Review Article

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Budd-Chiari Syndrome: Narrative Insights into Clinical Spectrum, Genetics and Contemporary Management Strategies

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ABSTRACT

Budd-Chiari Syndrome (BCS) is a rare potentially life-threatening disorder that occurs due to the hepatic venous outflow obstruction at the hepatic veins and/Inferior Vena Cava (IVC) causing hepatic congestion, portal hypertension, and inconsistent liver dysfunction. Clinical manifestations are heterogeneous, such as acute fulminant hepatic failure, chronic insidious ascites and hepatomegaly, and its manifestation varies with the rate of obstruction, the presence of veins, and other co-morbidities. Diagnosis depends on clinical examination, laboratory test and imaging, the first-line method is Doppler ultrasound and Contrast Enhanced Computed Tomography (CECT), Magnetic Resonance Imaging (MRI), or venography are used as second-line imaging modalities. Etiology is in most cases prothrombotic by nature and inherited, somatic, and polygenic risk factors play the role such as Factor V Leiden, JAK2 V617F and other molecular defects. Management is based on the combined multidisciplinary approach; anticoagulation is administered in the first step, followed by endovascular procedure, including angioplasty, stent placement, or Transjugular Intrahepatic Portosystemic Shunt (TIPS) and finally liver transplantation is carried out in refractory cases. Early detection and early intervention are important for improving clinical outcomes. This narrative review offers a clear overview of clinical, genetic and management aspects of BCS with a special focus on the recent developments and the emerging treatment plans.

Keywords: Anticoagulation, Hepatic venous outflow obstruction, Imaging, Myeloproliferative neoplasms, Thrombophilia

INTRODUCTION

Budd-Chiari Syndrome (BCS) is a life-threatening rare disorder caused by the obstruction of the hepatic venous outflow at the level of hepatic veins and/or Inferior Vena Cava (IVC) level, resulting in hepatic congestion, portal hypertension and the degree of liver dysfunction [1]. The clinical manifestations include fulminant hepatic failure to chronic ascites and mild hepatomegaly; the typical triad of the manifestations is pain in the abdomen, ascites and hepatomegaly, but the manifestation depends on geography and etiology [1,2]. Early identification of BCS is important since timely anticoagulation, endovascular therapy (angioplasty/stenting/thrombolysis), Transjugular Intrahepatic Portosystemic Shunt (TIPS) and, in a few instances, liver transplantation, have a significant impact on the outcomes [3]. In the past few decades with better imaging, increased use of hypercoagulability testing and an increasing range of therapeutic options, the natural history of BCS has altered in most centres [4].

The BCS is named after George Budd who in 1845, described a group of patients with obstructive lesions of hepatic veins, and Hans Chiari who made further clinicopathological observations in the late 19th century [5]. A meta-analysis and systematic review of articles estimated that the pooled incidence of BCS is approximately 1 case per million population per year and the pooled prevalence approximately 11 cases per million suggesting that BCS is a rare disease, though these figures may vary significantly by region and healthcare system [6]. The frequency of BCS is largely based on case-series conducted in hospitals as opposed to population-based registry investigations [7]. In an older Eastern Indian study 30 consecutive BCS patients in 5 years were analysed and reported causes for it such as, age (approximately 32.7 years), male predominance and type of obstructions, but not incidence or prevalence relative to the general population [7]. A different study of 49 patients studied obstruction patterns and established hepatic vein thrombosis as commonest type, however, giving no denominator to calculate incidence [8]. Therefore, the incidence and prevalence per million population cannot yet be estimated with any degree of certainty because of the lack of data on BCS in India [8]. The narrative review gives a proper detail of Budd-Chiari syndrome, its clinical spectrum, diagnostic methods, genetic and molecular etiology, classification, prognostic index, and current management interventions.

Beyond all the reported pooled estimates, epidemiology of BCS still remains incompletely defined considering global level because of marked geographic heterogeneity along with the methodological limitations in available studies [6,8]. Very significant regional variation exists in etiological patterns in which IVC-predominant obstruction is reported more commonly in regions such as Asia usually in India and China; whereas hepatic vein thrombosis associated with underlying prothrombotic disorders is more usually described in Western populations [6,9]. In India, studies have shown that hepatic vein thrombosis is the most common form of obstruction, with IVC involvement less frequent [7,8]. Furthermore, inconsistencies into diagnostic criteria, variability regarding access to advanced imaging modalities along with differences in referral patterns contribute into potential underdiagnosis, misclassification usually which is seen in low- and middle-income countries [10,11]. The absence of properly standardised international registries as well as uniform reporting frameworks restrict comparisons across various countries worldwide [10,11].

These persistent knowledge gaps have important implications. Uncertainty regarding true distribution of etiological subtypes as well as regional risk factor profiles limits development of targeted strategies into prevention, region-specific algorithms of BCS management [6]. It also hampers planning of health policy, allocation of specialised vascular as well as transplant services, design of adequately powered multicentric studies [6]. Addressing all these gaps through coordinated international registries along with harmonised criteria of diagnosis is essential for better defining global burden, improve outcome-oriented research in BCS [10,11].

