



Budd-Chiari syndrome: Prognostic scores, special populations, and management challenges

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Abstract

This editorial narrative review discussed Budd-Chiari syndrome (BCS), which represents a rare but critical vascular liver disease resulting in an obstruction of hepatic venous outflow. Despite having a unifying mechanism, the syndrome shows a large heterogeneity across presentation, cause, and disease trajectory, complicating diagnosis and management. Based on established prognostic scoring systems, the New Clichy Score, the BCS-transjugular intrahepatic portosystemic shunt Index, the Zeitoun Score, and the Pediatric End-stage Liver Disease score were examined. These scoring systems are used for risk stratification and therapeutic decision-making. Although these models deliver suitability information, their static parameters, narrow validation, and limited generalizability reduce their usefulness in diverse populations. Specific challenges are highlighted in pediatric patients, pregnant females, and individuals with myeloproliferative neoplasms for whom current tools often fall short. Moreover, there remains uncertainty regarding the durability of Pediatric End-stage Liver Disease score response and longer-term risks, such as hepatocellular carcinoma. There is a need to have a dynamic prognostic model that uses imaging and genetic factors in future studies. The article discussed enhancing recruitment to improve research. Overall, this article provided a contemporary, evidence-based approach for clinicians to aid in the evaluation and treatment of BCS.

Key Words: Budd-Chiari syndrome; Prognostic scoring system; Pediatric liver disease; Myeloproliferative neoplasms; Hepatocellular carcinoma; Transjugular intrahepatic portosystemic shunt; Advanced imaging techniques; Dynamic prognostic models

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Core Tip: This narrative review provided a comprehensive update on Budd-Chiari syndrome and discussed the advantages and disadvantages of current prognostic scores. It also emphasized special conditions in children, pregnancy, and cases related to myeloproliferative neoplasms. This paper identified key research gaps and advocated for future work on personalized risk assessment, long-term outcome evaluation, and standardized response criteria to improve clinical decision-making in this heterogeneous disease.

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INTRODUCTION

Budd-Chiari syndrome (BCS) is a rare but potentially life-threatening disorder of the liver that results in obstruction of the hepatic venous outflow. This obstruction is not secondary to right heart failure, pericardial disease, or sinusoidal obstruction syndrome (previously known as hepatic veno-occlusive disease). Obstruction can be either thrombotic or non-thrombotic. The obstruction causes hepatic congestion, portal hypertension, and progressive liver dysfunction[1]. BCS may present clinically with a wide spectrum from asymptomatic incidental discovery to fulminant hepatic failure. If it is not treated, it may lead to ascites, hepatomegaly, cirrhosis, and hepatocellular carcinoma (HCC). Early diagnosis and personalized teamwork must be conducted to improve survival and the cure-end outcomes[1].

The occurrence of BCS is different across countries because of different risk factors, access to health care, and diagnostic ability. In adults in Europe the yearly incidence has remained level at between 0.35 and 0.8 cases per million inhabitants (pmi)/year[2-4]. On the other hand incident rates in Asia are more heterogeneous; Japan had an estimated incidence of 0.13 pmi/year in 1989 while Kathmandu, Nepal had a higher incidence of 2.50 pmi/year[5,6].

BCS is broadly classified into primary and secondary forms. The primary form of BCS happens when hepatic veins or the inferior vena cava is obstructed due to intrinsic thrombus and often occurs with other prothrombotic conditions. Some examples are myeloproliferative neoplasms (MPNs), especially polycythemia vera, essential thrombocythemia, paroxysmal nocturnal hemoglobinuria, inherited thrombophilias (*e.g.*, deficiency of protein C or S or antithrombin III), the factor V Leiden mutation, and prothrombin G20210A mutation. Disease pathogenesis is aggravated by acquired risk factors like pregnancy, use of oral contraceptives, and antiphospholipid syndrome[7-9]. Secondary BCS occurs from external compression or invasion of the hepatic venous outflow tract, most notably by malignancies such as HCC, renal cell carcinoma, and adrenal tumors or metastatic disease[10,11].

