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CASE REPORT

Direct intrahepatic portocaval shunt as a salvage strategy in Budd–Chiari syndrome

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Summary

Budd–Chiari syndrome (BCS) remains a rare disease with varied presentations. It follows hepatic venous outflow obstruction and can be considered primary or secondary, depending on the obstruction's aetiology. The median age at diagnosis is approximately 35 years, and diagnoses are often delayed. Proper diagnosis is crucial to enable a full spectrum of treatment options, including interventional radiology, to improve outcomes.[1-3]

Case

Ms NN was initially referred in 2003 with hepatosplenomegaly of unknown cause. She was human immunodeficiency virus (HIV)-positive and remained virologically suppressed on a variety of antiretroviral therapy regimens over > 20 years. In 2003, an initial, extensive investigation, including two liver biopsies and imaging, was non-contributory to a clear diagnosis. Follow-up was planned, but she disengaged from care and was not seen for ~ 15 years between 2005 and 2021. Her antiretroviral therapy continued. She consulted dermatology in the interim and was diagnosed with bullous linear immunoglobulin A (IgA) disease of the skin, for which she received doxycycline, prednisone, and dapsone at different intervals.

The patient was again referred in December 2021, following a first episode of hepatic decompensation. She presented to her local hospital with a month-long history of jaundice and recent-onset ascites. At this time, she was diagnosed with advanced chronic liver disease with evidence of clinically significant portal hypertension. Her complications included jaundice, coagulopathy, hypersplenism, and newly described portal and splenic vein thromboses. Autoantibodies, including antinuclear antibody (ANA), anti-smooth muscle antibody (Ab), and anti-liver/kidney microsomal 1 antibody (LKM1 Ab), were all negative, but serum IgG was raised at 33 g/L. Liver biopsy was repeated, suggesting chronic venous outflow obstruction. A diagnosis of chronic BCS was made. A thrombophilia workup excluded a myeloproliferative disorder, but a combined protein C and S deficiency was documented. Gastroscopy demonstrated grade 2–3 oesophageal varices requiring banding. She was initiated on nonselective beta blockers and diuretics, with paracentesis.

Post-discharge, she again disengaged from care until a second decompensation in November 2022, when she required admission and treatment for spontaneous bacterial peritonitis (SBP). *Escherichia coli* was cultured, and she responded well to appropriate antibiotics. After this, repeated admissions for decompensation followed, and imaging was concerning for progression of previously documented portal vein (PV) thrombosis, now probably extending into the superior mesenteric vein (SMV) and splenic vein (Figures 1 and 2). Repeated visits for large-volume paracentesis ensued despite diuretic and salt intake management. Diuretic dose-up titration was limited by electrolyte derangement and worsening creatinine. The patient was evaluated for intervention, and the decision was discussed at length with interventional radiology. Her Child–Pugh score was B8 at baseline with a Model for End-Stage Liver Disease Sodium score (MELD-Na) of 15.

Interventional radiology

Following a multidisciplinary team meeting, it was agreed to offer Ms NN the option of attempting to recanalise the PV and placing a transjugular intrahepatic portosystemic shunt (TIPS). The patient consented. However, during the procedure, after multiple failed attempts, it was deemed infeasible. Although not anticipated, the SMV was fortunately recanalised (Figures 3 and 4). Due to the caudate lobe enlargement and the balloon's position, a transcaval direct intrahepatic portocaval shunt (DIPS) procedure was attempted. This was decided after gaining access to the right hepatic vein (HV) and being unable to puncture the balloon in the SMV/splenic vein due to the distance between the structures (Figures 5 and 6).

After a subhepatic transcaval puncture, a wire was snared through the transsplenic approach and a floss created (Figures 7 and 8). A Viatorr stent was urgently deployed to tamponade the puncture in the subhepatic inferior vena cava (IVC), and a balloon was also used to prevent haemorrhage into the peritoneum (Figure 9). Once the Viatorr stent was deployed, a second venous stent was deployed in the SMV, and on portogram, there was good, clear in-line flow of blood with no extravasation of contrast (Figure 10).