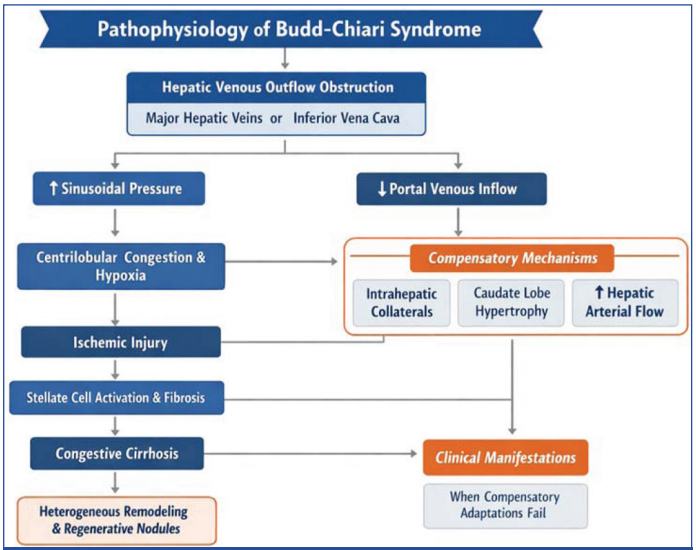
Search Strategy

Relevant literature was identified through searches of PubMed, Scopus, and Google Scholar using keywords including 'Budd-Chiari

Syndrome', 'hepatic venous outflow obstruction', 'thrombophilia', 'JAK2 mutation', and 'TIPS'. Articles published in English were reviewed and relevant contemporary evidence was narratively synthesised.

Pathophysiology of Budd-Chiari Syndrome

The pathophysiology of primary BCS is usually driven through hepatic venous outflow obstruction resulting into increased sinusoidal pressure and impaired hepatic drainage [12]. According to recent integrative models, obstruction whether inclusive of major hepatic veins or IVC produces retrograde transmission of pressure to hepatic sinusoids leading to sinusoidal dilatation, centrilobular congestion as well as hepatocellular hypoxia [12]. Reduced portal venous inflow occurs secondary to elevated sinusoidal resistance further aggravating ischaemic injury [12]. This haemodynamic imbalance contributes into activation of hepatic stellate cells thus promotes perivenular fibrosis ultimately progressing to congestive cirrhosis in chronic cases [12,13]. Compensatory mechanisms which include intrahepatic collateral formation, caudate lobe hypertrophy due to preserved direct venous drainage into IVC also the increased hepatic arterial flow ("arterialisation") partially mitigate congestion but contribute to heterogeneous parenchymal remodeling, regenerative nodule formation [12,13]. Clinical manifestations in case of BCS arise when all these compensatory adaptations fail to counteract persistent venous hypertension [13]. Pathophysiology of Budd-Chiari Syndrome is depicted in [Table/Fig-1].



[Table/Fig-1]: Pathophysiology of Budd-Chiari Syndrome (Image created by authors).

In addition to thrombosis and stenosis of hepatic veins or the IVC, non thrombotic etiologies which are inclusive of extrinsic compression by space-occupying lesions (including hepatic cysts, benign or malignant tumors) can produce functional hepatic venous outflow obstruction thereby mimicking BCS physiology through similar mechanisms of increased sinusoidal pressure along with congestion leading to hepatomegaly [14,15]. Hepatomegaly is known as one of the most common manifestations of BCS arising from hepatic congestion along with capsular stretching secondary to impaired venous drainage often in combination with ascites, abdominal pain [16].

Clinical Spectrum of Budd-Chiari Syndrome

Budd-Chiari Syndrome (BCS) is a term used to describe the presence of the obstruction of hepatic venous outflow (hepatic veins and/or IVC) resulting in congestion of the liver, high sinusoidal pressure and secondary portal hypertension [17]. The clinical manifestation is very heterogeneous in relation to the rate of obstruction development (acute, subacute and chronic), the number of affected veins, the development or not of collateral venous drainage and presence or absence of underlying liver damage or co-morbidity [17,18].

In acute or fulminant cases, the symptoms are likely to present within days to weeks and includes severe right upper quadrant/epigastric abdominal pain, marked hepatomegaly, sudden onset ascites, jaundice, coagulopathy and in some cases hepatic failure with encephalopathy [18]. Chronic BCS, on the contrary, it develops over months to years; its characteristics are more insidious [19]. The patients may present with progressive ascites, abdominal distension, hepatomegaly, splenomegaly, signs of portal hypertension (e.g. variceal bleeding) and less evident jaundice or liver enzyme derangements [19]. Physical examination can reveal pedal oedema and collateral veins formation (e.g. varices on the abdominal wall) and occasionally truncal subcutaneous veins in the event of an obstruction of the IVC [19,20]. Clinical spectrum and presentation of BCS is mentioned in [Table/Fig-2].

