Abdominal Radiology (2025) 50:1307–1319
https://doi.org/10.1007/s00261-024-04558-4

REVIEW

Interventions in Budd-Chiari syndrome: an updated review

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Received: 3 June 2024 / Revised: 25 August 2024 / Accepted: 29 August 2024 / Published online: 26 September 2024

Abstract

Budd Chiari syndrome is a potentially treatable disease, and imaging is the key to its diagnosis. Clinical presentations may vary, ranging from asymptomatic to fulminant disease. Subacute BCS is the most common type encountered in clinical practice, characterized by ascites, hepatosplenomegaly, dilated abdominal wall veins, and varicosities in the lower limb and scrotum. While hepatic vein thrombosis is the leading cause in the West, membranous and short segmental occlusion are predominant in the Asian populations. These geographical variations have an impact on the treatment algorithm in managing BCS. Anticoagulation alone often fails to prevent disease progression, demanding further interventional therapy. Interventional therapy carries a lower morbidity and mortality than surgery. Anatomical recanalization and portosystemic shunting form the basis of endovascular management. Membranous or short-segment occlusion are best treated by angioplasty, which restores the physiological venous outflow and possibly disease reversal. Suboptimal results with angioplasty require stenting. Transjugular intrahepatic shunt (TIPS) or direct IVC to portal vein shunt (DIPS) decompresses the portal pressure and reduces the sinusoidal congestion, which in turn diminishes hepatocellular damage and hepatic fibrosis. Despite its ability to modify the disease course, TIPS carries several procedure and shunt-related complications, mainly hepatic encephalopathy. Thus, anatomical recanalization precedes TIPS in the traditional step-up approach in managing BCS. However, this concept is challenged by some authors, necessitating future reseach. TIPS is a valid bridge therapy in BCS with acute live failure awaiting liver transplantation. Despite all, interventional therapies fail in a subset of BCS patients, leaving them with only option of liver transplantation.

Keywords Budd Chiari syndrome (BCS) · Anatomical recanalization · Hepatic vein angioplasty · IVC angioplasty · Stenting · Transjugular intrahepatic portosystemic shunt (TIPS) · Direct IVC to portal vein shunt (DIPS)

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Introduction

Budd-Chiari syndrome (BCS) refers to hepatic venous outflow obstruction from the level of small hepatic venules to the right atrium-inferior vena cava junction. Venous outflow obstruction leads to venous congestion, increasing hepatic sinusoidal pressure and causing hypoxic damage to hepatocytes. Unless the obstruction is relieved, gradual hepatocellular damage leads to progressive centrilobular fibrosis, nodular regenerative hyperplasia, and, ultimately, cirrhosis of the liver [1]. Apart from the hepatic damage, chronic BCS raises the risk of hepatocellular carcinoma [2].

The etiology and level of obstruction in BCS varies among the different regions of the world. Hepatic vein thrombosis is the most common type of obstruction in the Western world, with myeloproliferative disorders remaining the leading cause. In contrast, hepatic vein ostial stenosis, short segment thrombosis, and membranous inferior vena cava (IVC) obstruction prevail in the Asian-pacific region. These variations in the type of venous obstruction provide a unique opportunity for anatomical restoration of venous outflow among the Asian-pacific populations [3, 4].

BCS has a female preponderance. Clinical presentations also vary from asymptomatic disease to fulminant liver failure. Acute BCS is more common in the West, while most BCS in the East, including Asia, have subacute and chronic presentation [4, 5]. Acute BCS presents with pain abdomen, hepatomegaly, acute ascites, and jaundice. Acute onset occlusion of all three hepatic veins with no time for venous collateralization leads to a fulminant course, presenting with rapid onset abdominal pain, ascites, and features of acute live failure. Subacute and chronic BCS frequently present with hepatomegaly, splenomegaly, ascites, dilated abdominal wall veins, and varicosities in the lower limb and scrotum [4, 6]. Nearly 15% of BCS remains asymptomatic with atypical symptoms and are detected while evaluating for chronic liver disease. Clinically, chronic BCS may mimic cirrhosis and may present with esophageal varices and variceal bleeding [4, 5].

Imaging is critical to diagnose BCS. It also helps identify the type of obstruction, portal vein (PV) thrombosis, and other associated features, such as hepatosplenomegaly, ascites, hepatic lesions, and collateral pathways. Hepatic vein patency and flow dynamics are best evaluated using Doppler ultrasound. Cross-sectional imaging, i.e., contrast-enhanced CT and MRI, allows detailed evaluation of hepatic morphological changes, liver lesions, vascular abnormality, and mapping of vascular anatomy required before endovascular intervention or surgery. IVC obstruction is better evaluated on CECT/MRI than Doppler ultrasound. Although catheter venography is considered the gold standard, it is seldom required for diagnosis due to improvements in ultrasound and cross-sectional imaging [7]. Imaging features of BCS are summarized in Table 1 [7–9].

Management of BCS

Anticoagulation therapy is usually the first therapy initiated upon diagnosis of BCS. However, anticoagulation alone has a poor outcome, and often, the disease progresses, leading to cirrhosis and portal hypertension [10]. Endovascular interventions are the cornerstone therapy in managing BCS due to their minimally invasive nature, with lower morbidity and mortality than surgery. Although no unified treatment consensus exists, a step-up approach is commonly followed for BCS [10, 11]. The presence of liver failure necessitates liver transplantation. Underlying causes, such as myeloproliferative disorder and other hypercoagulable conditions, should also be addressed concurrently [4]. Currently, treatment of portal hypertension-related complications in BCS follows the same guidelines as that of cirrhosis due to any cause [12].

Step-up approach in BCS

The current European and American guidelines follow the step-up approach in managing BCS [13, 14]. The step-up strategy includes a gradual escalation in the invasiveness of therapies according to non-response, starting with anticoagulation, then HV/IVC recanalization, then transjugular intrahepatic portosystemic shunt (TIPS)/direct IVC to portal vein shunt (DIPS), and finally, liver transplantation [15, 16]. However, the definition of response to therapy is still undefined, presently based on arbitrary clinical criteria. Furthermore, the step-up approach ignores the concepts of venous congestion-driven chronic microvascular ischemia perpetuating hepatocellular necrosis and hepatic fibrosis [10, 11, 16, 17].

