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Case Report

Radiology Section

Accessory Hepatic Vein Stenting in the Management of Budd-Chiari Syndrome: A Case Report

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ABSTRACT

The Budd-Chiari Syndrome (BCS) is associated with hepatic venous outflow obstruction. A 22-year-old female patient presented with abdominal pain and backache for 12 days, along with generalised weakness. Physical examination revealed pitting pedal oedema and abdominal distension. An abdominal ultrasound with Doppler Ultrasonography (USG) was performed, revealing liver parenchymal disease with gross ascites and occlusion of all three Hepatic Veins (HV), suggestive of BCS. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) of the liver, as well as a right hepatic venogram using Digital Subtraction Angiography (DSA), showed chronic total occlusion at the ostium. An Accessory Hepatic Vein (AHV) was identified, joining the Right Hepatic Vein (RHV) to the Inferior Vena Cava (IVC). The accessory RHV exhibited high-grade stenosis (approximately 90-95%) at its junction with the IVC. Recanalisation of the AHV (balloon dilatation/stent insertion) was performed. Post-procedure accessory hepatic venogram showed a patent stented segment with a significant reduction in adjacent venous collaterals. No procedure-related complications were observed. The present case highlights the importance of AHV stenting in BCS patients, as it helps maintain normal physiology, in contrast to Direct Intra-hepatic Porto-systemic Shunt (DIPS), which alters normal physiology by allowing portal venous blood to mix directly into the systemic circulation, bypassing the liver parenchyma.

Keywords: Internal jugular vein, Liver, Transjugular intrahepatic porto-systemic shunt

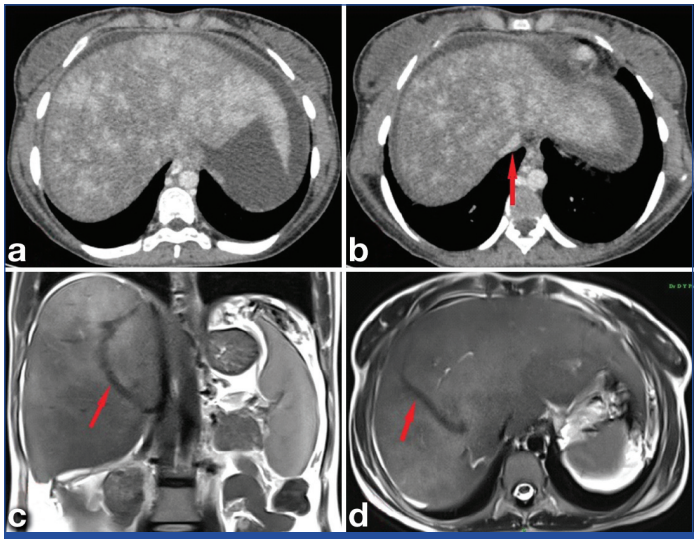
CASE REPORT

A 22-year-old female patient presented to the emergency department with complaints of generalised weakness, pedal oedema, and abdominal pain, which had been ongoing for 12 days and was associated with progressive abdominal distension. The abdominal pain was chronic in nature, a dull ache that was aggravated by walking or coughing. The pain was relieved in a supine position. She also experienced back pain for 12 days. All other systems reviewed were non-contributory. The patient's past medical history, hormonal therapy, abdominal surgeries, smoking, and alcohol drinking were non-contributory.

On examination, the patient appeared pale. Vital signs were as follows: pulse rate of 92/min, blood pressure of 110/70 mmHg, respiratory rate of 20/min, and a normal temperature. Abdominal examination revealed generalised tenderness. Gross ascites and an enlarged tender liver were noted. Basic haematological investigations revealed a haemoglobin level of 9.0 gm/dL, leukocyte count of 3700/mm^3, and platelet count of 206,000/mm^3. The liver enzymes were as follows: Aspartate Aminotransferase (AST) 16 U/L, Alanine Aminotransferase (ALT) 8 U/L, alkaline phosphatase 78 U/L, total bilirubin 0.94 mg/dL, and renal function test (serum creatinine) 0.62 mg/dL.