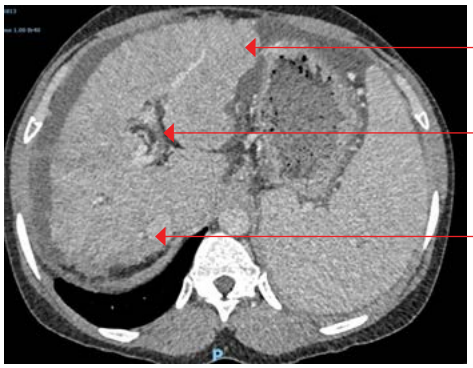

Figure 1

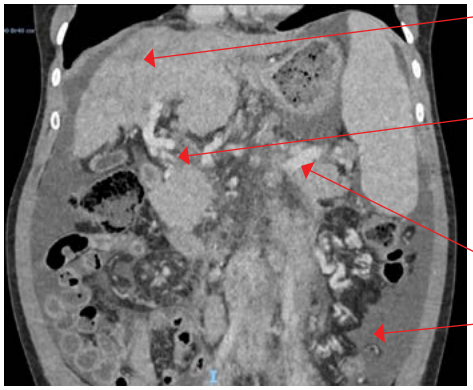

Figure 2

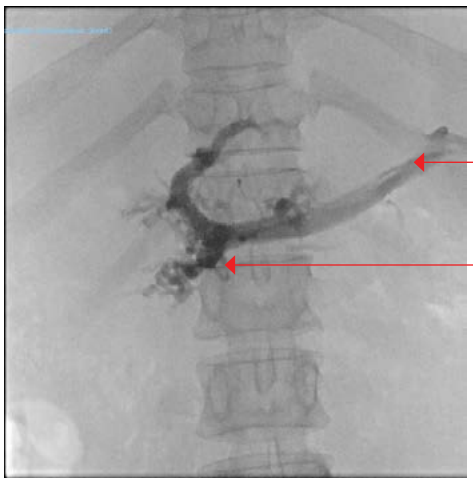

Figure 3

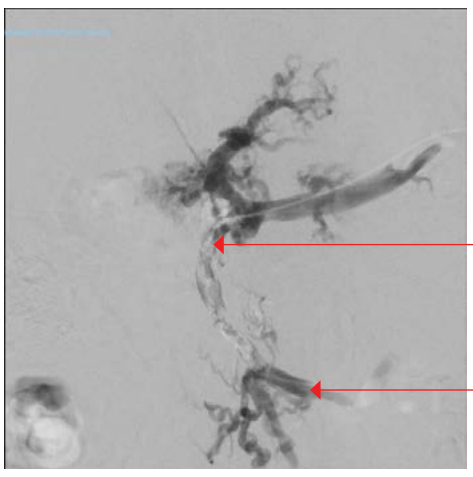

Figure 4

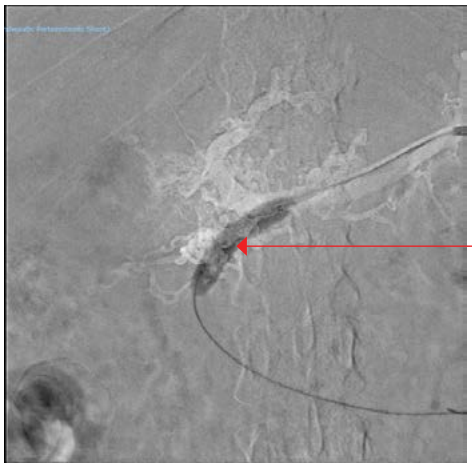

Figure 5

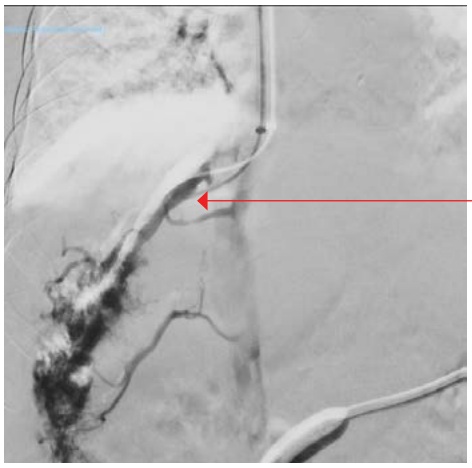

Figure 6

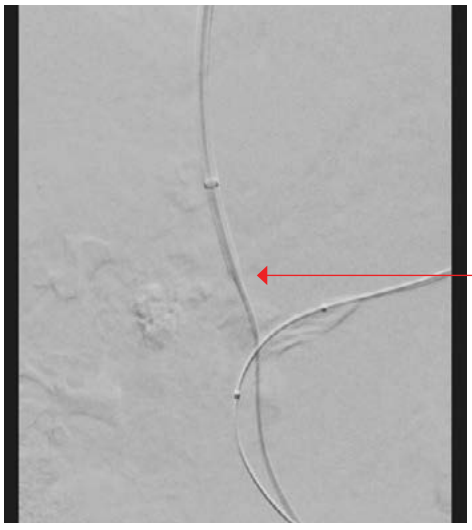

Figure 7

Despite a turbulent post-procedural course, the patient did extremely well. No further paracentesis was required. Expectedly, grade 1 (at most 2) hepatic encephalopathy (HE) became evident, and she responded well to a combination of lactulose and rifaximin. Computed tomography venogram six months post-procedure confirmed patency of the DIPS and SMV stent (Figures 11 and 12). Similarly, gastroscopy performed five months post-DIPS confirmed the absence of oesophageal or gastric varices.

