Advances and Challenges in Budd-Chiari Syndrome Management: A Review of the Literature

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ABSTRACT

Budd-Chiari syndrome (BCS) is a medical condition caused by a partial or complete obstruction of hepatic venous outflow. Epidemiologically, BCS was considered rare, and its infrequent diagnosis was attributed to limited diagnostic tools and low clinical awareness. Therefore, this review aimed to enhance clinicians' awareness, from accurate diagnosis to appropriate management of patients with BCS. The symptoms experienced by patients ranged from asymptomatic cases to fulminant forms of the disease. Imaging methods, such as Doppler ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI), were often used to establish the diagnosis. The primary treatment for BCS included anticoagulants, typically warfarin or low-molecular-weight heparin (LMWH). Depending on the patient's clinical status, more invasive therapeutic interventions could also be necessary.

Keywords: *Budd-Chiari syndrome (BCS), Doppler ultrasonography (US), Hepatic Venous Outflow Obstruction*

ABSTRAK

Budd-Chiari syndrome (BCS) adalah kondisi penyakit yang disebabkan oleh obstruksi parsial atau total pada hepatic venous outflow. Berdasarkan data epidemiologi BCS jarang terjadi, yang mungkin bisa terjadi karena instrumen diagnosis yang inadekuat dan kewaspadaan dokter yang rendah sehingga menjadi jarang terdiagnosis. Gejala klinis yang dialami pasien bervariasi, mulai dari asimptomatik sampai fulminant. Diagnosis dapat menggunakan batuan pemeriksaan radiologi seperti Doppler ultrasound (US), computed tomography (CT), dan Magnetic Resonance Imaging (MRI). Pengobatan pilihan utama pada BCS adalah penggunaan antikoagulan seperti warfarin dan low molecular weight heparin (LMWH). Sesuai dengan kondisi klinis pasien dapat juga disertai pengobatan dengan tindakan yang lebih invasif. Review article ini diharapkan membantu klinisi untuk meningkatkan kewaspadaan dalam diagnosis hingga tatalaksana pasie dengan BCS.

Kata Kunci: *Budd-Chiari syndrome (BCS), Doppler ultrasonography (US), Hepatic Venous Outflow Obstruction*

INTRODUCTION

Budd-Chiari syndrome (BCS) is a disorder caused by partial or complete obstruction of hepatic venous outflow, which may occur at the level of the small hepatic venules and the inferior vena cava.[1,2] The obstruction leads to venous stasis and congestion, causing increased sinusoidal pressure and hypoxic damage that affect both hepatocytes and sinusoidal endothelial cells. These pathological changes can lead to hepatocyte necrosis, centrilobular fibrosis, nodular regenerative hyperplasia, and may eventually progress to liver cirrhosis. The clinical presentation of BCS varies significantly, ranging from asymptomatic cases to fulminant hepatic failure.[2]

BCS remains underdiagnosed in many parts of Asia, primarily due to limited clinical awareness and lack of adequate diagnostic facilities, leading to delays in diagnosis and unreliable epidemiological data.[2]Although the prevalence of BCS is low, its consequences may be severe, ranging from portal hypertension, ascites, and variceal bleeding to progressive hepatic impairment and acute or fulminant liver failure.[2,3] The condition underscores the need to review current management strategies that may facilitate timely treatment selection and improve patient outcomes.[2] Therefore, this review aimed to raise clinical awareness and support healthcare providers in improving the diagnosis and treatment of patients with BCS.