Imaging modalities while becoming more advanced and easier to obtain are often not adequate for the diagnosis or treatment of BCS. Doppler ultrasound, contrast-enhanced CT, and magnetic resonance imaging (MRI) venography can all help. Management options vary from medical therapy with life-long anticoagulation to more invasive methods like angioplasty, transjugular intrahepatic portosystemic shunt (TIPS), and orthotopic liver transplantation (OLT). Yet making treatment decisions is not easy in view of the heterogeneous disease presentation. Currently, there are prognostic scoring systems like the Rotterdam, Clichy, and BCS-TIPS scores that help guide doctors. However, these may not be useful for everyone, especially kids and those getting TIPS.

In recent years the use of positron emission tomography (PET)/CT has been popularized as an adjunctive imaging modality to assess secondary causes of BCS. This is especially useful when conventional imaging is inconclusive. Although Doppler ultrasound, CT, and MRI remain the mainstay for diagnosis of BCS, PET/CT with its functional metabolic imaging may help detect occult malignancies, inflammatory vasculopathies, or systemic neoplastic processes contributing to obstruction of hepatic venous outflow[12].

Clinical evidence from numerous studies and case reports has pointed to the usefulness of PET/CT for identifying hypermetabolic thrombi, tumors with infiltration of venous walls, paraneoplastic thrombosis, lymphoproliferative disorder or intravascular lymphoma, all of which may present similar symptoms to BCS. PET/CT has also been found to be useful for biopsy site selection, systemic disease burden assessment, and staging and treatment planning for malignant BCS. Using PET/CT in the diagnostic algorithm seems warranted, especially in patients with atypical presentations, those who have poor responses to anticoagulation, and in instances with systemic illness of unknown primary cause. PET/CT effectively links morphological findings to pathophysiological processes by combining metabolic and anatomical imaging, leading to improved diagnostic confidence and tailored therapeutic strategies[13,14].

The purpose of this review was to provide a comprehensive and critical overview of current prognostic models used in BCS with an emphasis on their clinical usefulness and limitations in various patient populations. Moreover, it provided a stepwise, evidence-based management algorithm, discussed key predictive factors of a poor prognosis, and explored urgent research priorities, most notably the need for validated pediatric prognostic tools and standardized criteria for long-term TIPS patency assessment.

PROGNOSTIC SCORING SYSTEMS IN BCS

The prognosis of BCS can vary greatly from asymptomatic to fulminant hepatic failure. The outcome relies mainly on how significantly the blood from the liver is obstructed, whether any underlying conditions are present, and when treatment is started. A 3-year mortality rate without treatment approaches 90%. Nevertheless, progress in medications, blood vessel procedures, and liver transplantation (LT) has greatly enhanced this result. Reliable forecasting instruments are necessary to regulate therapy strategies and anticipate clinical pathways.

The types of data on which they are based can divide the prognostic systems used in BCS. Several validated scores or algorithms, including the New Clichy Score, BCS-TIPS Prognostic Index, Zeitoun Score, and Pediatric End-stage Liver Disease (PELD) Score rely primarily on clinical and laboratory characteristics [*e.g.*, severity of ascites, international normalized ratio (INR), serum bilirubin, renal function, age] (Table 1). Imaging-based variables are still undervalued in popular models. Certain studies have previously examined whether Doppler ultrasound and CT findings (*i.e.* hepatic vein patency and caudate lobe hypertrophy) have prognostic value; however, none have been formally integrated into a scoring system with validation. This distinction matters a lot when studying how reliable or comprehensive current models are[15].

Scoring systems proposed for BCS predict disease severity as well as clinical outcomes to help with management. Doctors use clinical, laboratory, and imaging data to estimate prognosis using scoring models. This means that studies may not apply to different types of people or people in a different area.

The New Clichy Prognostic score

The New Clichy Score is a validated prognostic model that predicts overall survival in patients with BCS[15]. It has five variables representing functions of the liver and the kidneys and disease severity as following: Ascites grade (0-3) reflects the severity of fluid accumulation in the peritoneal cavity; Child-Pugh Score (5-15 points), a composite measure of liver dysfunction, including bilirubin, albumin, INR, ascites, and encephalopathy; patient's age at time of diagnosis; serum creatinine (mg/dL), a surrogate for renal function; and Form III (acute-on-chronic presentation) is allocated a value of one if present and zero if absent.

The equation that includes these variables is $\text{Child-Pugh} + (0.047 \times \text{age}) + (0.0045 \times \text{creatinine}) + (2.2 \times \text{Form III})$. A threshold score of ≥ 3.75 has been linked to poorer prognosis and overall survival[15]. The New Clichy Score was first created and validated in an Egyptian cohort. It is clinically useful for the identification of patients who are high risk and may benefit from an early referral for LT or more aggressive therapy[16]. Even so the external validity of the findings is limited, and further multicenter studies are required to evaluate its generalizability.