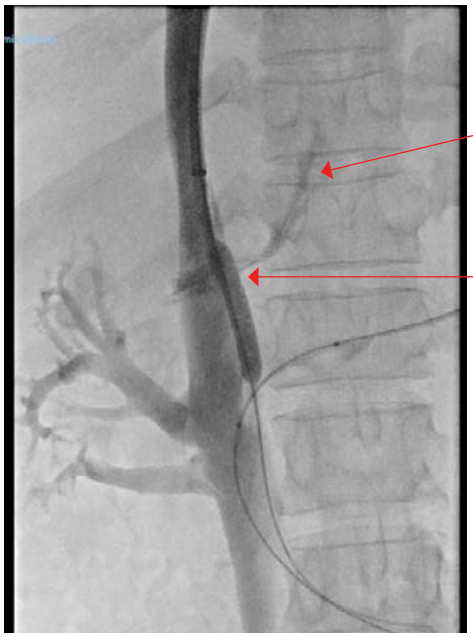

Figure 8

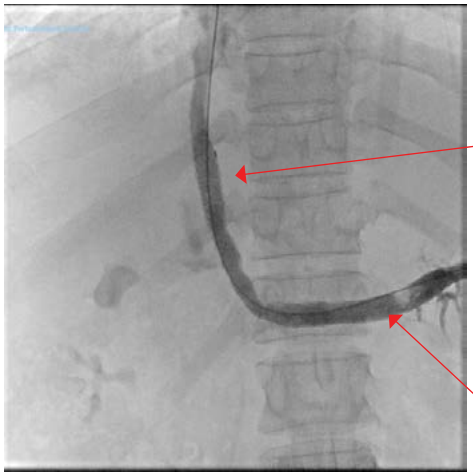

Figure 9

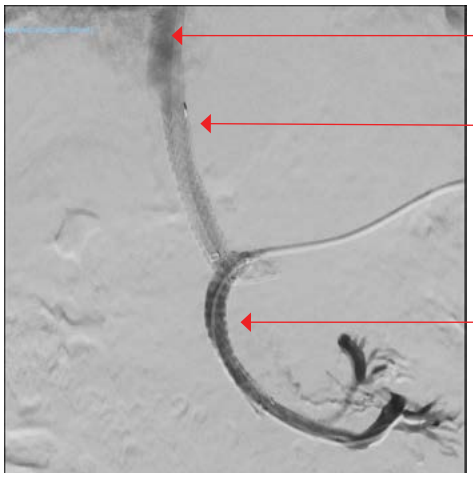

Figure 10

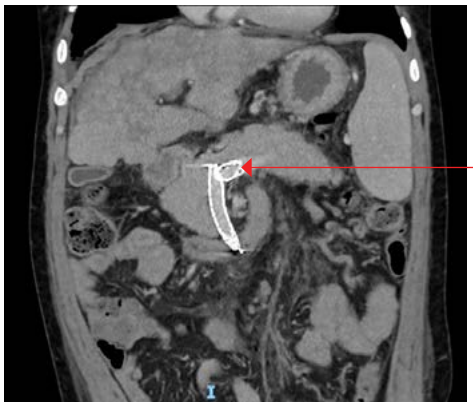

Figure 11

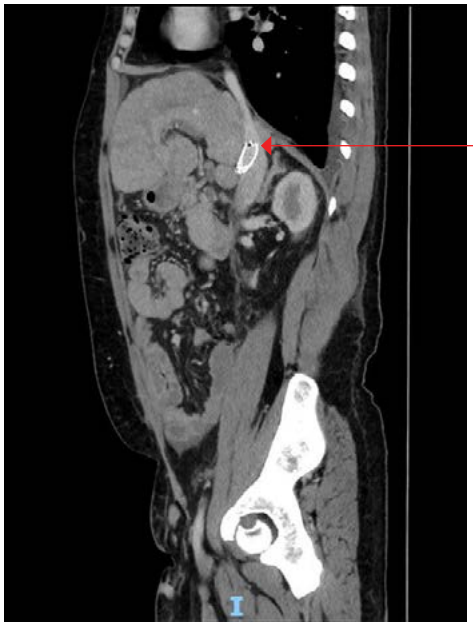

Figure 12

Discussion

BCS is a rare disease with a prevalence of 11 cases per million and an annual incidence of one per million.[1,3] Minimal data exists for sub-Saharan Africa and is largely limited to case reports. The median age at diagnosis is 35–40 years, and both sexes are equally affected.[3] Despite its rarity, BCS should be considered as part of the differential diagnosis in all patients with liver disease or failure of unknown cause – including acute, subacute, and chronic presentations – and systematically sought.[1]