Firstly, restoration of hepatic sinusoidal flow by hepatic vein recanalization is uncommon with anticoagulation only and probably does not justify a wait-and-watch strategy [10, 15]. Second, early anatomical recanalization restores the regular hepatic flow, preventing the disease progression. Thus, early anatomical recanalization can not be ignored in feasible cases (e.g., HV or IVC web), considering its potential benefits of disease reversal [18, 19]. Third, obliterative changes in the intrahepatic PV branches add to the disease progression in BCS, which TIPS may prevent [15, 20]. Fourth, TIPS is probably superior to spontaneous venous collaterals with respect to timing and capacity in patients with acute or subacute BCS. It relieves venous congestion,

Table 1 Summary of imaging features in Budd-Chiari syndrome

Acute BCS	USG	CT	MRI
Vascular	• Non-visualization of veins • Thrombus filling the lumen (in acute form) • HV/IVC narrowing or web • Venous stenosis with proximal dilation • Absent/flat flow, reversed flow in HV/IVC on Doppler • Reduced or hepatofugal portal flow • Caudate vein ≥ 3 mm	• Non-opacification or filling defect within HVs • IVC compressed by enlarged caudate lobe • Venous stenosis with proximal dilation	• Loss of normal T2 flow void in HVs • Other features same as CT and USG
Parenchymal	• Hepatomegaly • Altered hepatic echogenicity • Caudate hypertrophy • Ascites	• Decreased and patchy peripheral hepatic parenchymal enhancement with stronger central part of liver parenchymal • Other features same as seen in USG	• Heterogeneously decreased signal on T1W and increased signal on T2W within the peripheral liver • Increased enhancement in the caudate with decreased enhancement in the peripheral liver • Other features same as seen in USG
Chronic BCS			
Vascular	• Occluded or stenosis of hepatic veins or IVC • Fibrous cord replacing the vein • Comma-shaped veno-venous collaterals • Caudate vein ≥ 3 mm	• Enlarged hepatic artery • Veno-venous collaterals and portosystemic collaterals better depicted than USG	• Same as that of CT
Parenchymal	• Features of cirrhosis • Isoechoic NRH nodules may be seen	• Features of cirrhosis • Arterially enhancing NRH nodules with no washout in portovenous phase	• Changes of cirrhosis • NRH nodules-T1 hyper/T2 iso to hypointense with arterial hyperenhancement, no washout
Subacute BCS			
Vascular	Same as chronic BCS		
Parenchymal	• Mixed features of both acute and chronic BCS • NRH nodules are less common than chronic BCS • Volume redistribution changes seen • Liver parenchymal enhancement pattern is more like acute BCS		

BCS Budd Chiari syndrome, *HV* Hepatic vein, *IVC* Inferior vena cava, *NRH* Nodular regenerative hyperplasia

may stabilize hepatocyte function, and prevent disease progression [17]. Furthermore, early TIPS may improve outcomes in BCS patients with a high risk of liver failure and variceal bleeding [15].

Patients with chronic BCS often have adequate collaterals for decongestion. However, normalization of sinusoidal pressure may not occur, leading to progressive hepatocellular injury. The signs and symptoms of portal hypertension usually guide treatment in such patients. Another important consideration in such patients is lifelong anticoagulation therapy, which has an increased risk of bleeding complications, especially in those with large varices [13]. Thus, TIPS placed earlier than usual may be a reasonable option to prevent bleeding in patients with large varices requiring anticoagulation.

Considering these aforementioned facts, some authors argue against the traditional step-by-step treatment algorithm. Manusco proposed a new treatment algorithm suggesting early interventional therapy in symptomatic patients [17]. Early intervention may prevent disease progression, improve liver function, prevent portal hypertension, and possibly improve one's chance of survival. TIPS, however, may be associated with several procedural complications, including an increased risk of hepatic encephalopathy. Furthermore, liver function may worsen despite TIPS requiring liver transplantation [10, 15, 17]. These factors must be considered before deciding the type of intervention in BCS. Nevertheless, the choice between early and late intervention in BCS is still debatable, necessitating prospective comparative studies.

Interventions in BCS

The two interventional strategies in BCS include anatomical recanalization and portal flow diversion. Anatomical recanalization consists of thrombolysis and HV/IVC angioplasty with or without stenting. Portal flow diversion, also called derivative treatment, is usually achieved by creating a TIPS or DIPS. Surgical shunting is rarely considered nowadays.

Ultimately, BCS not responding to interventional therapies requires liver transplantation [21]

I. Anatomical recanalization

a. HV recanalization

In the vast majority of cases, one patent native HV or accessory HV suffices for adequate hepatic decongestion. A comprehensive imaging evaluation, including USG Doppler and CECT, is required to identify the best HV amenable to recanalization. The best HV should have a straight course, echo-free lumen, caliber of at least 7–8 mm, and draining sizable liver parenchyma with multiple veno-venous collaterals joining it [22].

The transjugular approach is most commonly preferred. A long sheath into IVC may be required to provide adequate catheter support during the negotiation of the stricture. Membranous occlusion and short segment stenosis are easy to negotiate using a combination of angled hydrophilic guidewire and 4F/5F diagnostic catheter. A curved metallic cannula and 5 F catheter-and-stylet assembly of Rosch-Uchida transjugular liver access set (RUPS-100, Cook Medical) may be required to cross the occluded segment for fibrotic and segmental obstruction. In a few cases, crossing HV stricture may be possible antegradely after access into the target HV via venous collateral communicating with IVC. Once the occlusion is crossed, serial angioplasty is performed using a high-pressure, non-complaint balloon (usually 10 to 14 mm). A post-angioplasty venogram is taken, and the pressure gradient should be measured across the occlusion with a target decrease in pressure gradient to < 5 mm Hg. A successful angioplasty shows a good antegrade flow with the disappearance of collaterals (Fig. 1).