EPIDEMIOLOGY

Global epidemiological data on BCS remains limited, particularly regarding its incidence, prevalence, geographic distribution, and patterns of occurrence across different populations. A study conducted in Italy between 2002 and 2012 reported 287 hospitalized cases of BCS, with female accounting for 54.4% and a mean age of approximately 50 years.[4] In France, a study identified 178 cases of primary BCS, equivalent to a prevalence of 4.04 cases per million population. Among the participants, 53% were male, with a mean age of 40 ± 14 years. Thrombosis was located in the hepatic veins in 121 cases (76.6%), in both the hepatic veins and inferior vena cava in 37 cases (23.4%), and in the portal vein in 5 cases (3.2%). Myeloproliferative neoplasms (MPNs) were the most commonly identified risk factor, present in 48% of cases.[5] A meta-analysis including two Asian and four European countries estimated a pooled prevalence of BCS at 11 cases per million population.[6]

CLASSIFICATION

Budd-Chiari syndrome (BCS) is classified into primary and secondary types based on the underlying cause of hepatic venous outflow obstruction. **Primary** BCS is characterized by endoluminal obstruction within the hepatic venous outflow tract, often caused by thrombosis associated with a prothrombotic state, such as hematologic disorders. Meanwhile, **secondary** BCS arises when the obstruction is caused by external compression or invasion of the hepatic venous system by lesions originating outside the vessel, such as cysts, tumors, abscesses, or other extrinsic factors.[7,8]

Based on clinical presentation and duration, BCS can be categorized into acute, subacute, chronic, and fulminant types. Acute BCS typically develops within less than one month and is characterized by abdominal pain, hepatomegaly, ascites, and the absence of collateral vein formation. Subacute BCS also presents with a relatively rapid onset, but symptoms such as ascites and hepatic necrosis are less pronounced due to the development of collateral venous circulation. Chronic BCS is often difficult to distinguish from liver cirrhosis, as it commonly presents with complications associated with cirrhotic progression. Magnetic resonance imaging (MRI) is considered superior in visualizing and differentiating between acute, subacute, and chronic forms of BCS.[9–11]

Fulminant BCS is a rare clinical condition caused by acute obstruction of all three hepatic veins, with rapid progression preventing the development of collateral circulation. It presents with a sudden onset of symptoms, including ascites, abdominal pain, and features of acute liver failure such as hepatic encephalopathy and coagulopathy.[2]

RISK FACTOR

BCS is associated with various systemic and local predisposing factors. Hematological neoplasms represent the most frequent acquired prothrombotic disorders associated with BCS. These include Philadelphia chromosome-negative MPNs, such as polycythemia vera, essential thrombocythemia, and primary myelofibrosis. Other hematological and thrombophilic conditions include paroxysmal nocturnal hemoglobinuria, antiphospholipid syndrome, Factor V Leiden mutation, prothrombin G20210A mutation, deficiencies of protein C, protein S, or antithrombin, and hyperhomocysteinemia. Drug-related and hormonal factors, particularly estrogen-containing oral contraceptives and pregnancy, may further increase

the risk of hepatic venous thrombosis. Inflammatory or immune-mediated conditions associated with BCS include Behçet's disease, inflammatory bowel disease, vasculitis, sarcoidosis, systemic lupus erythematosus, and other connective-tissue diseases. Meanwhile, intra-abdominal infections or inflammatory conditions and cytomegalovirus infection have been less commonly reported. Solid malignancies may cause secondary BCS through direct invasion or external compression of the hepatic veins or inferior vena cava. Identification of one risk factor should not preclude evaluation for additional underlying causes because multiple predisposing factors may coexist.[2,7,12,13]

PATHOPHYSIOLOGY

Obstruction of hepatic venous outflow leads to elevated sinusoidal pressure and reduced portal blood flow, thereby worsening hepatic congestion. Tissue hypoxia caused by impaired perfusion induces hepatocyte necrosis and activates hepatic stellate cells. This process contributes to the development of fibrosis, which progresses to congestive cirrhosis in more severe cases. As a compensatory mechanism, collateral circulation may form to alleviate venous congestion. However, this adaptation can promote the development of dysplastic nodules and potentially progress to hepatocellular carcinoma.[2,12] The pathophysiological mechanism underlying the development and progression of BCS is shown in **Figure 1.**

The prothrombotic state observed in BCS is associated with various underlying disorders, including protein C or S deficiency, antiphospholipid syndrome, Factor V Leiden mutation, paroxysmal nocturnal hemoglobinuria, PT gene mutation, antithrombin deficiency, and hyperhomocysteinemia. Among MPNs, PV is the most frequently identified cause of BCS, accounting for approximately 18-43% of cases. This is followed by essential thrombocythemia (ET), which is implicated in 8-14% of cases.[12]

