



Direct intrahepatic portocaval shunt for treatment of portal thrombosis and Budd-Chiari syndrome

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ABSTRACT

Budd-Chiari syndrome (BCS) refers to hepatic venous outflow obstruction that in severe cases can lead to acute liver failure prompting consideration of revascularization or transplantation. Here, a 22 year old female with angiographically proven BCS secondary to JAK2/V617F positive *Polycythemia vera* on therapeutic warfarin presented with acute liver failure (ALF). Imaging revealed a new, near complete thrombotic occlusion of the main portal vein with extension into the superior mesenteric vein. An emergent direct intrahepatic portocaval shunt (DIPS) was created and liver function promptly normalized. She has been maintained on rivaroxaban since that time. Serial assessment over 1 year demonstrated continued shunt patency and improved flow in the mesenteric vasculature on ultrasound as well as normal liver function. DIPS is a viable alternative in the treatment of ALF from BCS when standard recanalization is not feasible. Improved blood flow may also improve portal/mesenteric clot burden. While further investigation is needed, new targeted anticoagulants may be viable as a long term anticoagulation strategy.

Key words. Hepatic vein thrombosis. BCS. DIPS. Acute liver failure.

INTRODUCTION

Budd-Chiari syndrome (BCS) is an eponym describing any condition of hepatic venous outflow tract obstruction, regardless of mechanism. In patients with refractory ascites, direct intrahepatic portocaval shunt (DIPS) is an alternative to transjugular intrahepatic portosystemic shunt (TIPS) in which the caudate lobe is used to create a side-to-side shunt. Shunting is indicated in BCS when patients do not improve with medical therapy but is generally applied before more severe disease/acute liver failure develops.¹ Irrespective of vascular intervention, lifelong anticoagulation with warfarin is recommended in those with a demonstrable hypercoagulable state.² Here, a young female with acute BCS refractory to medical therapy and further complicated by mesenteric thrombosis was successfully treated with direct intrahepatic portocaval shunt (DIPS) creation.

CASE REPORT

A 22-year-old female without past medical history presented with one month of increasing abdominal swelling and two days of nausea and vomiting. Initial exam revealed tachycardia with pulse ranging 90–110, *scleral icterus*, jaundice, and distended abdomen with right upper quadrant tenderness and positive fluid wave. She had no asterixis and was alert and oriented to person, place, and time. Initial labs were significant for white blood cell count of 20,400 cells/ μ L, hemoglobin 162 g/L (16.2 g/dL), platelets 534,000 units/ μ L, international normalized ratio (INR) 1.4, aspartate aminotransferase 200 IU/L, alanine aminotransferase 206 IU/L, and total bilirubin 23.9 μ mol/L (1.9 g/dL).

A bedside ultrasound confirmed the presence of ascites, and a diagnostic paracentesis revealed a serum-ascites albumin gradient of 1.9, protein of 3.1, and no evidence of spontaneous bacterial peritonitis. Magnetic

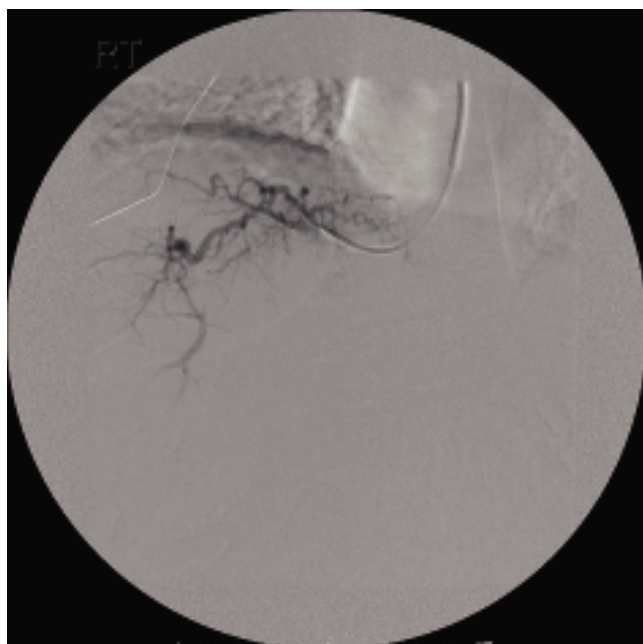


Figure 1. Hepatic venogram. This hepatic venogram demonstrates absent hepatic veins, which have been replaced by a web-like network of venous collaterals characteristic of Budd-Chiari syndrome.