Aspect	Description
Definition	BCS refers to obstruction of hepatic venous outflow (hepatic veins and/or IVC), leading to hepatic congestion, elevated sinusoidal pressure and secondary portal hypertension [17].
Pathophysiology	Obstruction of hepatic venous drainage results in hepatic congestion, impaired venous return, increased sinusoidal pressure and subsequent portal hypertension [17].
Clinical variability	The clinical presentation is heterogeneous and depends on the rate of obstruction development (acute, subacute, or chronic), the number of veins involved, the presence or absence of collateral venous drainage, and the presence of underlying liver disease or co-morbidities [17,18].
Acute/Fulminant BCS	Onset typically occurs within days to weeks. Symptoms include severe right upper quadrant or epigastric pain, marked hepatomegaly, sudden-onset ascites, jaundice, coagulopathy and in severe cases, hepatic failure with encephalopathy [18].
Chronic BCS	Develops gradually over months to years. Characterised by progressive ascites, abdominal distension, hepatomegaly, splenomegaly, signs of portal hypertension (e.g., variceal bleeding), and less prominent jaundice or liver enzyme derangements [19].
Physical examination findings	Findings may include pedal oedema, collateral venous formation such as abdominal wall varices, and occasionally truncal subcutaneous veins in the event of IVC obstruction [19,20].

[Table/Fig-2]: Clinical spectrum and presentation of BCS.

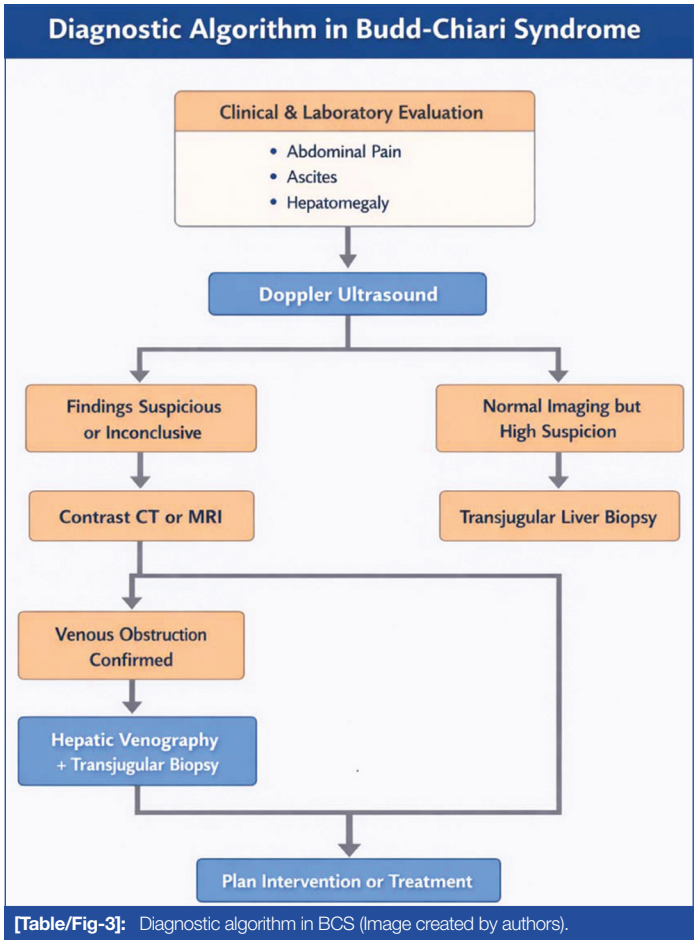
Diagnosis and Imaging Approach in Budd-Chiari Syndrome

The diagnosis of BCS is mainly clinicoradiological as there is high clinical suspicion due to the triad of abdominal pain, rapid onset of ascites and hepatomegaly (or less vigorous subacute/chronic manifestations), and laboratory tests of liver injury/coagulation and an initial imaging tests such as Doppler ultrasound are necessary since it is readily available, non invasive and the preferred first-line test [21]. Doppler findings include absent or reversed hepatic-venous flow, flat/monophasic hepatic veins waveform, slow or hepatofugal portal flow, and signs showing disturbance in IVC flow [22,23]. In circumstances, where Doppler is uncertain or to establish extent and manage intervention, cross-sectional imaging by means of CECT and most importantly MRI is employed to verify and map venous obstruction and secondary hepatic changes [22]. Characteristic cross-sectional appearances are non visualisation or abrupt termination of one or more of the hepatic veins or the suprahepatic IVC, caudate-lobe hypertrophy (which may be relatively spared due to autonomous venous drainage), non uniform / mosaic parenchymal enhancement because of segmental perfusion defects, intrahepatic and extrahepatic collateral vessels and marked ascites [22]. MRI is better at depicting thrombosis and parenchymal perfusion patterns whereas CT is meta-analytic results indicate that Doppler, CT and MRI are highly diagnostic with MRI having the highest discriminative ability in pooled studies [22,24].

Invasive venography (hepatic venography/IVC cavography) is the reference standard in cases when non invasive imaging is not conclusive or endovascular therapy is under consideration as it directly records the level and length of obstruction and allows pressure measurements, immediate intervention (angioplasty/

stenting, TIPS) [25]. In cases where there is a suspicion of outflow obstruction (such as short-segment occlusion or obstruction confined to small intrahepatic veins) is present, but normal imaging has been obtained, transjugular liver biopsy or histology may be needed to demonstrate centrilobular congestion, sinusoidal dilatation or evidence of chronic venous outflow impairment [26]. Thus, a realistic diagnostic algorithm to be applied in practice is as follows: clinical/laboratory examination and testing → Doppler ultrasound as the initial test) → confirmatory contrast CT or MRI with mapping → hepatic venography (with or without associated transjugular biopsy) in cases when planning intervention or when non invasive tests are discordant [26].