A failed transjugular approach necessitates a percutaneous transhepatic route. Target hepatic vein access is obtained using a micropuncture set, followed by stricture negotiation using an angled tip hydrophilic guidewire/5F catheter combination. In case of tight stricture, the stiff back end of the hydrophilic guidewire or a long 21G/22G Chiba needle (sharp recanalization) might be used to cross the stricture. An inflated balloon can be placed as a target at the HV-IVC junction to minimize the risk of non-target punctures, such as capsular transgression or pericardial puncture [23]. Cone beam CT (CBCT), if available, aids in sharp recanalization technique. Once the stricture is crossed, the guidewire is snared from the jugular or femoral approach, and the rest of the procedure is completed through the transjugular or transfemoral route. Some operators prefer angioplasty and stenting through the percutaneous transhepatic route [24, 25]. However, it may require a larger sheath placement, raising the bleeding risk. In the end, the percutaneous transhepatic tract is obliterated using coils or glue to avoid life-threatening hemorrhage (Fig. 2).

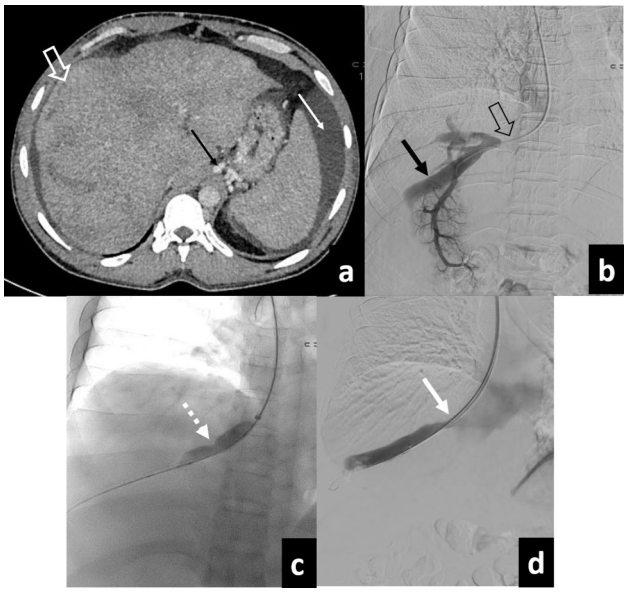

Fig. 1 RHV angioplasty for membranous occlusion in a patient with subacute BCS presenting with deranged liver function and ascites. **a** Axial CECT shows an enlarged and congested liver (open white arrow **a**). In addition, ascites (thin white arrow **a**) and lower esophageal venous collaterals (thin black arrow **a**) are seen, suggesting portal hypertension. **b** RHV venogram through a transjugular route reveals membranous ostial occlusion of RHV (open black arrow **b**) with the dilated proximal vein (thick black arrow **b**). **c** Obstruction was dilated using a 12 mm balloon (dash white arrow **c**). **d** Post–angioplasty venogram showing free flow (thick white arrow **d**) across the stenotic segment into IVC *RHV* Right hepatic vein, *CECT* Contrast-enhanced CT, *IVC* Inferior vena cava

HV stenting While angioplasty alone suffices for short segment and membranous HV occlusion, the persistence of residual stenosis > 30% and/or pressure gradient > 5 mm Hg necessitates stenting. Self-expanding metallic stent (SEMS) is most commonly used for HV stenting, with stent diameter ranging from 10 to 16 mm Hg and length varying from 40 to 60 mm Hg. The stent diameter should be about 2 mm more than the vein and extended by 1 cm proximal and distal to the stenotic segment [4]. Stent dysfunction during follow-up requires thrombolysis, balloon angioplasty, or a combination of both. Placement of a new stent may be required in some cases.

b. IVC recanalization

Short segment/membranous occlusion of suprahepatic IVC is one of the leading causes of BCS in Asian populations and is best treated with IVC angioplasty. In contrast to HV angioplasty, IVC angioplasty is commonly performed through the transfemoral route (Fig. 3). Tight stricture necessitates

