 4 (A,B) Fluoroscopic images demonstrate snare technique.

Figure 5 Fluoroscopic image shows balloon angioplasty of occlusion.

or tumor) or intraluminal invasion by tumor or parasite. The most frequent cause of BCS in Western countries is thrombotic occlusion in a hypercoagulable state.

Figure 6 Digital subtraction angiography image reveals reestablished flow following balloon angioplasty of occlusion, although persistence of collateral vessels is seen.

Primary Budd-Chiari Syndrome

HEPATIC VEIN THROMBOSIS AND HYPERCOAGULABLE STATES

Hypercoagulable states and myeloproliferative disorders cause the majority of BCS cases, ~75%. The most common hypercoagulable causes of BCS include protein C or S deficiency, essential thrombocythosis, systemic lupus erythematosus, factor V Leiden, myelofibrosis, polycythemia vera, paroxysmal nocturnal hemoglobinuria, antiphospholipid syndrome, and oral contraceptive use.

HEPATIC VEIN OR INFERIOR VENA CAVA WEBS

Studies suggest that hepatic vein webs or IVC webs are mostly due to postthrombotic sequelae. Histopathology of webs has revealed replaced intima with fibrous laminar structure, fresh and organized thrombi, recanalizations, and calcifications. This correlates well clinically with late onset of disease in these patients.

Secondary BCS

EXTRINSIC COMPRESSION OF HEPATIC VEINS OR INFERIOR VENA CAVA

Mass effect from regional malignancy or infectious processes can compress the IVC leading to BCS. Infectious processes include tuberculosis, syphilis, aspergillosis, and parasites.

STENOSIS OF THE HEPATIC VEIN OR INFERIOR VENA CAVA

BCS can also be acquired, such as hepatic venous stenosis as a complication of liver transplantation. Tumor thrombi from hepatocellular carcinoma, renal cell carcinoma, or retroperitoneal sarcoma growing into the intrahepatic IVC can also cause hepatic venous obstruction leading to BCS.

Clinical Presentation

Patients with BCS typically present with abdominal swelling, abdominal pain, ascites, hepatomegaly, and absent hepatojugular venous reflex. BCS should be suspected if there is simultaneous presentation of abdominal pain, ascites, and hepatomegaly. Patients with BCS can also be separated by the chronicity and severity of symptoms. These include acute fulminant, subacute, and chronic BCS. Subacute or chronic presentations are more common than acute fulminant course. The chronic presentations include those of chronic venous congestion of the liver leading to sequelae of portal hypertension and cirrhosis. Patients with subacute or chronic BCS may rarely be asymptomatic. Patients with acute hepatic venous obstruction leading to acute fulminant liver failure typically present

Figure 7 (A,B) Digital subtraction angiography images demonstrate placement of bare 8 mm x 3.8 cm hepatic vein stent with good results.

with nausea, vomiting, and jaundice, with markedly abnormal liver function tests such as aspartate transaminase, alanine transaminase, and bilirubin levels.

Budd-Chiari Syndrome Diagnosis

ULTRASOUND

The physiological flow patterns in hepatic veins reflect pressure variations in the right atrium. Hepatic venous flow is triphasic on Doppler ultrasound. The triphasic flow pattern is altered in BCS and right heart failure. Liver cirrhosis leads to decreased hepatic compliance, altering the hepatic venous waveform from a triphasic to

a monophasic form. In addition, severe portal hypertension or portal vein thrombosis may cause hepatofugal flow. Specific diagnostic signs of hepatic vein involvement in BCS include nonvisualization with flow void signal, fibrous cord in place of hepatic vein, and thrombus appearing as echogenic material in the hepatic veins. Evidence of intrahepatic collateral vessels is highly suggestive of BCS. These small spiderweb early collaterals, collateral subcapsular vessels, and intrahepatic venous collaterals appear as “bicolored hepatic veins.” These veins appear bicolored due to opposing flow directions seen in adjacent veins on color Doppler imaging. Other features include collateral vessels to the IVC, hepatic and portal vein collateral vessels, and prominent caudate vein measuring ≥ 3 mm in the absence of heart failure. Hepatic veins may also appear tortuous, curvilinear, fragmented, stenotic, or as a fibrous cord with slow, turbulent, or reversed flow. A combination of these ultrasound findings may be seen in other conditions such as regenerative nodules, caudate lobe hypertrophy, heterogeneous hepatic parenchyma, or portal vein thrombosis. Direct sonographic analysis of the IVC can also reveal enlargement, stenosis, or obstruction of the IVC by a thrombus or web.

COMPUTED TOMOGRAPHY AND MAGNETIC RESONANCE EXAMINATIONS

CT and MR angiography provide noninvasive anatomical definition of the hepatic artery that may be important for operative or interventional planning. CT and MR examinations may also reveal secondary causes of BCS. CT and MR portography provide important information regarding portal venous system patency or thrombosis that will dictate treatment options for

Figure 8 Axial computed tomography image shows patent hepatic vein stent with normal-size liver, patent hepatic veins, normal caliber inferior vena cava, and lack of ascites.

portocaval shunt or transjugular intrahepatic portosystemic shunt (TIPS). CT and MR examinations can also reveal information regarding intra- and extrahepatic collateral vessels, as well as IVC morphology and patency. When performing hepatic imaging in the setting of BCS, the sluggish venous flow should be accounted for when acquiring delayed venous phase imaging.