Despite having advancements in Doppler ultrasonography along with cross-sectional imaging further diagnosis of BCS remains challenging in routine practice [10]. Early, partial hepatic venous obstruction can be radiologically subtle resulting into underdiagnosis whereas chronic cases can mimic cirrhosis of other etiologies due to regenerative nodules as well as portal hypertension [10]. Doppler ultrasonography is operator-dependent which can further yield false-negative results in technically difficult examinations usually in obese or ascitic patients [27]. Cross-sectional imaging along with CECT and MRI improves sensitivity but can not reliably help to distinguish acute thrombosis from chronic fibrotic occlusion [11]. Invasive hepatic venography which was once considered as gold standard is now reserved for equivocal cases or prior to endovascular intervention due to its procedural risks [10,27]. Furthermore, variability into access to advanced imaging as well as thrombophilia testing in low-resource settings contributes in delayed-missed diagnosis [11]. These diagnostic uncertainties can thereby postpone timely anticoagulation or interventional therapy thus adversely affecting outcomes in BCS patients [11,27]. Diagnostic algorithm in BCS is depicted in [Table/Fig-3].



[Table/Fig-3]: Diagnostic algorithm in BCS (Image created by authors).

Few recent advances in imaging technology can help in refining precision of diagnosis in BCS patients [28]. Contrast Enhanced Ultrasound (CEUS) allows dynamic evaluation of hepatic perfusion without need of ionising radiation which can prove helpful to differentiate acute thrombosis from chronic fibrotic occlusion [28,29]. Elastography techniques which are inclusive of transient elastography and MR elastography provides non invasive assessment of liver stiffness which can further assist in evaluating progression toward fibrosis or cirrhosis in chronic cases of BCS [30,31]. Advanced MRI perfusion imaging, Three-Dimensional (3D) venographic reconstruction can further improve pre-interventional planning for angioplasty as well as TIPS procedures [28,32]. Adequate standardisation, broader validation still remains an important aspect before routine incorporation into clinical algorithms for BCS [28].

Classification and Prognostic Scoring of BCS

The classification of BCS is essential in the prognosis and treatment courses. It is possible to classify it in terms of anatomic, temporal, and prognostic. The anatomical classification of BCS is mainly based on location of the obstruction: obstruction on the hepatic vein, obstruction on the IVC, and a combination of both [24,33]. Hepatic vein thrombosis is a condition with acute symptoms, whereas insidious IVC obstruction can progress into the fibrotic changes with time [24]. The membranous IVC obstruction is a frequent cause of BCS that is linked to various clinical manifestations as compared to thrombosis of the hepatic vein [24,33].

Temporally, BCS is further classified due to its duration: acute (which is below 2 months), subacute (between 2 to 6 months) as well as chronic (more than 6 months); however, precise temporal cut-offs are not standardised uniformly across literature [34]. The temporal classification of BCS has practical-clinical utility as it broadly reflects regarding severity of hepatic injury, haemodynamic adaptation and urgency of intervention [34]. Acute BCS usually presents having rapid onset of hepatic dysfunction characterised by severe abdominal pain, hepatomegaly, ascites, and, in some cases, fulminant hepatic failure [34]. The cases of acute BCS thereby requires an urgent anticoagulation as well as early consideration of endovascular therapy [34]. In contrast, chronic BCS is more usually associated having long-standing hepatic venous outflow obstruction further leading to progressive fibrosis, cirrhosis, and established portal hypertension with collateral formation [34]. In chronic BCS, management strategies further focus on long-term complications inclusive of ascites control, variceal surveillance also evaluation for TIPS or liver transplantation [34]. Thus, although temporal cut-offs are not rigidly standardised such classification helps into stratification of risk, therapeutic planning also prognostic assessment [34]. Rotterdam BCS index, Clichy score and BCS-TIPS prognostic index are the scoring systems have been created to determine outcomes in BCS patients prognostically [35]. These indices take into account liver functioning, presence of ascites and required interventions to predict outcomes [35]. As an example, Rotterdam score has been found as the most effective index of discrimination in regard to 3-month mortality in BCS and should be considered first when deciding on the urgency of treatment [35]. Also, it has been observed that the BCS-TIPS score is a good predictor of survival in patients who have undergone TIPS surgeries [35]. Classification systems and outcome prediction in BCS is described in [Table/Fig-4].