resonance imaging of the abdomen demonstrated hepatomegaly with absence of blood in the hepatic vein and an enlarged caudate lobe with enlarged collaterals. The portal vein was patent at this time. A hepatic venogram corroborated hepatic outflow tract obstruction, confirming the diagnosis of Budd-Chiari syndrome (Figure 1).

During the hepatic angiogram, several attempts were made to catheterize the right and middle hepatic veins, but only small collateral veins to the inferior vena cava (IVC) were identified. There was no normal-appearing hepatic vein, nor any short segment amenable to recanalization or creation of a TIPS. A liver biopsy was also performed, demonstrating marked centrilobular congestion with focal hepatocellular degeneration, atrophy, and central vein fibrosis (Figure 2). Workup from common viral, autoimmune and inherited metabolic etiologies of chronic liver disease was negative; however, hypercoagulable work-up revealed JAK2/V617F mutation. This mutation in the presence of an elevated hemoglobin was diagnostic of *Polycythemia vera*. She was initiated on low molecular weight heparin (enoxaparin) bridge to warfarin and discharged with a therapeutic INR of 2.5. Her mentation was clear, and her bilirubin had trended down to 18 $\mu\text{mol/L}$ (1.1 g/dL). She remained well over the following month with weekly monitoring of her INR (ranging from 2.5 to 2.8). Her hemoglobin on discharge was 125 g/L (12.5 g/dL). Her ascites was well controlled with spironolactone 100 mg daily and furosemide 40 mg daily.

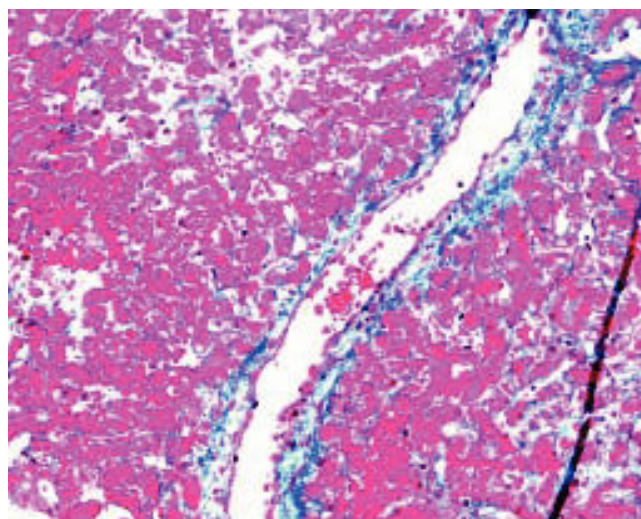


Figure 2. Liver biopsy. This liver biopsy demonstrates centrilobular congestion with focal hepatocellular degeneration, atrophy and central vein fibrosis.

She returned to the hospital 3 weeks later with abrupt onset of diffuse abdominal pain and disorientation. She was afebrile with pulse 80 beats per minute and blood pressure 124/82 mmHg. Initial laboratories included hemoglobin of 153 g/L (15.3 g/dL), alanine aminotransferase 1,638 IU/L, aspartate aminotransferase 1,267 IU/L, bilirubin 42.8 $\mu\text{mol/L}$ (2.5 g/dL), and INR of 11.4. She was admitted for observation and given a dose of vitamin K 10 mg IV with no change in INR over 48 h. Doppler ultrasonography of the liver demonstrated absent blood flow in the hepatic veins but visualized a new eccentric, nearly occlusive thrombus in the main portal vein (mean systolic velocity: 8 cm/sec) and distal superior mesenteric vein. The patient grew increasingly lethargic and began to display asterixis. She underwent evaluation for urgent shunt placement with simultaneous listing for orthotopic liver transplantation. A decision was made to proceed with the creation of a DIPS.