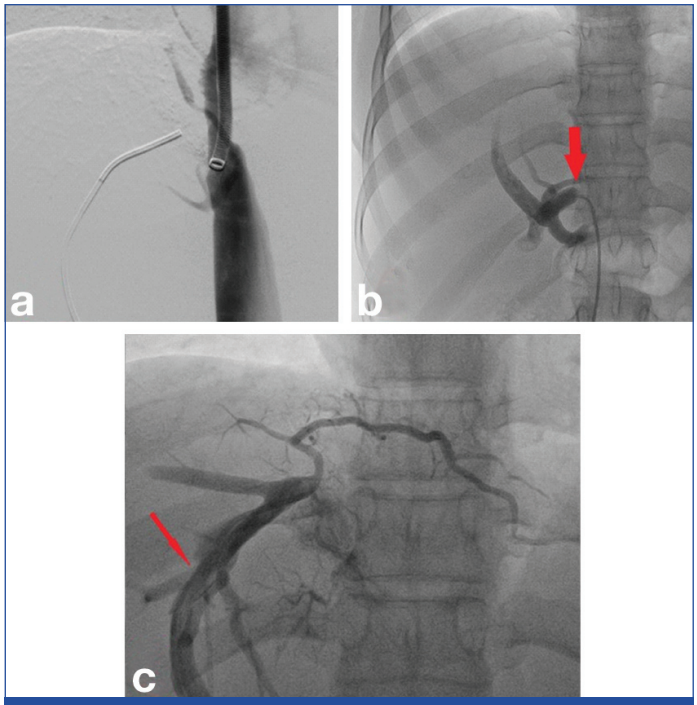
Abdominal and pelvic USG revealed liver parenchymal disease and gross ascites. Contrast-enhanced CT and MRI scans revealed hepatomegaly with a flip-flop enhancement pattern, occlusion of the HV, and a patent AHV joining the IVC [Table/Fig-1]. A right Internal Jugular Vein (IJV) was accessed using a 10F sheath, and the right common femoral vein was accessed using an 8F sheath. A right hepatic venogram showed chronic total occlusion at its ostium. A large AHV was joining the RHV to the IVC. This accessory RHV showed high-grade stenosis (approximately 90-95%) at its junction with the IVC [Table/Fig-2].

The AHV stenosis was crossed using a Terumo 0.035-inch wire and a 4F H1 diagnostic catheter. Balloon angioplasty of the stenosis was performed using an 8×40 mm angioplasty balloon. However, the check venogram showed persistent stenosis. Subsequently, an 8×25 mm express LD stent was deployed across the stenosis [Table/Fig-3]. The stent was post-dilated using an 8 mm balloon inflated up to 12 atm. The post-procedure accessory hepatic venogram showed a patent stented segment with a significant reduction in adjacent venous collaterals [Table/Fig-4]. The procedure was uneventful.

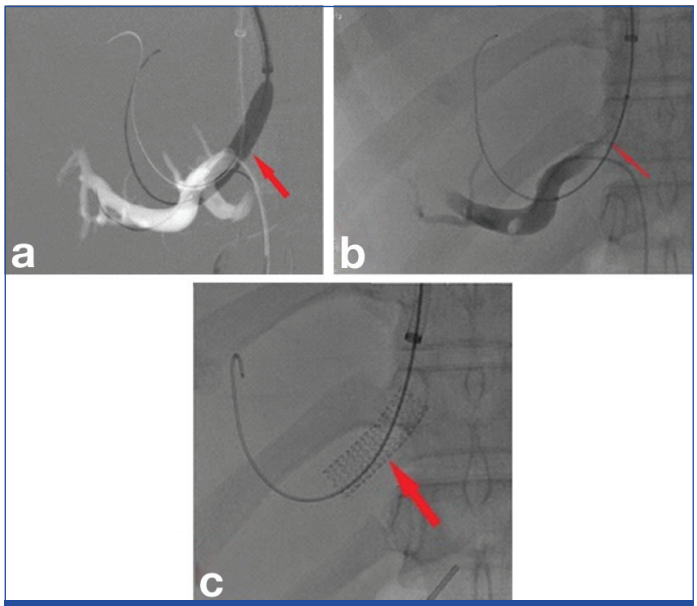


[Table/Fig-1]: Contrast CT images (a and b) in axial plane show flip-flop enhancement of liver with normal IVC (red arrow). No visualisation of Hepatic Veins (HV). Non-contrast MRI images in Coronal (c) and Axial (d) planes show Accessory Hepatic Vein (AHV) (red arrows).