Clinical relevance and limitations: Despite its usefulness the New Clichy Score should be interpreted in the clinical context. Scores predicting prognosis in BCS are inherently flawed due to the presentation of the disease ranging from fulminant hepatic failure to chronic compensated states. In addition anticoagulation, TIPS, or LT will alter the course of the illness. In such cases the prediction may be inaccurate if calculated with static scores. Recently, individuals have been working on dynamic prognostic models that consider therapy response and changing clinical parameters to help provide more personalized and timely prognostic assessments. In medical practice the use of scoring systems along with clinical experience is key to optimal management of BCS[16,17].

BCS-TIPS Prognostic Index

The BCS-TIPS Prognostic Index score was developed with the aim of predicting the 1-year survival of patients with BCS following placement of a TIPS. This tool is useful for identifying those who are more likely to benefit from TIPS. It can also be used to help identify patients at high risk who require expedited listing for LT[17].

The index contains three clinical variables, including age (years), serum bilirubin (mg/dL), and INR. These parameters are combined into the following formula: $(\text{age} \times 0.08) + (\text{bilirubin} \times 0.16) + (\text{INR} \times 0.63)$.

Based on this calculation, a score under 7 is related to excellent 1-year survival approaching 96%. Therefore, TIPS is very successful in patients with low model for end-stage liver disease (MELD) scores. When the score is more than 7, it indicates high risk, showing poorer outcomes in 1 year. In addition it may indicate a need for aggressive or alternate intervention, including LT[17].

The BCS-TIPS Index is useful for stratifying patients by risk based on survival following TIPS. It helps make clinical decisions including timing of TIPS and LT. It is important to bear in mind that the index predicts post-TIPS survival only and does not predict overall BCS prognosis, especially in conservatively managed patients or with anticoagulation alone. Moreover, the BCS-TIPS Index may not be generalizable. The index was first confirmed in a European population, but different results were demonstrated in non-European populations from Asia and Middle Eastern countries[16,17]. Notably, the score is not related to shunt patency at 1 year, which is an important contributor to long-term clinical success in patients undergoing TIPS. In general the BCS-TIPS Index is a useful prognostic tool in a defined clinical context, but it should not replace clinical judgment and assessment of the individual patient.

Table 1 Variables, formulas, and interpretation for each score

Score	Type	Parameters used	Formula/cutoffs	Clinical interpretation
New Clichy Score	Clinical/Laboratory-based	Bilirubin, INR, creatinine, ascites, encephalopathy	Weighted scoring system; score > 2 indicates poor prognosis	Stratifies diseases severity; guides timing of intervention
BCS-TIPS Index	Clinical/Laboratory-based	Bilirubin, INR, creatinine, age	Score = $(1.03 \times \text{creatinine}) + (0.8 \times \text{bilirubin}) + (0.8 \times \text{INR}) + (0.3 \times \text{age})$; > 7 Leads to poor outcome	Predicts survival after TIPS
Zeitoun Score	Clinical/Laboratory-based	INR, bilirubin, ascites	Categorical system: Class I (mild) to Class III (severe)	Assesses prognosis pre-TIPS
PELD Score	Clinical/Laboratory-based (pediatrics)	Albumin, bilirubin, INR, growth failure, age	Standard UNOS formula; higher scores correlate with worse outcomes	Used for pediatric LT prioritization
Imaging parameters	Imaging-based (proposed/research)	HV patency, caudate lobe hypertrophy, hepatic congestion signs	Standard UNOS formula; higher scores correlate with worse outcomes	Not part of current scores; proposed for future integration

BCS: Budd-Chiari syndrome; TIPS: Transjugular intrahepatic portosystemic shunt; PELD Score: Pediatric End-stage Liver Disease Score; INR: International normalized ratio; HV: Hepatic vein; LT: Liver transplantation; UNOS: United Network. for Organ Sharing.