Using ultrasound guidance, transjugular access was established and a 10-French angle tipped sheath was advanced over a 0.035 inch working wire to the IVC. Guided by a transfemoral intravascular ultrasound catheter positioned in the IVC, a Colapinto needle was guided from the IVC through the caudate lobe and into the cephalad portion of the main portal vein, just below the main portal vein bifurcation. A wire was then advanced into the portal vein as well and the needle removed (Figure 3A). Pressures were measured in the main portal vein (28 mmHg) and right atrium (5 mmHg). The intrahepatic tract was dilated to 6 mm with a balloon catheter. The 10-French sheath was then passed through the intrahepatic tract to the portal vein. A Viatorr 10 mm x 6 cm TIPS (Gore

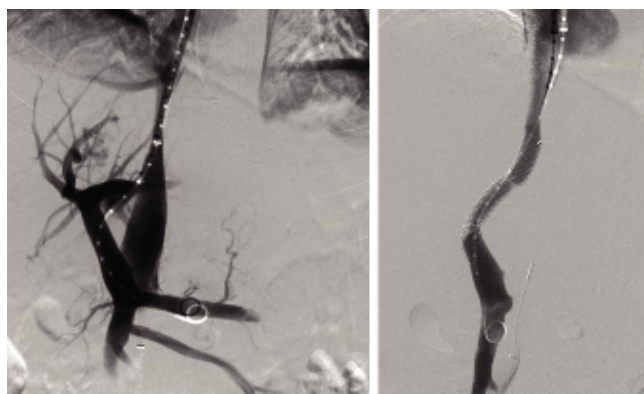


Figure 3. Direct intrahepatic portocaval shunt (DIPS) creation. **A.** Simultaneous portal venogram and cavogram show transcaval access into the main portal vein in preparation for DIPS placement. There is moderate narrowing of the intrahepatic inferior vena cava (IVC) due to liver hypertrophy. **B.** Portal venogram after DIPS placement shows brisk hepatopedal flow through the shunt into the IVC.

medical, Flagstaff, AZ) covered stent was then deployed (Figure 3B). The stent was balloon dilated to 8 mm. Completion pressures in the main portal vein and right atrium were 16 and 11 mmHg, respectively.

The day following DIPS creation, the patient's mentation began to improve and INR trended down. By post-procedure day two, the INR decreased to 2.5, her mentation returned to normal, her abdominal discomfort resolved. Given the formation of a portal vein thrombus despite systemic anticoagulation on warfarin, the patient was discharged on rivaroxaban for long-term anticoagulation four days after her procedure. At one year follow up, the patient is well with normal aminotransferases and INR of 1.2. Doppler ultrasonography at 1 year follow up shows a patent DIPS shunt and improved patency of the portal vein (mean systolic velocity: 54 cm/sec).

DISCUSSION

BCS is characterized by the abrupt onset of right upper quadrant pain and ascites. Patients with severe BCS can develop acute liver failure with coagulopathy and encephalopathy. Disease severity is estimated by the Rotterdam score determined as shown in figure 4.