BCS may be primary or secondary and includes any hepatic venous outflow obstruction from the confluence of the IVC and right atrium to the HVs. It is important to note that the definition excludes sinusoidal obstruction syndrome and hepatic outflow obstruction secondary to right-sided or congestive cardiac disease or pericardial disease.[1-3] BCS is considered primary if the obstruction is due to an intraluminal thrombus or primary disease of the vessel wall. It is considered secondary if the hepatic venous outflow is obstructed secondary to extrinsic compression, such as a tumour compression of the vessel wall.[2,4] Primary BCS is the focus for the remainder of this discussion. However, one should note that membranous obstruction of the inferior vena cava (MOIVC) is a distinct clinical entity and, while rare in many countries, does occur more frequently in eastern countries, such as Nepal, China, Japan, India, Korea, and

South Africa, and should be identified as treatment options and outcomes may differ.[5]

Regardless of the cause of the hepatic venous outflow obstruction, the resultant effects are largely the same. There is increased hepatic sinusoidal pressure and consequent portal hypertension with varying degrees of ascites, gastroesophageal varices, and HE. Venous congestion leads to initial ischaemic damage of the perisinusoidal hepatocytes and, eventually, nodular regeneration, fibrosis, and, ultimately, cirrhosis.[2] The presentation is highly varied and depends on the onset speed, extent of obstruction, and the presence of venous collaterals to decompress the portal system. Presentation is traditionally described as fulminant, acute, subacute, or chronic; however, histological findings often indicate significant fibrosis and even cirrhosis despite the acuity of the initial presentation.[2]

Geographical differences between Western and Eastern patients have been well described. While Western cohorts generally have HV obstruction secondary to thrombophilia, cohorts from the East and South Africa report a significantly higher rate of IVC obstruction due to endoluminal abnormalities and venous webs. In countries where MOIVC occurs more frequently, it is the most common cause of hepatic venous outflow obstruction.[5] This may be related to environmental factors, including poverty and higher infection rates.[2,4] Despite these regional differences, it is imperative that patients presenting with BCS undergo a thorough search for underlying thrombophilia.[1-3]

A hereditary or acquired hypercoagulable state is found in approximately 75% of patients, and two or more disorders are identified in 25–46%.[2,3] A reasonable initial screen should include testing for a Janus kinase 2 (JAK2) mutation associated with multiple myeloproliferative disorders, an antiphospholipid syndrome screen, and flow cytometry for paroxysmal nocturnal haemoglobinuria. Protein C and S and antithrombin III (AT III) deficiency, as well as a Factor V Leiden mutation, are also important.[2,3] Notably, the liver produces protein C and S and AT III; therefore, diagnosis based on plasma levels in the setting of liver dysfunction and thrombus may be difficult. Levels well below 10–20% of normal values, or low levels in close family members, would support the diagnosis of an acquired deficiency in the index patient.[2]

The classic presentation of new-onset abdominal pain, hepatomegaly, and recent-onset ascites attributed to acute BCS is rare, and some patients may even be asymptomatic at diagnosis. Many may present with "new"-onset liver dysfunction only to find they have decompensated, advanced chronic liver disease, often due to a new thrombus superimposed on previous obstruction.[3] Large cavocaval collaterals running posteriorly on the trunk are one of the few specific clinical signs of IVC obstruction, which may be a clue to MOIVC diagnosis in a subset of patients.[5] Laboratory values are highly variable and also depend on the rapidity of onset and the degree of underlying liver disease.[3] Doppler ultrasound is considered the initial investigation of choice with a high sensitivity and specificity in identifying hepatic venous outflow obstruction.[1,2,4]

Computed tomography and magnetic resonance imaging may be used to confirm the diagnosis in cases of uncertainty and to evaluate the anatomy to plan further interventions. They may also be necessary to evaluate for suspicion of hepatocellular carcinoma, particularly in patients with large regenerative nodules. Liver biopsy is generally unnecessary but may be required in those with small HV BCS, as the larger vessels may still appear patent on imaging.[1] Diagnosing MOIVC requires inferior vena cavography, which can provide information on the obstruction, the involvement of the HVs, and aid in planning specific treatment.[5] Finally, it is important to note that imaging should comment on the PV, as more than 20% of patients will have combined HV and PV thrombosis at diagnosis.[3,6]