CLINICAL MANIFESTATION AND LABORATORY FINDINGS

Clinical manifestations of BCS are highly variable and often nonspecific, with up to 20% of cases presenting without symptoms. Symptoms can be categorized based on the site of venous reflux. First, hepatic venous reflux may lead to hepatomegaly, splenomegaly, abdominal pain, distension, ascites, and portal hypertension. Second, reflux in the inferior vena cava (IVC) may present with bilateral lower extremity edema, varicose veins, including those on the chest wall, abdomen, flanks, and back, along with skin pigmentation and leg ulcers. A third category includes patients exhibiting a combination of both patterns.[14] Patients frequently present with the classical triad of abdominal pain, ascites, and hepatomegaly.[15] Furthermore, variceal bleeding occurs in approximately 5-21% of cases.[2]

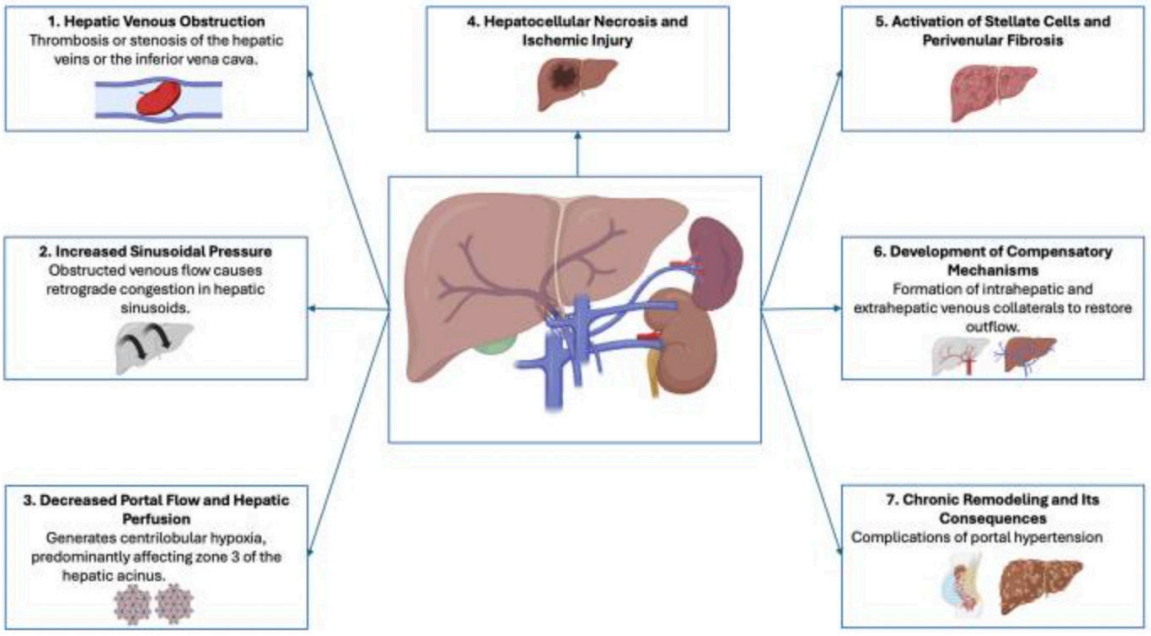

Figure 1. Pathophysiology of Budd–Chiari syndrome. Reproduced from Montes López et al. Pathophysiology of Primary Budd-Chiari Syndrome: A Narrative Review. Cureus. 2025;17(11):e96539, under the Creative Commons Attribution 4.0 International License.[12]

Evaluation for possible obstruction of the hepatic venous outflow should be considered in patients presenting with acute or persistent liver dysfunction. In acute cases, serum transaminase levels are typically elevated, although a rapid decline may also be observed. Laboratory results such as albumin, bilirubin, and international normalized ratio (INR) may vary depending on the disease course. In cases where MPNs are the underlying cause, splenomegaly is often present, and platelet counts frequently exceed $200{,}000 \times 10^9$ per liter. An elevated serum-ascites albumin gradient (SAAG) may be indicative of BCS in patients with ascites.[3]