Although several prognostic indices have been developed for BCS, their practical-clinical application needs further contextual interpretation [32]. The Rotterdam score which is inclusive of encephalopathy, ascites, prothrombin time and bilirubin is usually useful at initial presentation to stratify short-term (3-month) mortality risk as well as guide urgency of intervention [32]. In clinical practice, a high Rotterdam score should prompt an early evaluation for TIPS as well as liver transplantation rather than prolonged conservative management [32]. The Clichy score which is derived from earlier cohorts is less usually used into contemporary practice because of its limited validation in diverse populations [36]. Comparatively,

Classification type	Subtypes/Description	Clinical significance	References
Anatomical	- Hepatic vein obstruction - Inferior vena cava (IVC) obstruction - Combined obstruction (hepatic vein + IVC)	Hepatic vein thrombosis presents acutely; IVC obstruction may progress insidiously to fibrosis; membranous IVC obstruction has distinct clinical manifestations.	[24,33].
Temporal	- Acute (<2 months) - Subacute (2-6 months) - Chronic (>6 months)	Acute BCS often leads to fulminant liver failure; chronic BCS may result in cirrhosis and portal hypertension.	[34].
Prognostic/ Scoring Systems	- Rotterdam BCS Index - Clichy Score - BCS-TIPS Prognostic Index	Predicts outcomes based on liver function, ascites, and intervention needs; Rotterdam score effective for 3-month mortality; BCS-TIPS predicts survival after TIPS.	[35].

[Table/Fig-4]: Classification systems and outcome prediction in BCS.

the Rotterdam score has showed superior discriminative ability for short-term mortality prediction in modern cohorts whereas Clichy score has limited external validation as well as it is less frequently applied [32,36]. The BCS-TIPS prognostic index is most specifically applied in patients who are undergoing TIPS it also helps predict post-procedural survival, thereby assisting in selection between TIPS and early transplantation [36,37]. The BCS-TIPS score is usually said to be very useful after TIPS placement for identifying patients at higher risk of early mortality who can benefit from closer surveillance or expedited transplant evaluation [36,37]. However, these models were derived from relatively small cohorts which may not fully account for modern interventional advances as well as have limited external validation across different ethnic populations. Additionally, none of the currently available scoring systems uses molecular or genetic risk factors which can further limit their prognostic precision in era of personalised medicine [37,38]. Therefore, prognostic scores must complement rather than replace clinical judgment, imaging findings as well as dynamic assessment of treatment response while determining therapeutic escalation [37,38]. Serial reassessment which is done using these indices during follow-up can help to provide greater clinical value than a single baseline calculation [37,38].

Beyond clinical scoring systems, circulating biomarkers are also further being explored for prognostic stratification in case of patients having BCS [36,39]. Elevated D-dimer levels can reflect ongoing thrombosis whereas markers of hepatic fibrosis such as hyaluronic acid as well as procollagen peptides correlate with chronicity while progression into cirrhosis [36,39]. Inflammatory mediators, endothelial dysfunction markers are also under investigation for their potential helpful role in predicting response to TIPS; risk of hepatic decompensation [37,40]. Although no biomarker is currently incorporated into standard prognostic indices; integration of molecular-biochemical markers having established scores such as the Rotterdam index can help to enhance risk prediction in the era of personalised medicine [32,35].

Genetic and Molecular Basis of BCS

A prothrombotic milieu usually contributes in the development of BCS, where inherited and somatic/genetic mutations are critical factors [41]. Factor V Leiden (the Arg506Gln mutation in the F5 gene) is the most frequently inherited risk factor, in BCS patients [41]. There are also deficiencies of the natural anticoagulants like protein C, protein S, and antithrombin III, which are directly associated with familial BCS by having particular point mutations leading to impairment of the anticoagulant effect of protein C [42]. Furthermore, Methylenetetrahydrofolate Reductase (MTHFR) C677T variation and prothrombin G20210A polymorphism are associated with specific populations [43]. Screening for inherited thrombophilia is usually relevant in younger age patients, individuals without any identifiable provoking factors, or those presenting having recurrent thrombotic events [43]. However, routine universal screening in all patients of BCS remains debated due to cost-effectiveness considerations as well as the variable impact on long-term therapeutic decisions [42,43].

Myeloproliferative Neoplasms (MPNs) is a highly notable risk factor considering the somatic mutation of BCS. The JAK2 V617F mutation is found in a massive proportion (between 40-60%) of BCS patients [43]. From a clinical point-of-view; identification of JAK2 V617F mutation must further prompt comprehensive hematological evaluation even in absence of cytosis as an occult or early-stage MPN can be present [43]. Importantly, this has direct implications clinically as current evidence further supports routine screening for JAK2 V617F mutation in all patients diagnosed having BCS, even in the absence of overt hematological abnormalities [43]. Identification of an underlying MPN thereby influences long-term management inclusive of consideration of cytoreductive therapy, prolonged or indefinite anticoagulation as well as structured hematology follow-up [41,43,44].

In such patients, management extends beyond anticoagulation alone as well as it can require risk-adapted cytoreductive therapy based upon bone marrow findings, haematologic parameters [41,44]. Moreover, detection of JAK2 mutation can precede clinical manifestation of overt MPN thereby helps providing significant diagnostic value [45]. Other driver genes of MPNs like Calreticulin (CALR), MPL are also advised to be tested for consideration of BCS [41,44]. Testing for CALR, MPL mutations is mainly important in JAK2-negative cases for avoiding underdiagnosis of MPN-associated BCS [41,44,45]. Although MPN-associated mutations are said to be important for etiological classification, therapeutic planning but their independent prognostic impact on overall survival in BCS patients remains incompletely defined [41,44]. Furthermore, all these mutations also assist in etiologic clarification while guiding hematologic management and their direct correlation with severity of hepatic disease, response to interventional therapies in BCS remains uncertain [44].