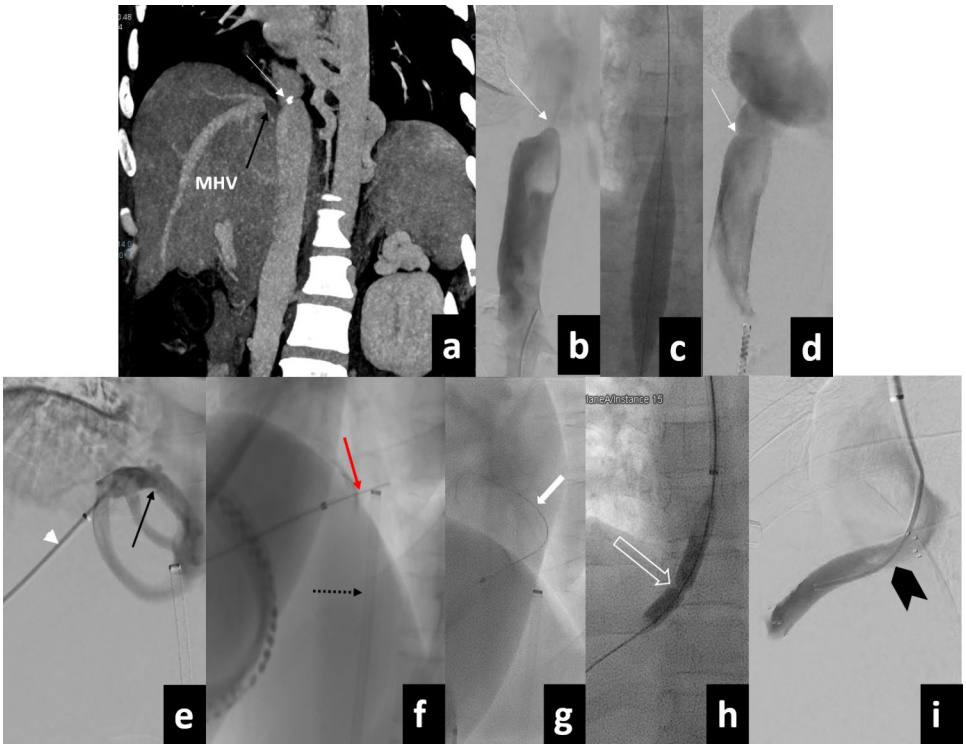

Fig. 2 Combined IVC and HV recanalization (sharp recanalization of HV using Chiba needle). **a** Oblique coronal MIP image showing a thin & calcified IVC web and short-segment fibrotic stricture of MHV. **b–d** IVC venogram confirmed the IVC occlusion (white arrow **b**) which was then recanalized with an 18 mm balloon (white arrow **d**). **e–i** Upon failed transjugular MHV catheterization, percutaneous MHV access was obtained (arrowhead **e**), followed by an MHV venogram that showed ostial occlusion (black arrow **e**) with multiple collaterals. The fibrotic occlusion was crossed using a 21G 20 cm Chiba needle (red arrow **f**) under fluoroscopic guidance. Further, a 10F sheath (dash black arrow **f**) placed in IVC also guided IVC puncture. Once the occlusion was crossed and IVC punctured, a 0.018' guidewire (V-18, Boston Scientific) (thick white arrow g) was passed into the heart, snared from the jugular route, and then through and through access was obtained. Through the transjugular route, the stricture was serially dilated to 10 mm using balloons (open white arrow **h**) and then a 12 mm self-expanding metallic stent (black arrowhead **i**) was placed to complete the procedure *IVC* Inferior vena cava, *MIP* Maximum intensity projection, *MHV* Middle hepatic vein, *G* Gauze

additional maneuvers, such as advancing a long sheath till the occlusion and attempting stricture crossing using a guiding catheter and straight-tip hydrophilic guidewire or placing long sheaths till occlusion from both femoral and jugular route, followed by probing (Fig. 4). Notably, probing a tight stricture using a long Chiba needle/Colapinto needle mandates utmost precaution to avoid IVC or cardiac perforation, resulting in catastrophic hemorrhage. Hardwires on either side of the stricture must be aligned in a straight line, which is confirmed by obtaining orthogonal views in anteroposterior and lateral projection [22, 26]. CBCT also helps in guiding the right trajectory during probing.

IVC stenting Some operators prefer avoiding early stenting as it makes further interventions, such as TIPS and liver transplantation, difficult. Stenting is usually reserved for recurrent IVC occlusion or significant residual stenosis despite angioplasty. SEMS of 25–30 mm diameter is recommended for IVC stenting [4]. Few operators choose primary stenting, particularly in patients with long-segment IVC stenosis.

Follow-up

A standard approach includes follow-up at 1, 3, and 6 months, and subsequently every 6 months or annually. The follow-up visits must include clinical, laboratory, and imaging parameters [4, 27, 28]. USG Doppler is the widely used imaging modality. Inconclusive USG findings or findings suspicious of re-stenosis/re-occlusion necessitate further confirmation with cross-sectional imaging. Rarely, invasive venography may be needed.

During the post-intervention period, INR is maintained between 2 and 3 since most BCS patients often have underlying hypercoagulability. Initially, low molecular weight heparin (LMWH) is administered, followed by bridging with warfarin and long-term oral warfarin therapy thereafter. INR should be strictly monitored until stable INR

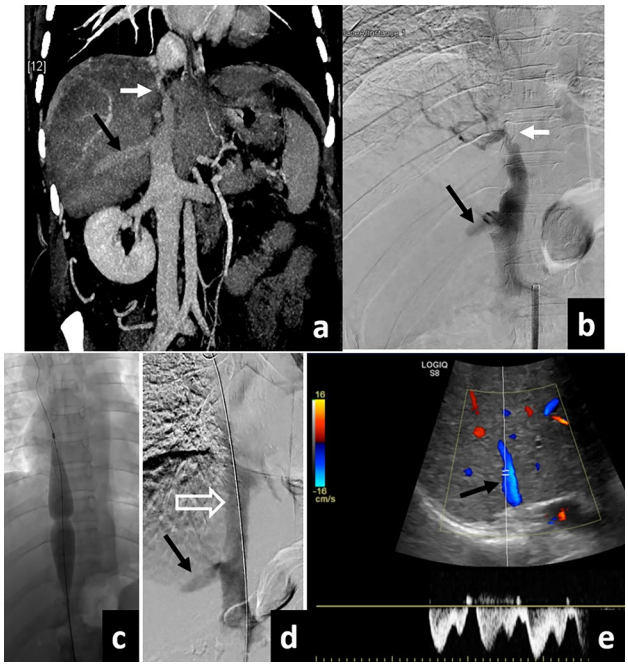

Fig. 3 Segmental IVC recanalization through transfemoral route in a patient with abdominal pain and lower limb swelling. **a** Coronal CECT showing short-segment intrahepatic IVC obstruction due to IVC web and associated chronic eccentric thrombus (white arrow **a**). The liver was draining mainly through right AHV (black arrow). **b** IVC venogram confirmed the CT findings (white arrow **b**). **c** Stricture was crossed and balloon angioplasty was done using a 20 mm balloon. **d** Immediate post-angioplasty venogram showing recanalized IVC (open white arrow **d**) with the normal drainage of AHV (black arrow). **e** 3-month follow-up USG showing the normal phasic flow in AHV (black arrow), suggesting patent intra and suprahepatic IVC. Symptoms gradually improved over a period of 1 to 2 months. *AHV* Accessory hepatic vein, *IVC* Inferior vena cava