Zeitoun prognostic score

The Zeitoun Score is a prognostic model for BCS that estimates overall survival and strategies risk for therapeutic decision-making. The score incorporates four key clinical variables[18]: (1) Age (years). Reflects baseline physiological reserve; (2) Response of ascites levels for diuretics (graded 0-2). Grade zero reflects no ascites, grade one means ascites under control with diuretics while grade two means refractory ascites; (3) Child-Pugh score. Assesses the synthetic function of the liver and the degree of portal hypertension; and (4) Serum creatinine (mg/dL). It is a marker of renal impairment, which is a critical prognostic marker in advanced liver disease. These parameters are integrated into the following formula: $(0.95 \times \text{Child-Pugh}) + (0.35 \times \text{age}) + (1.35 \times \text{creatinine}) + (0.9 \times \text{ascites response grade})$. A cutoff value of 5.4 has been determined. Scores ≤ 5.4 indicate a good prognosis and may respond to conservative treatment (*e.g.*, anticoagulation and diuretics). A score over 5.4 indicates a poor prognosis, which suggests the need for an invasive procedure such as TIPS or OLT[18].

The Zeitoun Score was developed in a cohort of patients with BCS with different managements. The impact of each management and specifically shunt procedures was not clearly specified in the original work. Even though this model can be useful for estimating overall survival, it does not have dynamic reassessment. Further, this model may not reflect treatment response over time. Further proof is required for it to apply to more patient groups.

PELD Score

The PELD Score is a prognostic index widely used for prioritizing children under 12 years of age for LT based on estimated 3-month waitlist mortality. It is used for children with chronic liver diseases, including rare causes like pediatric BCS, and informs organ allocation policy under United Network for Organ Sharing (also known as UNOS) and other international frameworks. The score includes the following five variables: Total bilirubin (mg/dL) reflects cholestasis and hepatic excretory function; INR analysis evaluates hepatic synthetic dysfunction; serum albumin (g/dL) is a key indicator of nutritional status as well as liver function; if the child was added to the list before the age of 1 year, the age benefit extending until 24 months would be applicable; and growth failure (if the weight or height is more than 2 standard deviations below the mean, you will get a score of 0.667 points).

The equation for PELD is expressed as $\text{PELD} = 0.480 \times \ln(\text{bilirubin}) + 1.857 \times \ln(\text{INR}) - 0.687 \times \ln(\text{albumin}) + 0.436$ (if < 1 year) + 0.667 (if growth failure present) (Note: The logarithmic input is limited to a minimum value of one to prevent negative scores). A PELD Score ≥ 15 indicates a high risk of death within 3 months and supports urgent LT listing. A study of biliary atresia in children showed mortality of 47.9% for PELD ≥ 15 and 31.6% for PELD < 15[19,20].

The PELD Score does not consider some complications that warrant exception points in the transplant allocation systems. These complications include hepatopulmonary syndrome or renal failure. Moreover, it is not routinely used for prognostication in BCS unless there is consideration for transplant due to the underlying hepatic decompensation in a child with BCS. It can predict the risk of dying before transplant but not how someone will do after transplant or overall as they live with BCS.

Limitations of current scoring systems

Even though prognostic scoring systems help predict the outcome of BCS and stratify cohorts, they fail to be clinically used for several important reasons.

Limited individual prognostic accuracy, as most established scores, including the New Clichy Score, Zeitoun Index, and BCS-TIPS Score, demonstrate modest predictive performance with areas under the receiver operating characteristic curve often below 0.70 for orthotopic LT (OLT)-free survival. The value is much lower than the area under the receiver operating characteristic curve threshold of 0.80-0.90 that is considered necessary for accurate individual clinical prognosis [21]. The scores also have low reliability as predictive tools. For example, differences across the target populations limit the external validity of the scoring systems. The variability in genetic background and the causes of the disease (such as

MPNs *vs* thrombophilia) and treatment protocols as well as the health care infrastructure in the different regions hampers the generalizability of results. Although the New Clichy Score was validated in an Egyptian cohort, it may not yield similar results in Europeans or Asians. Study findings have also failed to make a prediction on 1-year TIPS patency using the BCS-TIPS score. Most often a difference in baseline disease severity and clinical management strategies may be the reason for this[16,21,22].