More recent whole exome and genome association studies are narrowing down risk further: a Chinese cohort of 500 patients found multiple Single Nucleotide Polymorphism (SNPs) (e.g. rs1042331 and many others) to be associated with BCS and created a Polygenic Risk Score (PRS) model with high discriminatory power (AUC >0.9 in external replication) to predict susceptibility [46]. PRS aggregate multiple low-penetrance genetic variants for further estimation of cumulative thrombotic susceptibility but their application in BCS cases remains investigational as most PRS models are derived from specific ethnic cohorts as well as lacks broad external validation [46]. At present, PRS lacks standardised thresholds for guiding duration of anticoagulation, interventional decision-making as well as transplant referral thereby it must not influence routine clinical practice outside research settings [46]. At the molecular level beyond thrombosis risk, gene expression analysis of liver tissue in BCS patients reveals dysregulation of extracellular matrix remodelling, growth factor signalling, angiogenesis genes and vascular remodelling genes; acute disease, chronic disease phases differ in their expression of genes (e.g. MMP7, SCG10 up in acute, thrombospondin-1 down in chronic) [47]. Ribonucleic Acid (RNA) sequencing studies of blood also suggest differential expression in genes involved in proteasome function, cellular stress responses, and immune components (e.g. GFI1B, lipocalin-2) [47,48]. Although all these molecular insights enhance understanding of disease pathogenesis which can further offer future therapeutic targets their immediate clinical applicability for risk stratification or prognostication remains limited [47,48].

Global Disparities and Barriers to Care in BCS

BCS shows marked geographic heterogeneity in etiology, diagnostic access as well as in treatment availability [7,49]. In Western countries like United States-France, hepatic vein thrombosis associated with myeloproliferative neoplasms predominates whereas in regions such as China and India, IVC obstruction is more usually reported. Disparities in access to advanced imaging, molecular diagnostics (e.g., JAK2 testing), interventional radiology services as well as liver transplantation usually influence outcomes [7,9,49,50]. In many low- and middle-income settings, delayed diagnosis, limited availability of TIPS or transplant programs further results into advanced-stage presentation as well as restricted therapeutic escalation [9]. Socio-economic barriers, limited infrastructure for anticoagulation monitoring also variable clinical awareness among people further contributes into inequities in BCS cases [9,49]. Addressing all these issues through regional registries, standardised diagnostic pathways along with an expansion of interventional capacity remains very essential for improving global outcomes in BCS patients [9,49,50].

Digital health and telemedicine in BCS care: Digital health strategies can play role in offering opportunities to improve continuity of care in BCS usually in resource-limited settings [51]. Telemedicine platforms can help to facilitate remote anticoagulation monitoring, early identification of ascites progression as well as multidisciplinary coordination between hepatologists, haematologists and interventional radiologists [51]. Mobile health applications for INR tracking-symptom reporting can enhance adherence of patients while also reducing delayed escalation of therapy in BCS patients [51]. The formal evidence in BCS is limited, integration of telehealth into chronic liver disease management frameworks can prove helpful to mitigate geographic disparities, improve long-term outcomes [51].

Management Approaches in BCS

Treatment of BCS by the first-line management methods typically entails anticoagulation therapy, in which the Direct Oral Anticoagulants (DOAC) like apixaban and rivaroxaban are becoming more popular as compared to the traditional low-molecular-weight heparin and vitamin K antagonists [52,53]. DOACs are as effective and have benefits in respect of ease of administration, but additional randomised trials are justified to verify these results [52].

Given the important role of anticoagulation in management of BCS, therapy is recommended lifelong usually in cases when an underlying prothrombotic disorder or myeloproliferative hepa is identified [54]. The primary therapeutic goals of anticoagulation are prevention of propagation of thrombus, recurrence as well as progression to hepatic decompensation rather than complete radiological recanalisation [54]. Initial treatment is inclusive of low-molecular-weight heparin which is followed by transition to vitamin K antagonists; emerging data further suggest that DOACs such as apixaban as well as rivaroxaban shows comparable efficacy, safety in selected patients although randomised trials also remain limited [54]. Among DOACs, apixaban can further help to offer a relatively favorable bleeding profile in patients having underlying liver dysfunction though robust head-to-head comparative trials in BCS are lacking [54,55]. In patients having cirrhosis, anticoagulation is not contraindicated per se but it further requires individualised assessment of bleeding risk usually in the presence of varices, thrombocytopenia as well as prior hemorrhage [54,55]. Endoscopic screening for esophageal varices, also consideration of prophylactic band ligation can be done before or during anticoagulation in high-risk individuals having BCS [55]. Endpoints of therapy are not only defined by thrombus resolution but it is also through prevention of progression, recurrence, hepatic decompensation. In cases having concomitant portal vein thrombosis, anticoagulation still remains indicated unless active bleeding is present in patient [55]. Even in idiopathic cases of BCS without any identifiable thrombophilia, extended or indefinite anticoagulation is usually favored due to the risk of recurrence although optimal duration in such cases remains still debated [55]. Careful monitoring of INR (for warfarin), renal function (for DOACs) along with periodic reassessment of bleeding risk are very essential aspects to optimise long-term outcomes [54,55]. Dynamic reassessment of thrombotic versus haemorrhagic risk throughout follow-up is very crucial mainly in patients having fluctuating liver function or portal hypertension [54,55].