SELECTIVE HEPATIC VENOGRAPHY

Cannulation of hepatic veins in BCS can be a challenging task. Moderate sedation with local anesthesia for adults and general anesthesia for children is preferred to facilitate the procedure. Transjugular or percutaneous transhepatic injection of contrast using fine needle access is performed with iodinated contrast or CO₂ injection of liver parenchyma. The latter may be better to evaluate collateral vessels and may be used in patients with a history of adverse contrast reaction or renal insufficiency. Hepatic vein access allows for pressure measurements, analysis of hepatic venous flow dynamics, localization of the site of obstruction, catheter-directed thrombolysis, and hepatic venous stent placement. Venacavography is often performed with a jugular or femoral vein approach often combined with hepatic venography.

Treatment of Budd-Chiari Syndrome

Despite proposed guidelines for the management of BCS by an expert hepatic vascular disorder panel in 2003, the treatment of BCS remains somewhat controversial. The traditional approach to treatment involves systemic thrombolysis in all BCS patients without a specific contraindication. Patients undergoing systemic or catheter-directed thrombolysis should be closely monitored for clot propagation using a combination of Doppler sonography, CT, and MR examinations. Surgical portosystemic shunt or TIPS is performed in progressive cases of BCS or as a bridge to transplantation. Patients with acute or fulminant BCS are best served with liver transplantation. Recently, there have been an increasing number of successful reports involving endovascular techniques including angioplasty and stent placement. These techniques are especially effective in cases of hepatic or IVC webs, as demonstrated in our case.

ACKNOWLEDGMENTS

Regie Gonzalez, M.D., Department of Pediatric Gastroenterology, University of Florida—Gainesville, for case referral to interventional radiology.

SUGGESTED READINGS

- Bargallo X, Gilabert R, Nicolau C, Garcia-Pagan JC, Ayuso JR, Bru C. Sonography of Budd-Chiari syndrome. *AJR Am J Roentgenol* 2006;187:W33–W41
- Bogin V, Marcos A, Shaw-Stiffel T. Budd-Chiari syndrome: in evolution. *Eur J Gastroenterol Hepatol* 2005;17:33–35
- Bozkurt G, Cil B, Akbay A, Turk CC, Palaoglu S. Intractable radicular and low back pain secondary to inferior vena cava stenosis associated with Budd-Chiari syndrome: endovascular treatment with cava stenting—case report and review of the literature. *Spine* 2006;31:E383–E386
- Desser TS, Sze DY, Jeffrey RB. Imaging and intervention in the hepatic veins. *AJR Am J Roentgenol* 2003;180:1583–1591
- Erden A, Erden I, Karayalcin S, Yurdaydin C. Budd-Chiari syndrome: evaluation with multiphase contrast-enhanced three-dimensional MR angiography. *AJR Am J Roentgenol* 2002;179:1287–1292
- Janssen HL, Garcia-Pagan JC, Elias E, et al. Budd-Chiari syndrome: a review by an expert panel. *J Hepatol* 2003;38:364–371
- Kage M, Arakawa M, Kojiro M, Okuda K. Histopathology of membranous obstruction of the inferior vena cava in Budd-Chiari syndrome. *Gastroenterology* 1992;102:2081–2090
- Kavanagh PM, Roberts J, Gibney R, Malone D, Hegarty J, McCormick PA. Acute Budd-Chiari syndrome with liver failure: the experience of a policy of initial interventional radiological treatment using transjugular intrahepatic portosystemic shunt. *J Gastroenterol Hepatol* 2004;19:1135–1139
- Lee BB, Villavicencio L, Kim YW, et al. Primary Budd-Chiari syndrome: outcome of endovascular management for suprahepatic venous obstruction. *J Vasc Surg* 2006;43:101–108
- Mahmoud AE, Mendoza A, Meshikhes AN, et al. Clinical spectrum, investigations and treatment of Budd-Chiari syndrome. *QJM* 1996;89:37–43
- Menon KV, Shah V, Kamath PS. The Budd-Chiari syndrome. *N Engl J Med* 2004;350:578–585
- Rerksupphol S, Hardikar W, Smith AL, et al. Successful stenting for Budd-Chiari syndrome after pediatric liver transplantation: a case series and review of the literature. *Pediatr Surg Int* 2004;20:87–90
- Sanchez-Recalde A, Sobrino N, Galeote G, Calvo Orbe L, Merino JL, Sobrino JA. Budd-Chiari syndrome with complete occlusion of the inferior vena cava: percutaneous recanalization by angioplasty and stenting. *Rev Esp Cardiol* 2004;57:1121–1123
- Sharma S, Texeira A, Texeira P, Elias E, Wilde J, Olliff SP. Pharmacological thrombolysis in Budd Chiari syndrome: a single centre experience and review of the literature. *J Hepatol* 2004;40:172–180
- Zhang CQ, Fu LN, Zhang GQ, et al. Long-term effect of stent placement in 115 patients with Budd-Chiari syndrome. *World J Gastroenterol* 2003;9:2587–2591