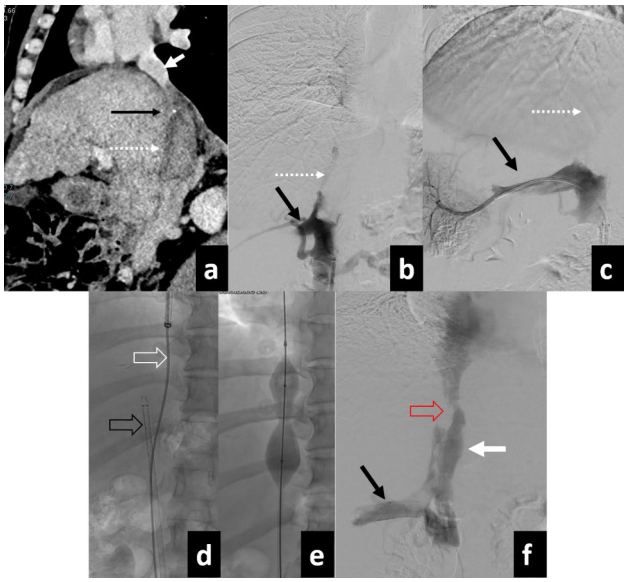

Fig. 4 Transjugular IVC recanalization after a failed transfemoral approach. **a** Sagittal CECT showing a calcified (thin black arrow a) web within the intrahepatic IVC with proximal thrombus (dash white arrow **a**) and a patent suprahepatic IVC (short white arrow **a**); **b**, **c** Transfemoral venogram revealing intrahepatic IVC occlusion (dash white arrow **b**) with no forward flow to heart. A dominant right inferior accessory hepatic vein is also seen (black arrow **b**, **c**). **d** Following a failed transfemoral approach, stricture was negotiated after wedging an MPA (open white arrow **d**) catheter in the suprahepatic IVC through a transjugular route. **e** Balloon angioplasty was performed after crossing the obstruction. **f** Post-angioplasty venogram depicting recanalized IVC (thick white arrow **f**) with drainage of AHV (black arrow **f**). Although IVC was partially recanalized (open red arrow **f**), leg swelling improved after angioplasty, and the patient was kept on long-term anticoagulation *CECT* Contrast-enhanced CT, *IVC* Inferior vena cana, *MPA* Multipurpose angiographic catheter, *AHV* Accessory hepatic vein

is achieved in the range of 2–3 [28, 29]. Direct-acting oral anticoagulants (DOACs) have also been tried in BCS since they do not require monitoring and dose adjustment. Preliminary data are encouraging; however, evidence is limited to formulate recommendations [4, 30]. Moreover, routine usage of long-term anticoagulation following angioplasty in patients with only membranous occlusion requires further validation.

Liver stiffness (LSM) value is a promising noninvasive tool for monitoring response to endovascular intervention. Decongestion of the liver reduces liver stiffness; thus, a decrease in LSM value following endovascular therapy indicates a response to therapy. Notably, LSM value can be unreliable in the presence of gross ascites and obesity [31–33].

Outcomes of HV/IVC recanalization

The pooled technical success rate of HV/IVC recanalization was found to be 97.9% (95%CI: 95.9–99.9%) in the largest recent meta-analysis [34]. The clinical success rate varies considerably among studies, ranging from 80 to 100% [18, 19, 25, 27, 28, 34]. The long-term treatment outcomes of anatomical recanalization are found to be superior in membranous than segmental obstruction. About 10% of HV/IVC recanalization cases require re-intervention within five years. The 1-year and 5-year survival rates are > 90% and > 80% respectively [34].

Angioplasty vs. stenting

Most studies on BCS support initial balloon angioplasty for anatomical recanalization, with stenting reserved for significant residual and recurrent stenosis. The only available RCT comparing angioplasty alone vs. primary stenting

in BCS demonstrated the superiority of primary stenting over angioplasty alone. During a median follow-up time of 27 months, the primary stenting group had a significantly higher proportion of patients free of re-stenosis than the angioplasty alone group [42/43 (98%) vs. 27/45 (60%)]. The 3-year re-stenosis-free survival was about 1.5 times higher in the primary stenting (96%, 95%CI: 88.6–100.0) compared to the angioplasty alone group (60.4%, 95%CI: 46.4–78.7) (log-rank $p < 0.001$). Notably, there were no reports of stent fracture or migration [35]. Results from another large retrospective analysis ($n = 177$; angioplasty alone = 51, angioplasty + stenting = 117) on anatomical recanalization in BCS by Han et al. also showed that the re-occlusion rate was nearly four times higher (31% vs. 7.7%, $p < 0.001$) in the angioplasty alone group compared to the angioplasty plus stenting group. Multivariate analysis in that study indicated that angioplasty alone was an independent risk factor for re-occlusion [36].

According to a study evaluating post-intervention IVC patency rate in BCS, the long-term patency following balloon angioplasty alone was found to be lower in segmental obstruction of IVC (SOVC) than membranous obstruction of IVC (MOVC) ($p = 0.001$); however, no significant difference was found between the two groups when they underwent stent placement ($p = 0.687$) [37]. 38 BCS patients with MOVC were successfully treated by balloon angioplasty alone in a study by Yang et al. Only one patient had recurrence during eight years of follow-up [38]. Similarly, in a different series of patients with membranous hepatic vein obstruction with or without IVC obstruction, Ding et al. ($n = 93$) demonstrated good long-term results with angioplasty alone. During the follow-up, re-occlusion occurred in only 8.9% of patients [28].