Percutaneous transluminal angioplasty may be effective in restoration of the venous outflow in patients with short-segment hepatic vein stenosis, and stenting may be implemented in the event that only angioplasty is inadequate [56]. TIPS has proved to be effective reducing portal pressure and increase survival in patients who fail to respond to medical and interventional therapy interventions, as well as in patients with diffuse involvement of the hepatic vein [57]. Liver transplantation is still a final treatment option in patients who have end-stage liver disease or patients who do not respond to other interventions [58]. A consensus guideline has been offered by Asian Pacific Association for the Study of the Liver (APASL) and suggests early intervention through clinical severity and anatomical considerations and not just an expectant management approach [59]. The present changing paradigm highlights the significance of a multidisciplinary approach in the treatment of BCS, which focuses on maximising patient outcomes by responding to them in a timely and appropriate manner [59]. Stepwise management of BCS is depicted in [Table/Fig-5].

Treatment modality	Description/Indication	References
Anticoagulation therapy	First-line management. Direct oral anticoagulants (DOACs) such as apixaban and rivaroxaban are increasingly used due to ease of administration and comparable efficacy to low-molecular-weight heparin and vitamin K antagonists.	[52,53]
Percutaneous transluminal angioplasty	Effective for restoring venous outflow in patients with short-segment hepatic vein stenosis.	[56]
Stenting	Implemented when angioplasty alone is insufficient to maintain venous patency.	[56]
Transjugular Intrahepatic Portosystemic Shunt (TIPS)	Reduces portal pressure and improves survival in patients who fail medical or interventional therapy or with diffuse hepatic vein involvement.	[57]
Liver transplantation	Considered in patients with end-stage liver disease or failure of other interventions.	[58]
Guideline-based Early Intervention	APASL recommends early intervention based on clinical severity and anatomical considerations rather than expectant management. Emphasises a multidisciplinary approach to optimise outcomes.	[59]

[Table/Fig-5]: Stepwise management of BCS.

Cost-effectiveness of therapeutic strategies for BCS varies usually across healthcare systems, resource settings [8,9]. Anticoagulation represents most cost-efficient first-line intervention due to its wide availability as well as relatively low expense [54]. Endovascular procedures inclusive of angioplasty, TIPS involve higher initial procedural costs; however, early intervention can further reduce long-term expenditures by prevention of hepatic decompensation, recurrent hospitalisations as well as need for transplantation [37,56]. Liver transplantation is known as the most resource-intensive modality having substantial perioperative costs and lifelong immunosuppressive therapy [58]. In low and middle-income countries, financial as well as infrastructural limitations can further delay escalation beyond anticoagulation thus potentially affecting outcomes [9,10]. Despite all these implications, formal cost-effectiveness analyses specific to BCS are scarce thus, highlighting about an important area for future health-economic research [10].

Management of BCS follows a stepwise strategy which is based upon clinical response, disease severity [27]. Anticoagulation is initiated promptly in all patients without any contraindications which are continued long term [33,54]. In those patients having short-segment stenosis, web-like obstruction, angioplasty with or without stenting can thereby restore hepatic venous outflow as well as prevent progression [33]. Patients who fail medical therapy, develop persistent portal hypertension, refractory ascites and hepatic dysfunction are mainly candidates for TIPS which has helped to significantly improve transplant-free survival in modern cohorts [37]. Liver transplantation thereby remains definitive therapy for patients having fulminant presentation, progressive liver failure despite usage of TIPS [58]. Importantly, early identification of non responders as well as timely escalation are critical as delayed intervention is associated with poorer outcomes [27]. Thus, management needs dynamic reassessment rather than a static treatment approach [27].

Budd-Chiari Syndrome in Pregnancy

Pregnancy represents a very unique as well as high-risk scenario in context of BCS due to physiological hypercoagulable state inherent to gestation which increases susceptibility to venous thrombotic events inclusive of BCS [60]. Pregnancy-related BCS can arise either as a first presentation during pregnancy or puerperium or be encountered in women having pre-existing BCS planning conception as well as has been identified in a notable proportion of BCS patients in pooled analyses of observational studies [60,61]. The prevalence of BCS which is pregnancy-associated ranges widely among cohorts having a pooled prevalence of approximately 6.8% among all BCS patients thereby underscoring significance of pregnancy as a contributory risk factor for hepatic venous outflow obstruction in susceptible women [61]. Diagnosis during pregnancy is usually challenging due to presence of clinical manifestations inclusive of abdominal pain, ascites as well as hepatomegaly can overlap with other pregnancy-related liver disorders (e.g., pre-eclampsia or acute fatty liver of pregnancy) [60]. Doppler ultrasonography remains main aspect of imaging which is done during pregnancy due to its safety profile while cross-sectional imaging (CT/MRI) can be further considered when necessary for detailed vascular mapping [60]. Early, accurate recognition is said to be very essential to differentiate BCS from other aetiologies, also guide management [60].