DIAGNOSIS

The diagnostic evaluation of BCS should include confirmation of hepatic venous outflow obstruction and investigation of the underlying prothrombotic conditions. Non-invasive imaging is sufficient to establish the diagnosis in most patients, while catheter venography and liver biopsy are reserved for selected cases.[7,13,16]

LABORATORY WORKUP

Initial laboratory assessment should comprise a complete blood count, hepatic and renal biochemical profiles, serum albumin and bilirubin measurements, and coagulation testing, including prothrombin time and the INR.[7] As underlying prothrombotic disorders are frequently identified in BCS, all patients should be systematically assessed for both local and systemic predisposing conditions. Thrombophilia evaluation should cover protein C, protein S, and antithrombin activities, Factor V Leiden and prothrombin G20210A variants, and antiphospholipid antibodies, including lupus anticoagulant, anticardiolipin antibodies, and anti-β2-glycoprotein I antibodies. Positive antiphospholipid antibodies should be confirmed using a second sample collected no earlier than 12 weeks.[13] Investigation for an underlying myeloproliferative neoplasm should include JAK2 V617F mutation analysis, regardless of whether the peripheral blood cell counts are within the normal range. For JAK2-negative patients with persistent clinical suspicion of an MPN, selective CALR mutation analysis and bone marrow examination may be warranted. Paroxysmal nocturnal hemoglobinuria should also be investigated by peripheral blood flow cytometry. Protein C, protein S, and antithrombin measurements require cautious interpretation because hepatic dysfunction, an acute thrombotic event, and ongoing anticoagulation may change the levels. In particular, vitamin K antagonists may compromise the reliability of protein C and protein S assessment.[7,13]

IMAGING WORKUP

Doppler US should be used as the first-line imaging examination for diagnosing BCS. Subsequently, contrast-enhanced CT or MRI should be carried out to confirm the diagnosis, assess hepatic morphological and vascular abnormalities, and evaluate the feasibility of available therapeutic interventions.[13,16] In chronic BCS, focal liver lesions are common and most frequently represent regenerative or focal nodular hyperplasia-like nodules that develop in response to chronic vascular disturbances and altered intrahepatic perfusion. Because benign regenerative nodules may show arterial phase hyperenhancement and even portal venous or delayed phase washout, conventional enhancement criteria alone may be insufficient to distinguish BCS-related lesions from HCC.[17,18] In patients with chronic BCS, surveillance for HCC should include liver imaging and serum alpha-fetoprotein measurement every six months. Suspicious or indeterminate nodules should be further evaluated using multiphasic CT or contrast-enhanced MRI. Inconclusive results on multiphasic CT warrant further lesion characterization using MRI, preferably with a hepatobiliary contrast agent. Lesions that remain suspicious for HCC should undergo histological confirmation.[13,18] **Table 1** shows the characteristic imaging results associated with BCS.

Ultrasonography (US)

Doppler ultrasonography (US) is considered an appropriate first-line imaging modality for patients with suspected BCS.[19] Its advantages include widespread availability, low cost, and the absence of ionizing radiation.[16] The sensitivity and specificity of Doppler-US for diagnosing BCS are reported to be approximately 87.5% and 85%, respectively. However, its limitations include dependence on operator expertise, technical difficulties in patients with obesity or excessive bowel gas, and the inability to sufficiently provide consistent anatomical detail required for planning endovascular or surgical interventions.[2]

Table 1. Imaging Findings of Budd-Chiari Syndrome[16–18,20]