Therefore, the requirement of stenting primarily depends upon the type of venous outflow obstruction. Short-segment stenosis responds better to balloon angioplasty than long-segment stenosis. Furthermore, segmental occlusion carries a greater risk of re-occlusion with angioplasty alone compared to membranous occlusion. Primary stenting may be preferred in such patients with better success rates.

c. Accessory hepatic vein recanalization in BCS

Hepatic venous outflow obstruction prompts the development of venous collaterals and accessory hepatic vein (AHV) dilation. Of note, a single AHV of caliber > 5 mm in diameter often suffices for liver decongestion due to the development of collaterals between main HVs and AHV [39, 40]. When AHV compensatorily dilates, its ostium remains of the same caliber due to constraint by the IVC wall, leading to relative focal ostial stenosis. In such a case, AHV recanalization effectively achieves positive long-term outcomes in patients with BCS. A recent meta-analysis, including retrospective studies, found that AHV recanalization had a longer primary patency and a lower re-stenosis rate than native hepatic vein recanalization. The primary clinical success rate (96%), 1-year (97%), and 5-year survival (96%) rates were comparable to that of HV recanalization [41]. Balloon angioplasty often suffices for AHV recanalization owing to its focal membranous nature in most instances. Failed angioplasty necessitates stent placement. It is worth noting that compensatory AHVs are not present in all HV-type BCS patients, limiting the utility of this recanalization strategy only to a subset of BCS patients [42]. In some individuals, other dominant collateral vein recanalization may be contemplated if found suitable on imaging [40].

d. HV/IVC thrombolysis

Thrombolysis is typically performed in patients with acute BCS. Catheter-directed thrombolysis is carried out via a jugular or femoral route with t-PA (5 mg bolus followed by 0.5 mg/hour for six hours) or urokinase (3000 units/kg bolus followed by 50,000 units/hr. for 6–12 h). Mechanical thrombolysis may also be required in some cases [43, 44]. Notably, stenting is avoided in acute BCS.

II. Transjugular intrahepatic portosystemic shunt (BCS-TIPS)

In patients with complete occlusion of all hepatic veins, hepatic blood flow depends on the adequacy of the intrahepatic and extrahepatic venovenous collaterals. However, slow or insufficient collateralization could worsen the disease and necessitate prompt interventional therapy [4, 45]. TIPS diverts the portal flow from the liver and immediately relieves portal hypertension. In the long term, TIPS may also reduce or prevent the development of cirrhosis and regenerative hyperplasia by alleviating microvascular ischemia resulting from sinusoidal congestion [17]. Presently, TIPS is preferred over surgical shunt since it is less invasive and is associated with less morbidity and mortality [46].

The indications of TIPS in BCS include (1) the absence of recanalizable hepatic veins, (2) no response to anticoagulation and angioplasty, and (3) worsening disease despite successful anatomical recanalization [47]. Patients receiving BCS-TIPS often have refractory ascites (up to 100%) and variceal hemorrhage (up to 30.9%) [48]. The technical success of BCS-TIPS is reported to be > 95% [49].

Symptomatic acute BCS should be treated as early as possible. TIPS remains a rescue therapy in acute BCS when thrombolysis or anticoagulation fails. The presence of acute liver failure necessitates liver transplantation, and TIPS can be utilized as a bridge therapy in this situation [47, 50, 51]. Notably, TIPS has a dismal prognosis in acute liver failure.

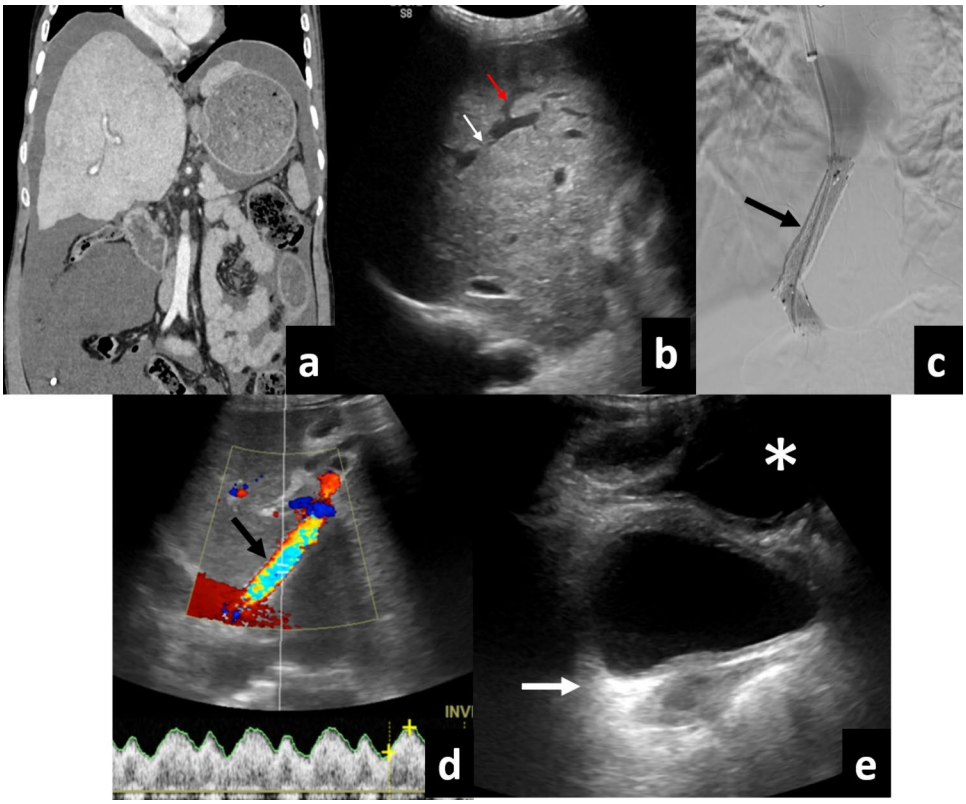

Fig. 5 BCS-DIPS in a patient with gross ascites and non-recanalizable hepatic veins. **a** Coronal CECT showing enlarged liver with mild heterogeneous parenchymal enhancement and gross ascites; **b** USG showing intrahepatic venous collaterals (red arrows **b**) and thrombus within the hepatic vein (white arrow **b**); **c** Direct IVC to PV shunt (black arrow **c**) was created using a combination of 10 mm stent graft and 10 mm bare metallic stent; **d**, **e** Follow up USG at 1 month depicting patent DIPS with phasic flow and Doppler with the resolution of ascites (white arrow e showing no fluid in pelvis). *Denotes a resolving abdominal wall hematoma that occurred during a previous paracentesis *DIPS* Direct IVC to portal vein shunt, *CECT* Contrast-enhanced CT, *USG* Ultrasonography

The presence of severe jaundice and hepatic encephalopathy do not preclude TIPS in acute BCS patients. Instead, TIPS should be performed as soon as possible [50, 51].