Encephalopathy and ascites are scored as either 0 (absent) or 1 (present), prothrombin time as 0 (INR ≤ 2.3) or 1 (INR > 2.3), and bilirubin in $\mu\text{mol/L}$. A score 0 to 1.1 por-

tends good prognosis (class I, five year survival 89%); between 1.1 and 1.5: intermediate prognosis (class II, five year survival 74%); and < 1.5 : poor prognosis (class III, five year survival 42%).¹ In this case, her initial Rotterdam score of 1.14, intermediate prognosis, increased to 3.20, poor prognosis, and highlighted the need for urgent intervention.³

For patients in need of intervention, recanalization is the least invasive approach and is usually first attempted. In Asia, where suprahepatic stenosis of the IVC is more common, recanalization comprises 98.62% of all interventional radiology procedures performed for BCS.² In Western nations, hypercoagulable states often lead to complete hepatic vein occlusion, making recanalization less feasible (success rates around 20%).⁴ The TIPS is most commonly employed, whereby a shunt is created from the hepatic vein, through the liver parenchyma, and into the portal vein. Liver transplantation can be considered for those who continue to develop progressive liver injury.⁵

Occasionally, as in this case, the hepatic vein is entirely occluded and inaccessible to either recanalization or creation of a TIPS. In such instances, a DIPS (portal-caval shunt through the caudate lobe lying between the inferior vena cava and the portal vasculature) can be employed.⁶ Classically, DIPS is performed via a transjugular approach using endovascular ultrasound.⁷ DIPS has been created successfully through percutaneous puncture of the portal and caval system as well.^{8,9} In the majority of these cases, however, DIPS was employed in patients with subacute and chronic BCS with preserved liver function. Recently, indications for shunting in BCS have been expanding. A recent study examined 52 patients with severe BCS (Rotterdam class III) of whom 18 underwent surgical shunting and 14 underwent liver transplantation. No significant differences in survival existed between groups.¹⁰ Several cases and small series have suggested DIPS as a viable option for patients with acute and hyperacute BCS.^{11,12}

In a patient with stable hepatic function, a trial of anticoagulation is recommended before any of invasive interventions are attempted. Current recommendations call for the use of warfarin to maintain an INR between 2 and 3.⁵ As aforementioned, in Western nations where thrombosis is more common than stenosis, anticoagulation alone may be a durable treatment strategy.⁴ Little is known about the efficacy of target specific anticoagulants such as dabigatran, rivaroxaban, and apixaban, in the long term management of

$\text{Rotterdam score} = 1.27 \times \text{encephalopathy} + 1.04 \times \text{ascites} + 0.72 \times \text{prothrombin time} + 0.004 \times \text{bilirubin}$

Figure 4. Disease severity is estimated by the Rotterdam score.

BCS. A single case report describes the successful use of rivaroxaban in a patient with polycythemia secondary to elevated erythropoietin levels.¹³ All three of anticoagulants are metabolized by various cytochrome p450 isoforms in the liver while rivaroxaban is the only one primarily eliminated by the kidneys.¹⁴⁻¹⁶

This case was complicated by an acute portal vein thrombosis which formed despite first line therapy with systemic anticoagulation.¹⁷ We hypothesize that acute portal vein thrombus may have precipitated liver failure by further reducing inflow to an already compromised liver with outflow block. Endovascular ultrasound was used to create a DIPS simultaneously with listing for liver transplantation. We postulate that restoration of portal blood flow facilitated partial dissolution of portal vein thrombus. The patient was switched to rivaroxaban for long term anticoagulation strategy because of documented progression of thrombus on previous warfarin based therapy. Over 1 year of follow up, the patient has continued on rivaroxaban and with serial phlebotomy [goal hemoglobin of 130 g/L (13 g/dL)]. Doppler ultrasound at one year showed increased portal vein blood flow and the patient remains in good health with normal liver function.

This case provides evidence that shunting via DIPS may be viable for patients when hepatic blood flow is further compromised by portal vasculature thrombosis. For patients who either have a contraindication to warfarin or who have progressive disease despite therapeutic INR, rivaroxaban may be a reasonable alternative anticoagulant. Further studies are needed to clarify the role of target specific anticoagulants in the treatment of Budd-Chiari syndrome.

ABBREVIATIONS

- **BCS:** Budd-Chiari syndrome.
- **DIPS:** direct intrahepatic portocaval shunt.
- **INR:** Internationalized Normalized Ratio.
- **IVC:** inferior vena cava.

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