	Acute	Subacute and chronic
Doppler	• Absence of flow in part or all of the hepatic veins • Reversal of flow direction within some or all hepatic veins • High flow velocity detected at sites of stenosis • Alteration of flow pattern from normal phasic to absent, reversed, turbulent, or continuous • A continuous waveform pattern referred to as a "pseudoportal Doppler" signal	• Hepatic venous abnormalities including webs, stenosis, occlusion, or complete obliteration • Altered flow within the hepatic veins—absent, turbulent, or reversed; the "Miranet sign" may be observed in cases of severe stenosis or obstruction of the inferior vena cava • Intrahepatic veno-venous collateral formation, which is highly specific diagnostically, appearing as comma-shaped channels with color flow • Intrahepatic collaterals that connect to systemic circulation via subcapsular pathways • Development of collateral channels between an occluded hepatic vein and a patent hepatic vein
CT	• Dilatation of the vein accompanied by an intraluminal filling defect • Reduced enhancement of the peripheral liver parenchyma, producing a characteristic "flip-flop" pattern • Redistribution of liver volume with the development of a nodular surface • Homogeneous enhancement within the enlarged caudate lobe	• Alterations in overall liver morphology • Multiple regenerative nodules measuring approximately 0.5–4 cm in diameter • On multiphasic CT, the nodules appear homogeneous, demonstrate prominent enhancement during the arterial phase, and retain enhancement in the portovenous phase • HCC may demonstrate heterogeneous arterial phase hyperenhancement followed by portal venous or delayed phase washout, with or without an enhancing capsule; however, washout alone is not specific for HCC in BCS because it may also occur in benign regenerative nodules • Presence of extrahepatic collateral vessels • Enlargement of the hepatic artery relative to the splenic artery • Multiple collateral channels may also be identified, although with lower diagnostic confidence
MRI	• Abnormal signal intensity of the vein on T2-weighted imaging • Lack of contrast opacification in the hepatic veins and inferior vena cava • Maintenance of normal signal characteristics in the caudate lobe across all sequences • Following contrast administration, the caudate lobe shows relatively lower enhancement compared to the heterogeneously increased enhancement of the peripheral liver, with the overall pattern becoming more uniform on delayed-phase images	• Features consistent with fibrosis and chronic parenchymal liver disease • Regenerative or focal nodular hyperplasia-like nodules are commonly iso- to hyperintense on T1-weighted images and iso- to hypointense on T2-weighted images. They usually demonstrate homogeneous arterial phase hyperenhancement without diffusion restriction and may appear iso- or hyperintense during the hepatobiliary phase • HCC tends to show T1 hypointensity, T2 hyperintensity, diffusion restriction, heterogeneous arterial phase hyperenhancement, portal venous or delayed phase washout, and hypointensity during the hepatobiliary phase • Development of venous collaterals, detectable with sensitivity and specificity comparable to or even exceeding that of Doppler ultrasound

Computed Tomography (CT)

Computed tomography (CT) is the most frequently used imaging modality in cases with suspected BCS. This modality allows for visualization of vascular abnormalities, hepatic morphological changes, and vascular mapping, which is particularly useful before interventional procedures or surgery.[16] The imaging of CT can identify the location and extent of venous thrombosis or stenosis and is valuable for delineating vascular anatomy before transjugular intrahepatic portosystemic shunt (TIPS) placement.[20] The venography shows a sensitivity and specificity of approximately 86.1% and 97.3%, respectively. However, its main limitations include higher cost and reduced reliability in consistently and accurately detecting venous abnormalities.[2]

Magnetic Resonance Imaging (MRI)

Magnetic resonance imaging (MRI) offers higher diagnostic accuracy compared to both CT and Doppler US. This method uses a valuable imaging modality as it enables comprehensive assessment of vascular abnormalities, hepatic morphological changes, and detailed mapping of both intrahepatic and extrahepatic vasculature, as well as the evaluation of hepatic nodules. However, MRI may be challenging to carry out in patients with ascites due to the presence of artifacts and the difficulty of maintaining a supine position during the examination.[16,19]

Catheter Venography

Catheter venography is not routinely required when BCS has been clearly diagnosed using non-invasive imaging. This method is recommended when the diagnosis remains uncertain and may also assess the extent of hepatic venous outflow obstruction before endovascular treatment. When appropriate, diagnostic venography and endovascular intervention may be carried out during the same session.[13,16]