TIPS technique

Pre-procedural imaging, especially CECT or CE MRI, helps delineate the relevant anatomy and aids in procedural planning. TIPS increases the cardiac preload and may trigger congestive cardiac failure in patients with pre-existing cardiac dysfunction and pulmonary arterial hypertension. Hence, pre-TIPS workup must include a detailed cardiac evaluation, including ECG, echocardiography, and NT pro-BNP.

The absence of patent hepatic veins, volume redistribution, and ventral hilar displacement due to an enlarged caudate lobe pose greater technical difficulty for TIPS in BCS than in cirrhosis. In failed hepatic vein cannulation, the shunt is created directly between intrahepatic IVC and PV, also called direct IVC to PV shunt (DIPS).

TIPS is performed in the fluoroscopic suite under conscious sedation with adequate analgesia. General anesthesia may be preferred whenever available. First, an IVC venogram is taken to evaluate IVC patency and exclude the IVC web. It also helps identify the HV stump from where the parenchymal puncture is started during PV access. HV cannulation is then attempted through the transjugular route. After successful HV cannulation, PV is punctured using a stiff curved needle (RUPS-100, Cook Medical). In case of failed HV cannulation, PV is targeted directly from the intrahepatic IVC. Transabdominal USG guidance is an easy and safe technique for guiding PV puncture. Alternatively, intravascular USG guidance may be used. Once PV is accessed, venogram and hemodynamic monitoring are obtained. The parenchymal tract is then dilated, and the shunt is created using either a hybrid stent having covered and bare parts or a combination of covered and uncovered stent. An expanded polytetrafluoroethylene (e-PTFE) lined stent graft is recommended for TIPS creation. The aim is to line the parenchymal tract with a stent graft due to longer shunt patency. A 10 mm diameter shunt is considered optimal for BCS (Fig. 5) [49, 52].

Anatomically challenging cases may require procedural modification, such as the Gun-sight technique in which the PV and IVC are punctured using a 21G needle under the USG/CT guidance to obtain a through and through access, followed by snaring of the guidewire through the jugular access. Once the IVC to PV access is obtained, the procedure is the same as a regular TIPS/DIPS procedure, completed through the transjugular route [53].

About 15 to 20% of patients with BCS have concomitant PV thrombosis [54]. Although the traditional transjugular route is most commonly preferred for BCS-TIPS in such a situation, procedural modification, such as trans

splenic or transhepatic assistance, may be required during TIPS/DIPS creation. A gooseneck snare loop or a contrast-filled balloon is placed as a target at the intended site of the PV puncture via percutaneous transhepatic or transsplenic access. Then, the snare or balloon is targeted from the IVC or HV via the transjugular route. Once PV access is obtained, the remaining procedure is the same as that for conventional TIPS. PV thrombus may require pharmacological, mechanical thrombolysis, or a combination of both [55]. The step-by-step technical aspect of regular TIPS/DIPS, PVR-TIPS, and gun sight techniques is beyond the scope of this article and is described elsewhere in detail.

Post-TIPS follow-up

The post-BCS-TIPS follow-up protocol is the same as for anatomical recanalization (discussed before), and it includes ultrasound, biochemical, and clinical evaluations.

BCS-TIPS outcome

TIPS decreases the portal pressure and hepatic congestion by reversing the flow in intrahepatic portal branches. Improvement in clinical and biochemical parameters usually takes 2 to 4 weeks, especially in acute BCS patients [56].

The clinical success rate following BCS-TIPS varies significantly among studies, ranging from 86 to 94.6% [49]. Variable disease severity and clinical symptoms among studies may explain this wide range of reported clinical success rates. TIPS may fail to improve liver function, requiring liver transplantation in up to 5% of patients with BCS-TIPS [49].

BCS-TIPS prognostic index (BCS-TIPS PI) is the most widely validated prognostic score to predict OLT-free survival after TIPS [4]. This score is calculated using the formula: Age (years) x 0.08 + bilirubin (mg/dl) x 0.16 + INR x 0.63. Patients with BCS-TIPS PI < 7 have a 1-year OLT-free survival rate of 95%, while a score of > 7 is associated with a poor prognosis, with a 12% 1-year OLT-free survival rate. Thus, patients with a BCS-TIPS PI score of > 7 should receive liver transplantation as early as possible [4, 10].

Survival

Various studies have shown one and five-year survival rates of 80–100% and 70–80%, respectively, in BCS-TIPS patients [49, 51, 56–59]. Available data suggests that TIPS might improve survival [10, 57, 58]. However, there is dearth of comparison data on survival rates in BCS patients receiving TIPS versus non-TIPS treatment. In fact, a comparative study is not feasible unless the traditional step-up approach is violated.

Post-TIPS complications

Myriads of complications can occur during and after TIPS creation, with procedure-related complications ranging from 0 to 56% [49, 60]. However, major adverse events following BCS-TIPS are seen in only up to 10% of patients, and immediate procedure-related mortality is noted only in 0.5% of patients [49]. These complications include capsular transgression, IVC and extrahepatic PV injury, contrast-induced nephropathy, and stent migration. Notably, TIPS is associated with more procedure-related complications than angioplasty in BCS and may be a reason to be cautious about early TIPS as a first-line intervention.

Post-TIPS HE may occur due to portosystemic shunting. HE following TIPS in BCS is less common than in patients with liver cirrhosis since most of the patients with BCS have a better hepatic functional reserve. According to a recent meta-analysis, the incidence of HE following TIPS is seen only in 11.6% of patients [49]. Detailed discussion about TIPS complications and their management are beyond the scope of this article.