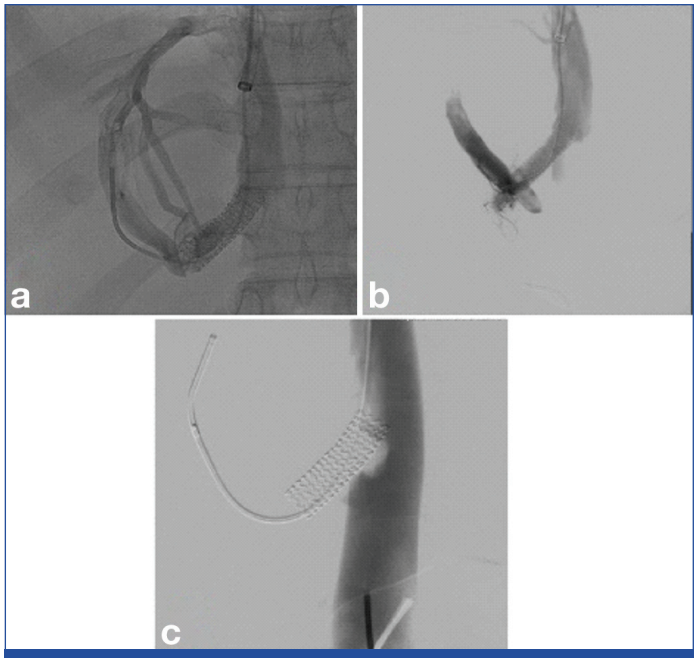
The patient was kept on anticoagulation with heparin-warfarin overlap for three days and subsequently continued on warfarin alone with a target INR of 2.5. After achieving a therapeutic INR range, the patient was discharged in stable condition after seven days. During the six-month follow-up, the patient was doing well with no evidence of ascites. The follow-up Doppler showed a patent stent with good velocity and in-stent flow [Table/Fig-5]. The patient continued anticoagulation therapy with warfarin tablets.



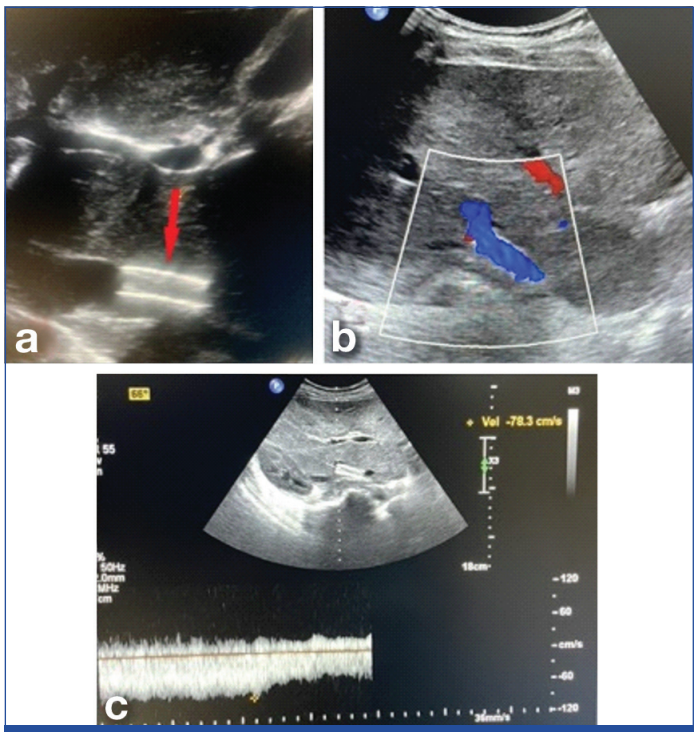
[Table/Fig-2]: Digital subtraction venogram shows normal IVC (a), high grade stenosis at ostium of Accessory Hepatic Vein (AHV) (red arrow in b) and course of Accessory Hepatic Vein (AHV) (red arrow in c).



[Table/Fig-3]: Balloon angioplasty of Accessory Hepatic Vein (AHV) ostium (red arrow in a) Residual stenosis after balloon angioplasty (red arrow in b). Stent across ostium of Accessory Hepatic Vein (AHV) (red arrow in c).