Management in pregnant patients prioritises anticoagulation with Low-Molecular-Weight Heparin (LMWH) for prevention of thrombus extension as well as recurrent thrombosis given contraindications, teratogenic risks associated along with vitamin K antagonists during gestation [60,62]. Interventional procedures which is inclusive of hepatic vein/IVC angioplasty, stenting or TIPS are usually reserved for refractory cases or undertaken after delivery for minimisation of foetal radiation exposure [60]. Maternal outcomes have also improved along with modern multidisciplinary care. In studies of women having known BCS who conceived, no maternal deaths were further observed as well as term live births were frequent along with live birth rates above 75% in several cohorts, particularly among women who received anticoagulation and close monitoring [60,61]. However, pregnancy loss, preterm delivery, intrauterine growth restriction as well as small-for-gestational-age infants remain more common than in general obstetric population thereby reflecting ongoing challenges in fetal outcomes [62].

Paediatric BCS

BCS in children is rare condition but it also represents a distinct clinical entity having unique etiological patterns, diagnostic challenges and management considerations as compared to adults [63]. Paediatric BCS accounts for a small proportion of hepatic vascular disorders as well as available data are largely derived from single-centre case series along with registry analyses [63,64]. Children more usually show inherited thrombophilias which is inclusive of protein C, protein S, or antithrombin deficiencies, congenital vascular anomalies also idiopathic causes [63].

Paediatric BCS further presents insidiously along with progressive type of ascites, hepatomegaly, abdominal distension, failure to thrive, signs of portal hypertension [36]. Acute presentations having fulminant hepatic failure are less usual but they have been described [36]. The symptoms can mimic chronic liver disease of other etiologies thus delayed diagnosis is common [36]. Doppler ultrasonography thus remains first-line diagnostic modality although smaller vessel caliber as well as technical limitations in children can further reduce sensitivity, thus necessitating cross-sectional imaging with MRI for confirmation [36,64].

Management principles in pediatric BCS cases still mirror adult stepwise strategies but it also needs few paediatric-specific considerations [36]. Lifelong anticoagulation is usually recommended when an underlying prothrombotic disorder is identified; low-molecular-weight heparin is commonly initiated which is followed by transition to vitamin K antagonists although experience with direct oral anticoagulants in children remains limited as well as it lacks robust safety data [65]. Endovascular interventions which are inclusive of angioplasty, TIPS can be technically challenging because of small vascular anatomy but have showed feasibility, improved survival in selected cases [65,66]. Liver transplantation is also indicated in children having decompensated cirrhosis, failure of interventional therapy and has been associated having acceptable survival rates comparable to other pediatric transplant indications [66].

Prognostically, early diagnosis as well as timely intervention are critical as untreated paediatric BCS carries significant risk of having progressive fibrosis, portal hypertension also growth impairment [36]. Future multicentre registries are very important for better defining optimal anticoagulation duration, interventional timing as well as long-term transplant outcomes in paediatric BCS population [36,66].

Future Directions and Research Priorities

Despite of various therapeutic advances there are still several important gaps remain in management of BCS [55]. Large prospective, population-based studies are thereby needed for better defining global incidence, regional etiologic variation as well as long-term outcomes as most current data is derived from retrospective tertiary-centre cohorts [55]. Randomised controlled trials which compare vitamin K antagonists with direct oral anticoagulants are lacking thus further leaving uncertainty regarding optimal long-term anticoagulation strategies [67]. Further validation of prognostic scoring systems across various type of BCS populations is required for improving stratification of risk as well as guide into early therapeutic escalation [62,66]. Additionally, molecular characterisation of underlying myeloproliferative neoplasms, other prothrombotic disorders can help into refining personalised treatment approaches in patients having BCS [10,68]. Finally, collaborative international registries, standardised diagnostic criteria are very essential for harmonisation of data collection, reduce epidemiologic uncertainty also inform evidence-based clinical guidelines [68].

Limitation(s)

The present narrative review is limited by potential selection bias inherent to non systematic literature synthesis and heterogeneity among included studies.

CONCLUSION(S)

Budd-Chiari Syndrome (BCS) is rare life-threatening hepatic vascular disorder, characterised by hepatic venous outflow obstruction leading hepatic congestion, portal hypertension, and fluctuating signs and symptoms. Multimodal imaging and clinical suspicion are

important to get the most desirable outcome. The progress in the knowledge of genetic predisposition and molecular mechanisms streamlined the risk stratification. Treatment should involve a multidisciplinary, stepwise approach to the management of the condition, including anticoagulation, endovascular interventions to TIPS or liver transplantation in severe instances. Early detection and timely treatment at an individual level are also important because they enhance the survival of people with BCS.

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