Intrahepatic Collateral Recanalization in Symptomatic Budd-Chiari Syndrome: A Single-center Experience

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The authors present a single-institutional experience with intrahepatic collateral vessel recanalization as a treatment option in symptomatic Budd-Chiari syndrome (BCS). Over a period of 26 months, this procedure was performed in four symptomatic patients in whom standard hepatic vein recanalization was not feasible or had failed, with a follow-up duration ranging from 7 to 44 months. Based on these cases, intrahepatic collateral vessel recanalization is a promising treatment option in suitable patients with symptomatic BCS and is deserving of further study.


Abbreviations: ALP = alkaline phosphatase, ALT = alanine aminotransferase, AST = aspartate aminotransferase, BCS = Budd-Chiari syndrome, IVC = inferior vena cava, TIPS = transjugular intrahepatic portosystemic shunt

HEPATIC venous outflow obstruction, also known as Budd-Chiari syndrome (BCS), is a rare condition that causes hepatic congestion, portal hypertension, hepatocyte necrosis, and eventual liver failure (1). Ultrasonography (US) and Color Doppler studies are the most widely used imaging studies for its diagnosis. The US findings in BCS include inferior vena cava (IVC) webs and thrombi, IVC narrowing, hepatic venous thrombosis, enlarged caudate lobe, ascites, and intrahepatic or extrahepatic collateral vessels. Color Doppler studies show monophasic to absent flow in the hepatic veins and high flow velocities in the areas of stenosis in the IVC or hepatic veins (2).

The reported incidence of intrahepatic collateral vessels in BCS is approximately 50% (3). These intrahepatic collateral vessels are alternative pathways for venous return from obstructed hepatic veins to the IVC (4). Hence, documentation of intrahepatic collateral vessels by imaging studies is important to understand the evolution of the disease in these patients.

The aim of treatment in patients with BCS is to decompress the liver by providing adequate venous outflow to the liver (5). This is usually achieved by endovascular recanalization of hepatic veins, IVC, or both; or by the creation of endovascular cavoportal shunts (6,7).

We present our institutional experience of intrahepatic collateral recanalization as a treatment option in symptomatic patients with BCS in whom hepatic vein recanalization was not possible or was unsuccessful. In our practice, all patients with clinical suspicion of BCS were evaluated with US, color Doppler imaging, and confirmatory venography. Based on the site(s) of venous occlusion, these patients were classified into three types: type 1 included cases in which the IVC and hepatic veins were involved; type 2 when only hepatic veins were involved, and type 3 when only the IVC (ie, hepatic and/or suprahepatic segments) was involved. From May 2005 to July 2007, four suitable patients underwent intrahepatic collateral recanalization.

PATIENT SELECTION

The endovascular intrahepatic collateral recanalization procedure was approved by our institutional review board. The patients with type 2 BCS who were not suitable for the standard hepatic vein recanalization because of its complete or nearly complete occlusion, or who had undergone failed hepatic vein recanalization procedures, were screened for intrahepatic collateral recanalization. This included mapping of intrahepatic collateral vessels that were dilated (≥ 7 mm) by stenosis or occlusion at the ostium with the IVC (Figs 1, 2). Patients deemed suitable based on the aforementioned criteria underwent intrahepatic collateral recanalization.

PROCEDURE

The initial access was a right femoral or right internal jugular venous approach based on the orientation of the
compromised ostium of the collateral to be recanalized. A percutaneous US-guided image of the collateral vessel can be obtained to establish the orientation of the collateral ostium in relation to the IVC and also to confirm the morphology of the obstruction. This also delineates the flow dynamics within these collateral vessels and serves as a valuable road map for the recanalization procedure (Fig 3).

After venous access was secured with a 7-F introducer, the ostium to be recanalized was engaged with a SIM-1 catheter (Boston Scientific, Cork, Ireland) when the right femoral venous approach was used (Fig 4) or a multipurpose or Cobra catheter (Cook, Bloomington, Indiana) when the right internal jugular approach was employed (Fig 5). After the ostium was engaged with the catheter, the stenotic/occluded segment was crossed with a 0.35-inch hydrophilic wire (Terumo, Tokyo, Japan), which was later exchanged for a 0.35-inch guide wire. Pressure measurements within the dilated collateral vessel, IVC, and right atrium and their gradients were routinely measured. The occluded segment was recanalized with an angioplasty balloon of suitable size and, if needed, a stent combination with restoration of normal hepatofugal flow (Figs 6, 7).

FOLLOW-UP PROTOCOL

The first follow-up study with US and color Doppler imaging was performed within the first week; these follow-up studies were repeated every 3 months during the first 6 months and every 6 months thereafter. Venography was performed only in case of clinical deterioration or nondiagnostic US and color Doppler studies. All patients underwent anticoagulation to a target International Normalized Ratio of 2.5–3.