HISTOPATHOLOGY WORKUP

Histological assessment is generally unnecessary once the diagnosis of BCS has been established by radiological imaging. Liver biopsy may be warranted when clinical suspicion persists despite apparently patent hepatic veins, particularly for identifying small hepatic vein BCS.[13] Microscopic abnormalities include sinusoidal dilatation and congestion, centrilobular hepatocellular injury or necrosis, and perisinusoidal or perivenular fibrosis. These abnormalities are not specific to BCS and may also occur in other hepatic vascular disorders. Therefore, interpretation should consider the clinical and radiological results, particularly when differentiating BCS from sinusoidal obstruction syndrome and hepatic congestion related to right-sided cardiac or pericardial disease.[21]

TREATMENT

In patients with primary, non-fulminant BCS, management follows a stepwise strategy beginning with anticoagulation and treatment of the underlying condition. This is followed by percutaneous angioplasty, TIPS, and liver transplantation in patients who do not achieve an adequate response to the preceding treatment.[13]

MEDICAL THERAPIES

Anticoagulation, as the first-line therapy, is administered to all patients with BCS. Low-molecular-weight heparin (LMWH) is recommended as the initial treatment for a duration of at least 5-7 days, followed by lifelong warfarin therapy, aiming for an INR target of 2-3. Potential complications should be closely monitored, such as bleeding. Heparin-induced thrombocytopenia (HIT) occurs more frequently with unfractionated heparin (UFH) than with LMWH. Therefore, UFH is best avoided, with LMWH preferred for initial anticoagulation. In pregnant patients, warfarin is contraindicated, and LMWH is the preferred anticoagulant.[2,13]

Direct oral anticoagulants (DOACs) offer the advantage of not requiring regular monitoring, as is necessary with vitamin K antagonists (VKAs), thereby improving convenience and potentially enhancing patient adherence. Although DOACs are not yet broadly recommended as standard therapy for BCS, some studies suggest relative safety and potential benefits for patients who struggle to maintain a therapeutic INR or require long-term anticoagulation.[22] A multicenter study showed that DOACs are generally effective and safe for long-term use. Four cases of spontaneous major bleeding were reported, while the overall transplant-free survival reached 91.6% at five years. However, further studies including larger populations are still required.[23]

A large comparative study presented at an international conference reported no significant differences between DOACs and warfarin in terms of safety, efficacy, need for TIPS, or incidence of spontaneous bacterial peritonitis. These results suggest that DOACs may be a potential alternative to warfarin, although the evidence is currently limited to conference-level data.[24] Various types of DOACs exhibit different metabolic profiles, as shown in **Figure 2.** However, the use in patients with renal impairment remains limited and requires careful monitoring due to reduced drug clearance and an increased risk of bleeding. Evidence regarding safety and use in advanced chronic kidney disease, particularly among patients undergoing dialysis, remains limited. [25]

THROMBOLYSIS

Thrombolysis should not be used as a standalone treatment for BCS, as both systemic and locally administered thrombolytic therapies have low success rates and carry a substantial risk of major bleeding. Catheter-directed thrombolysis may be used as an adjunct during percutaneous transluminal balloon angioplasty to facilitate initial venous recanalization or to manage immediate rethrombosis occurring during the procedure.[13]

ANGIOPLASTY RECANALIZATION

Angioplasty and venous recanalization are carried out to relieve hepatic venous outflow obstruction and improve venous hemodynamics. In accordance with the stepwise management strategy, percutaneous balloon angioplasty is the preferred first invasive