[Table/Fig-4]: a and b) Post-stenting venogram shows good flow across stent in Accessory Hepatic Vein (AHV); c) IVC gram showing dilution of contrast at the level of stent.



[Table/Fig-5]: Six months follow-up USG shows Accessory Hepatic Vein (AHV) stent in-situ (red arrow in a). Colour Doppler images; (b) shows good flow across stent. Spectral waveform (c) shows good velocity flow across stent.

DISCUSSION

BCS is a rare disease characterised by hepatic venous outflow obstruction at the level of the HV or IVC, resulting in portal hypertension [1,2]. The obstruction can be thrombotic or non-thrombotic and can occur anywhere along the venous course, from the hepatic venules to the junction of the IVC and the right atrium. BCS occurs in approximately one in 100,000 of the general population worldwide [3]. Patients may present with acute signs and symptoms such as abdominal pain, ascites, and hepatomegaly, or with more chronic symptoms related to long-standing portal hypertension. A proper clinical history and imaging are essential for a definitive diagnosis [3]. Diagnostic tools commonly used to diagnose BCS include Doppler USG of the liver, MRI, hepatic venography, and liver biopsy [3]. Various treatment options, including Transjugular Intrahepatic Portosystemic Shunt (TIPS), surgical shunts, and liver transplantation, have been described as potential treatment options for BCS [4-7]. Early diagnosis of the disease is crucial for appropriate treatment. More than half of the cases of classic BCS occur between the ages of 20 and 39 years [8]. The proposed etiologies of BCS, as described by Valla DC [3], include multifactorial disease in which several pro-thrombotic conditions, observed in atleast 35% of patients, contribute to the development of thrombosis in an uncommon location such as the HV. Myeloproliferative diseases are observed in approximately 50% of patients with BCS and a mutation in the JAK2 gene. Among other acquired conditions, Behcet's disease, paroxysmal nocturnal haemoglobinuria, antiphospholipid syndrome, oral contraceptive use, and pregnancy have been documented as aetiologies for BCS. In BCS due to Behcet's disease, IVC involvement is more common than HV involvement. Oral contraceptive use is associated with HV involvement rather than IVC involvement. Inherited conditions such as Factor 5 Leiden, protein C deficiency, and protein S deficiency also contribute to the development of BCS in a subset of the

population [3]. Various treatment options for BCS include treating the underlying disease, anticoagulation therapy, percutaneous angioplasty, TIPS, and liver transplantation [3]. Anticoagulation therapy is recommended for all patients with BCS, even in the absence of a prothrombotic disorder. Percutaneous angioplasty is recommended for patients with short-segment stenosis of either the HV or IVC. When HV involvement is predominant, anticoagulation along with angioplasty provides symptomatic relief in 20-30% of patients with BCS. TIPS using a covered stent graft is used for suitable patients who have failed anticoagulation/angioplasty. Liver transplantation is an option for patients in whom previous options have failed or are not possible [6,7].

Recanalisation of the HV has been increasingly used in the treatment of BCS in recent years with the aim of relieving hepatic congestion and resolving BCS symptoms [1,2,9,10]. Nowadays, TIPS is commonly used as the first-choice treatment for BCS. However, shunt dysfunction is a common problem, occurring in up to 13%-50% of patients when metal stents are used for TIPS [11]. In contrast, recanalisation of the HV is a physiological procedure, unlike TIPS, as it does not directly mix portal venous blood with systemic blood.

A recent study by Lv LL et al., in 2021 reported that recanalisation of the AHV can safely and effectively treat HV-type BCS, and may result in longer patency compared to recanalisation of the main HV (MHV) [12]. Similar findings were reported by Li DM et al., and Xia FF, with a sample sizes of 46 and 27 respectively [13,14]. The present case also showed similar results, with AHV recanalisation being performed without any complications and with good patency.

Patients with BCS may experience progressive liver dysfunction even if they are treated with a patent surgical shunt or TIPS shunt. Therefore, lifelong follow-up and monitoring with liver function tests are necessary for these patients. Some of them may eventually require liver transplantation [15].

CONCLUSION(S)

AHV stenting is a safe, physiological, and effective treatment option for patients with BCS. It provides an effective and satisfactory means of treating patients and is associated with good long-term outcomes.

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