CASE REPORTS

Case 1

A 20-year-old man presented with moderate ascites, hepatomegaly, jaundice, and hemorrhoidal bleeding for 3 months duration. At the time of presentation, his liver function test results were as follows: serum bilirubin, 2.5 mg/dL (normal range, 0.5–1 mg/dL); total protein, 7.5 g/dL (normal range, 6–8.5 g/dL); serum albumin, 3.6 g/dL (normal range, 3.5–5 g/dL); aspartate aminotransferase (AST), 120 U/L (normal range, 8–40 U/L); alanine aminotransferase (ALT), 158 U/L (normal range, 5–35 U/L); and alkaline phosphatase (ALP), 101 U/L (normal range, 40–125 U/L). After the standard routine imaging studies, he was diagnosed with the type 2 pattern of BCS and was subjected to right hepatic vein recanalization, which was unsuccessful. Further imaging studies confirmed the presence of suitable intrahepatic collateral vessels, of which recanalization was attempted via a right internal jugular venous approach. Angioplasty of the stenotic ostium was undertaken with a 7-mm × 4-cm angioplasty balloon catheter (ATB; Cook). The collateral vessel was seen to share the ostium with the accessory right hepatic vein. Repeated angioplasty performed at the ostium showed resistant residual stenosis, so a self-expanding 8-mm × 4-cm stent (Zilver; Cook) was placed across the collateral ostium, with good recanalization. He showed good resolution of ascites over a period of 7 days.
At 44-month follow-up he remained free of symptoms. His liver function test results at this time were as follows: serum bilirubin, 0.5 mg/dL; total protein, 8.6 g/dL; serum albumin, 4.9 g/dL; AST, 18 U/L; ALT, 15 U/L; and ALP, 57 U/L.

Case 2

A 22-year-old man presented with fever for 3 days, abdominal discomfort, moderate ascites, jaundice, and one episode of hemorrhoidal bleeding. His liver function test results were as follows: serum bilirubin, 1.1 mg/dL; total protein, 7.5 g/dL; serum albumin, 4 g/dL; AST, 55 U/L; ALT, 50 U/L; and ALP, 263 U/L. After his diagnosis was confirmed and the sites of hepatic venous occlusions were established (type 2 pattern), right hepatic vein recanalization was attempted, which was unsuccessful. He was noted to have adequate intrahepatic collateral vessels in the right lobe of liver with a short segment ostial occlusion suitable for recanalization. A percutaneous transhepatic puncture of the collateral vessel was performed under US guidance to study the collateral hemodynamics and the orientation of its ostium with the IVC to plan the venous approach for recanalization. In view of the downward orientation of the collateral vessel, a right femoral venous approach was used for recanalization. As the occluded segmental of the ostium was resistant to repeated angioplasty, a 9-mm \( \times \) 4-cm self-expanding stent (Zilver; Cook) was placed across the stenosis, with good recanalization of the collateral ostium.

The first screening US at 1 week showed good resolution of ascites (ie, minimal residual ascites), and the patient’s recovery was uneventful. At 42-month follow-up, the patient was free of symptoms and his liver function test results were as follows: serum bilirubin, 0.6 mg/dL; total protein, 9.2 g/dL; serum albumin, 5.2 g/dL; AST, 39 U/L; ALT, 40 U/L; and ALP, 114 U/L.

Case 3

A 17-year-old male patient presented with jaundice for 4 years, hematemesis, and abdominal distention for 3 months. He had been treated by variceal ligation and glue injection at another institution. He was diagnosed to have the type 2 pattern of BCS based on imaging studies. Preprocedural US and color Doppler imaging showed ostial occlusion of the right hepatic vein and large hepatic collateral vessels. His liver function tests were as follows: serum bilirubin, 2.6 mg/dL; total protein, 6.8 g/dL; serum albumin, 3.3 g/dL; AST, 50 U/L; ALT, 31 U/L; and ALP, 48 U/L. Right hepatic vein recanalization via a right internal jugular venous approach was unsuccessful, so intrahepatic collateral recanalization was attempted via a right femoral venous approach. The recanalized intrahepatic collateral vessel was seen to share a common ostium with the caudate lobe vein. The patient had good resolution of ascites at the time of discharge from the hospital 10 days after the procedure. He was prescribed oral anticoagulation treatment after the procedure, with a therapeutic International Normalized Ratio.

The patient remained asymptomatic for 14 months before a recurrence of ascites and variceal bleeding. Venography showed an adequately patent

Figure 3. Percutaneous transhepatic collateral injection in patient 2 opacifying multiple dilated intrahepatic collaterals with occluded ostium (1) with the IVC. The distal end of the introducer (via the right internal jugular vein) is seen in the region of the IVC.