Due to the rare nature of BCS, large randomised controlled trials evaluating treatment strategies have not been conducted, and much of the guidance is based on systematic reviews and meta-analyses of case reports and retrospective studies, as well as expert consensus. Untreated, BCS has a high mortality of up to 80% at three years, but this can be dramatically altered with diagnosis and treatment, resulting in five-year survival rates of 80–90%.[2,3,7] Therapeutic management options consist of medical management, including anticoagulation, angioplasty and stenting where possible, and insertion of a shunt, such as a TIPS or DIPS. Orthotopic liver transplant is reserved for patients who progress despite all alternative treatments or who present with fulminant hepatic failure.[3,7]

Medical management includes anticoagulation for all patients, regardless of whether a specific prothrombotic condition is identified. One of the main aims is to prevent further clot/thrombus propagation, particularly within the portomesenteric system, thereby precluding future options. Therefore, variceal surveillance and eradication, plus nonselective beta blockers, remain paramount for preventing complications of variceal bleeding. Diuretics for ascites are strongly recommended for any patient with portal hypertension. Finally, specific treatment directed at the underlying disease process, such as hydroxyurea in certain myeloproliferative disorders, is important because it improves outcomes.[1-3] Failure of medical management alone is seen in many patients with BCS, necessitating more invasive treatment options.[4,6-8] Those with short-segment stenoses or IVC webs may be candidates for angioplasty/stenting, but the majority of patients will require assessment for a shunt.[1,2,9]

TIPS was first proposed in a 1993 publication. These are percutaneous, image-guided shunts that have essentially replaced surgical shunts due to their significantly better morbidity and mortality.[2,6,7] TIPS patency has also significantly improved since the replacement of bare metal stents with polytetrafluoroethylene-covered stents.[1,2,4] TIPS placement may be technically challenging, and it should be performed in experienced centres, as a direct transcaval approach is required in up to 40% of patients since it is impossible to identify and

enter the remnant HV.[1,3,6] Despite this, complication rates remain relatively low, with five- and 10-year survival rates of 90% and 80%, respectively, and, in experienced hands, technical success is achieved in > 95% of cases.[6] The most common symptomatic indication for TIPS is refractory ascites, followed by variceal haemorrhage.[4] Jaundice, HE, and PV thrombosis are not considered contraindications.[4,6] Notably, TIPS may be considered as a bridge to liver transplant in those presenting with fulminant liver failure.[3,4,7]

The most common post-procedural complications include HE, which may be as high as 20%, and reversible post-procedural bleeding/haemoperitoneum. HE can generally be managed with standard medical treatment, with only a small minority having refractory, unmanageable symptoms. For those patients, TIPS reduction or occlusion may be required, and, ultimately, liver transplant is recommended.[1,4,6] Finally, if TIPS revision is required, it is often relatively easy and safe, with excellent secondary patency rates and durable outcomes.[6]

An ongoing debate surrounding TIPS in BCS is its timing. Many, including the latest European Association for the Study of the Liver (EASL) guidelines published in February 2026, recommend a step-up approach beginning with medical management and considering more invasive strategies only if persistent symptoms of portal hypertension exist; however, some experts recommend a more aggressive approach.[1,2,4,6] Persistent hepatic venous outflow obstruction is thought to drive ongoing ischaemia and, therefore, damage and fibrosis progression. Earlier relief of the obstruction may improve outcomes further and delay or halt liver disease progression before overt signs of portal hypertension appear.[8,10]

The rapid development of interventional radiology over the last two decades has led to significant advances in treatment, improving life expectancy for patients with BCS. TIPS provides an excellent treatment option and, when indicated, should be offered to improve both quality of life and survival in those with BCS. While anticoagulation is almost universally recommended in all patients with BCS, the patient discussed, Ms NN, is currently not on anticoagulation. Her international normalised ratio (INR) remains > 1.65, with a platelet count of 57 × 10^9/L. Importantly, her thromboelastogram demonstrates no procoagulant state, and a risk–benefit analysis was performed. Nonetheless, it is being monitored.

Conclusion

We present a case of chronic BCS with advanced chronic liver disease and clinically significant portal hypertension complicated by extensive portomesenteric thrombosis. Despite this, our patient had a dramatic improvement in her clinical condition with the insertion of a challenging DIPS. She remains well out of the hospital and has been able to partially recompensate despite extremely advanced disease.

Images courtesy of the interventional radiology team

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