BCS-TIPS dysfunction

The incidence of shunt dysfunction is reported to be higher in BCS-TIPS than TIPS in cirrhosis, mainly attributable to the associated prothrombotic states. A recent meta-analysis reported a cumulative TIPS dysfunction rate of 40.1%, and BCS-TIPS with bare metallic stent was associated with a three times higher rate of shunt dysfunction than with covered stent (68.8% vs. 22.8%, risk ratio = 2.67; 95% CI: 1.77–4.04) [49]. Notably, the rate of TIPS dysfunction varies considerably between studies, with a decreasing trend from bare stent to the stent-graft era. The shunt patency rate has improved significantly with the introduction of the e-PTFE stent graft [61].

Recurrence of portal hypertension-related complications raises suspicion of shunt function. Intra-stent flow velocity of < 60 cm/sec or > 180 cm/sec on Doppler indicates shunt dysfunction. A decrease in extrahepatic PV velocity < 30 cm/sec is an indirect feature of shunt dysfunction [56, 62]. Inconclusive Doppler findings may necessitate further evaluation with a catheter TIPS venogram and portosystemic pressure gradient (PSPG) measurement to confirm shunt dysfunction. Shunt dysfunction requires balloon dilatation ± thrombolysis, and restenting may be done if deemed necessary. A parallel TIPS may be considered if shunt revision fails [63].

Timing of BCS-TIPS

The ideal timing of TIPS in BCS is largely unknown. While the traditional step-up strategy is the recommended approach

in BCS, some authors argue in favor of early TIPS, considering its ability to reduce sinusoidal congestion by portal flow diversion, thereby preventing venous outflow-induced chronic microvascular ischemia and hepatic fibrosis [10, 11, 64, 65]. Nevertheless, TIPS is associated with several procedures and shunt-related complications, particularly hepatic encephalopathy. Further, liver function may worsen despite TIPS in a subgroup of BCS patients requiring liver transplantation [10, 56]. Thus, prospective trials with larger sample sizes are warranted to define the optimal timing of BCS-TIPS.

Anatomical recanalization vs. TIPS in BCS

Recanalization of HV and/or IVC restores the anatomical venous outflow while TIPS diverts the portal flow, reducing the hepatic sinusoidal pressure and further hepatocellular damage. HV/IVC recanalization is less invasive than TIPS and has fewer procedural complications. A study evaluating the long-term outcome following HV recanalization in patients with BCS demonstrated that HV recanalization resulted in similar patency and survival rates compared to TIPS but with significantly lower procedural complications (9.5% vs. 27.1%) and hepatic encephalopathy (0 vs. 18%). 72% of patients responded to anatomical recanalization, with only 25% of cases requiring further TIPS or surgery. It is worth noting that patients who received TIPS had a more severe baseline disease, as evident from lower albumin, high MELD score, CTP, and revised new Clichy PI [18]. A multicentric prospective European study reaffirmed similar findings. Interestingly, 63% of patients in this study required further interventions (TIPS, surgical shunt, or OLT) after anatomical recanalization. On the contrary, TIPS showed a good outcome, regardless of its timing, i.e., early vs. late during the disease course [10]. Contradictory to the findings of the study mentioned above, a Chinese study demonstrated excellent survival in BCS patients following anatomical recanalization [19]. These differences in outcomes of anatomical recanalization between Western and Asian populations could be due to differences in the occlusive mechanism of hepatic venous outflow. Membranous or short-segment occlusion is more frequent in the Asian population, where anatomical recanalization remains a potentially valuable and durable intervention [4]. Recently, a study by Mukund et al. revealed no significant difference in survival between anatomical recanalization and TIPS in BCS. However, unlike TIPS, anatomical recanalization significantly improved serum bilirubin, albumin, AST, and ALT levels after three months and two years [66].

It is evident that the efficacy of anatomical recanalization steadily declines with the progression of the disease and may require TIPS insertion. Nevertheless, the criteria for identifying patients who will not respond to upfront anatomical recanalization remains unidentified, demanding further research. However, whenever feasible, anatomical recanalization should be attempted first. TIPS should be reserved for cases when anatomical recanalization is not possible or does not alleviate symptoms.

III. Liver transplantation in BCS

Liver transplantation (LT) is indicated for progressive liver failure despite medical and interventional therapies. It is also considered in acute BCS with fulminant liver failure. Outflow reconstruction remains a crucial technical component in LT for BCS [4]. An improving trend in post-LT outcomes has been noticed worldwide, probably due to improved surgical techniques and post-transplant management. The 5-year post-LT survival rate in patients with BCS is around 75% [67–69]. Older donor or recipient age and a MELD score of >30 are associated with poor outcomes. Furthermore, long-term anticoagulation remains an essential adjunct medical therapy in BCS following LT [67, 68].

Conclusions

Imaging is essential for diagnosing BCS and guiding intervention. Short segment or membranous HV/IVC occlusion responds well to anatomical recanalization. Failed or infeasible anatomical recanalization prompts TIPS placement. TIPS is also a valid rescue option in BCS with acute liver failure awaiting liver transplantation. The step-up approach in BCS is yet controversial, as is the timing of BCS-TIPS, necessitating future research.

Undoubtedly, endovascular intervention has revolutionized the treatment of BCS with favorable long-term outcomes and has nearly replaced surgical intervention. Despite timely intervention, liver function may progressively deteriorate. Liver transplantation is the last resort when all the interventional measures have failed.

Acknowledgements I would like to thank all the residents and supporting staff of department of Radiodiagnosis, AIIMS-Bhubaneswar.

Author contributions R.K.P and T.P.T conceptualized and wrote the main manuscript. K.C, T.P.T, S.G prepared Figs. 1 and 2, 3 S.B, T.D, S.G prepared Figs. 4 and 5. M.K.P, H.K.N and B.P reviewed and edited the manuscript.

Data availability No datasets were generated or analysed during the current study.

Declarations

Competing interests The authors declare no competing interests.

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