Figure 4. Venogram from the right femoral venous approach in patient 3 shows the stenosed ostium (arrowhead) of intrahepatic collateral vessels (arrow) with retrograde opacification.
ostium of the previously recanalized intrahepatic collateral vessel. An IVC stenosis was suspected, which was treated by angioplasty and stent placement, with little clinical improvement. Left hepatic vein recanalization was attempted, which was unsuccessful. In view of the recurrence of symptoms and unsuccessful hepatic vein recanalization, a modified transjugular intrahepatic portosystemic shunt (TIPS) procedure was planned. After the TIPS procedure, he showed significant clinical improvement (ie, ascites regressed, no further variceal bleeding) and remained well clinically and radiologically at follow-up 20 months after the TIPS procedure. His liver function test results at the last follow-up were as follows: serum bilirubin, 1.6 mg/dL; total protein, 8.6 g/dL; albumin, 4.5 g/dL; AST, 78 U/L; ALT, 53 U/L; and ALP, 85 U/L.

Case 4

A 22-year-old man presented with jaundice, abdominal pain, ascites, and fever for 3 months. He was treated in another hospital by IVC angioplasty, with no clinical improvement. His liver function test results at presentation were as follows: serum bilirubin, 3.3 mg/dL; total protein, 6.8 g/dL; albumin, 3.1 g/dL; AST, 56 U/L; ALT, 37 U/L; and ALP, 179 U/L. After routine imaging studies at our hospital, he was found to have the type 2 pattern of BCS. Recanalization of the right and middle hepatic veins was attempted, but was unsuccessful. On screening US and color Doppler studies, the patient was found to be a suitable candidate for intrahepatic collateral vessel recanalization. Recanalization by angioplasty of the intrahepatic collateral vessels, which shared a common ostium with the right accessory hepatic vein, was performed via a right internal jugular venous approach and was uneventful. At 7-month follow-up the patient had mild icterus and was otherwise free of symptoms. He had gained 4.5 kg in the interim. His liver function test results at follow-up were as follows: serum bilirubin, 1.8 mg/dL; total protein, 7.9 g/dL; albumin, 4.4 g/dL; AST, 40 U/L; ALT, 31 U/L; and ALP, 118 U/L.

DISCUSSION

Formation of intrahepatic venovenous collateral vessels in patients with hepatic venous obstruction is the body’s own compensatory mechanism to decompress the liver, and if these collateral vessels are adequate, it can explain the absence of symptoms in these patients (8). The significance of intrahepatic collateral vessels in the treatment of patients with BCS was mentioned by Baijal et al (7), who observed in patients with combined hepatic vein and IVC obstruction that recanalization of the IVC alone can be an effective treatment option if at least one large hepatic vein or collateral vessel opens into the IVC below the level of its obstruction.

In the present larger series of BCS, two patients were incidentally detected to have hepatic venous obstruction and a confirmatory venogram documented well formed intrahepatic venovenous collateral vessels, suggesting adequate venous drainage by these vessels (Fig 8). In these asymptomatic patients with adequate intrahepatic collateral vessel formation, we believe a repeat thrombotic
episode resulting in ostial compromise could render BCS symptomatic.

Endovascular recanalization of the compromised ostium of these collateral vessels indirectly facilitates the body’s own compensatory mechanism, which is our hypothesis to justify the performance of these recanalization procedures. These intrahepatic collateral vessels reroute the obstructed hepatic venous system into the IVC and share a common ostium with an accessory hepatic vein, providing a venovenous collateral vessel formation (3). In the present series, the recanalized intrahepatic collateral vessels shared the ostium with the right accessory hepatic vein in three patients and the caudate lobe vein in one.

A good understanding of this compensatory intrahepatic collateral vessel formation and its mapping with US and color Doppler studies are key factors in the consideration of this recanalization option for suitable patients. A case-by-case approach is necessary because the incidence of accessory hepatic veins, as well as the varying extent of intrahepatic collateral vessel formation, makes this recanalization procedure suitable only for select patients.

In our recent practice we have attempted collateral vessel recanalization in suitable patients in cases of failed major hepatic vein recanalization, as well as reserve endovascular cavoportal shunt creation (ie, modified TIPS) for collateral vessel recanalization failures (11), as in case 3 described here. The theoretical advantages of collateral vessel recanalization versus a modified TIPS procedure are the lack of deprivation of portal blood to the liver, lack of alteration of hepatic hemodynamics (12), and no risk of hepatic encephalopathy.

The limitation of this procedure is that it is feasible only in cases of adequate intrahepatic collateral vessel formation with ostial stenosis/occlusion and a failed standard hepatic vein recanalization procedure; this represents just 5.1% of patients at our institution (four of 78). Larger study groups and long-term follow-up are needed to validate the long-term outcome of this recanalization procedure.

In conclusion, understanding intrahepatic collateral vessel formation in patients with BCS is not only essential in understanding the evolution of the disease but also important in planning suitable endovascular recanalization procedures. Intrahepatic collateral vessel recanalization is a promising treatment option in suitable patients with failed hepatic vein recanalization, and is deserving of further study.

References