	Dabigatran	Apixaban	Rivoraxaban	Edoxaban	Betrixaban
Mechanism of action	Direct thrombin inhibitor	Direct Factor Xa inhibitor			
Prodrug	Yes	No			
Half-life	12–17 hours	12 hours	5–9 hours	10–14 hours	20–27 hours
Cmax	1–2 hours	3 – 4 hours	2–4 hours	1-2 hours	3–4 hours
Renal excretion	80%	27%	66%	50%	10%
Liver metabolism	20%	75%	65%	50%	<1%
FDA approval	2010	2013	2011	2014	2017
FDA indications	• Stroke prevention in NVAF • DVT and PE treatment • DVT and PE secondary prevention	• Stroke prevention in NVAF • Post-operative DVT prophylaxis • DVT and PE treatment • DVT and PE secondary prevention	• Stroke prevention in NVAF • Post-operative DVT prophylaxis • DVT and PE treatment DVT and PE secondary prevention	• Stroke prevention in NVAF • DVT and PE treatment	• VTE prophylaxis
Interactions	P-gp inhibitors	Strong inhibitors of CYP3A4 and P-gp	Strong inhibitors of CYP3A4 and P-gp	P-gp inhibitors	

Figure 2. Comparison of direct oral anticoagulants (DOACs). Reproduced from Miceli G, Ciaccio AM, Tuttolomondo A. Challenges and Opportunities of Direct Oral Anticoagulant (DOAC) Therapy in Complex Clinical Scenarios: A Comprehensive Review and Practical Guide. *J Clin Med*. 2025;14:2914, under the Creative Commons Attribution 4.0 International License.[25]

intervention when a technically accessible short-segment, membranous, or segmental obstruction is identified in a hepatic vein or the inferior vena cava.[2,13] In patients with partial or localized narrowing of the upper hepatic veins or the suprahepatic inferior vena cava, percutaneous transluminal angioplasty, with or without stent placement, may effectively restore hepatic venous drainage.[26] Successful recanalization may reduce hepatic congestion and preserve liver function. Failure to recanalize the obstructed segment, procedural failure, or inadequate clinical response warrants consideration of TIPS as the next treatment option.[13]

TRANSJUGULAR INTRAHEPATIC PORTOSYSTEMIC SHUNT (TIPS) AND SURGICAL SHUNTING

TIPS should be considered for patients with primary, non-fulminant BCS who do not achieve an adequate response to anticoagulation and treatment of the underlying condition. This method is particularly appropriate when hepatic venous recanalization is not technically feasible, angioplasty has failed, or adequate clinical improvement has not been achieved after angioplasty. TIPS decompresses the congested hepatic and portal venous systems and has become the preferred shunting procedure in the stepwise management of BCS. Because hepatic vein obstruction may make the procedure technically challenging, TIPS should be carried out at an experienced referral center. A direct intrahepatic portocaval shunt may be required when conventional access through a hepatic vein is not possible.[13] TIPS may also be used as a bridging treatment in selected patients awaiting liver transplantation.[2,26] Surgical portosystemic shunting has largely been replaced by TIPS and is now considered obsolete in contemporary BCS management. Therefore, it is not recommended as a routine alternative when endovascular treatment is unsuccessful or technically impossible.[13]

LIVER TRANSPLANT

Liver transplantation represents the final treatment option for primary, non-fulminant BCS in patients who remain unresponsive despite anticoagulation, treatment of the underlying condition, percutaneous angioplasty, and TIPS. Evaluation for liver transplantation should be initiated early in patients presenting with BCS-related acute liver failure. This procedure should not be delayed until completion of the entire stepwise treatment strategy.[13,26] Liver transplantation should also be considered when TIPS is technically unfeasible or unsuccessful and progressive hepatic failure persists.[13] The reported 5-year and 10-year post-transplant survival rates are approximately 71%

and 68%, respectively. Recurrent BCS has been reported in 5–11% of patients, supporting continued anticoagulation after transplantation unless the underlying prothrombotic disorder has been corrected by the procedure.[2]

CONCLUSION

In conclusion, BCS is a condition caused by partial or complete obstruction of hepatic venous outflow. The clinical presentation varies widely, ranging from asymptomatic cases to fulminant liver failure. The management of this condition should be designed to suit the patient's clinical condition and disease severity.

CONFLICT OF INTEREST

The authors declare no conflicts of interest related to this work.

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DH, BJW, LR, FG, JW, AW contributed to the conception, design, drafting of the manuscript, data acquisition, literature review, and manuscript revision. All authors approved the final version of the manuscript.

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