Most BCS prognostic models are based on static baseline data and do not incorporate changes in liver function or renal status or responses to therapeutic efforts (anticoagulation, TIPS, LT). As such, their use in serial clinical decision-making is limited, especially in a disease that can evolve acutely or chronically[21]. Moreover, there is no integration of imaging data or biomarkers as the new biomarkers such as von Willebrand factor, D-dimer, and inflammatory cytokines and radiological findings such as Doppler flow characteristics and hepatic vein obstruction are not assessed in any of the current scoring systems although they might have some additive prognostic value. Techniques like contrast-enhanced ultrasound or MRI show promise for evaluating hepatic hemodynamics, but they are not yet included in current indices. In addition the thresholds for therapeutic decisions are absent. Currently, prognostic scores are not employed to enhance care escalation. For example, these systems lack validated thresholds for the determination of medical management to TIPS and LT referral. According to some experts, clinical decisions continue to be mostly based on subjective assessment of persistent symptoms like refractory ascites and progressive hepatic encephalopathy or laboratory deterioration rather than score driven protocols[21].

Many prognostic tools come from older datasets. The PELD Score is used to prioritize LT in children under the age of 12. This score does not predict BCS-specific outcomes and has not been validated in pediatric BCS cohorts. Additionally, while the Zeitoun Score includes pediatric patients, its external validation in children with different etiologies is lacking [22]. The limitations on lab parameters and anticoagulation impact is a problem as certain score components, particularly INR, may be less reliable in BCS. The increased blood clot tendency and common use of blood thinners in BCS can artificially increase INR values, wrongly assuming liver dysfunction. Patients with cirrhosis with low muscle mass and altered tubular handling may not have their renal function reflected accurately by creatinine[21-23]. Eventually, poor measurement of functional status and quality of life are other limitations because these models ignore patient-reported outcomes and functional capacity, two important prognostic indicators that are recognized, especially for patients suffering from chronic liver disease. Examining health-related quality of life and frailty status may help in patient-centered decision-making[22].

A summary of the limitations of major BCS scores is presented in Table 2.

BCS IN SPECIAL POPULATIONS

BCS although uncommon can affect different populations, each with differing pathophysiological considerations that affect diagnosis and management. BCS delivers unique challenges in diagnosis and management in special populations. We can optimize our outcomes by identifying and treating hidden prothrombotic states, managing pregnancy-associated risk, individualizing therapy in children, and managing malignancy-associated presentations. The key to success in treating these patients remains a personalized multidisciplinary approach.

Hypercoagulable states and thrombophilia

The most commonly identified cause of BCS is hypercoagulability. At least one prothrombotic factor is observed in 75%-80% of patients. Many others have two or more simultaneous abnormalities. Inherited or acquired, these factors are usually synergistic in predisposing to hepatic vein thrombosis[24].

Inherited thrombophilic disorders like Factor V Leiden mutation, which is one of the most common mutations, linked with BCS, are found in 25%-31% of the cases[25,26]. This leads to activated protein C resistance, which increases the risk of thrombosis. Prothrombin G20210A gene mutation is less often associated with BCS but affects plasma prothrombin levels. Deficiencies of protein C, protein S, and anti-thrombin III are inherited conditions associated with BCS. However, the involvement of concurrent hepatic dysfunction complicates the interpretation of the levels, which necessitates repeat testing in stable phases. MPNs are the most common causes of the BCS. The JAK2V617F mutation is present in up to 41% of patients with BCS, usually in the absence of gross hematological abnormalities[27]. To reach a definitive diagnosis, a bone marrow biopsy may be needed. In addition sometimes red blood cells burst at night in a sleep-related rare blood version paroxysmal nocturnal hemoglobinuria occurs. Diagnosis requires detection of CD55/CD59-deficient cells by flow cytometry. The involvement of autoimmune-mediated hypercoagulability requires patients with antiphospholipid syndrome to undergo repeated testing of lupus anticoagulant, anticardiolipin antibodies, and anti-beta2-glycoprotein I antibodies for confirmation. It is important for clinicians to recognize that many patients can have multiple thrombophilic conditions and require a full hematological workup. A trial of anticoagulation is the mainstay of management, but the management may have to be individualized based on the etiology, bleeding risk factors, *etc.*[24-27].

Pregnancy and hormonal factors

Pregnancy refers to a physiological state of excessive blood clotting (hypercoagulability) due to high levels of certain clotting factors (fibrinogen, factor VIII), less breakdown of these clots, and low levels of certain natural anticlotting proteins (for example, protein S). Moreover, the weight of the uterus during pregnancy suppresses blood flow[28]. Pregnancy may reveal or worsen BCS in females with genetic disorders predisposing to blood clots. If the condition is not handled quickly, it may cause serious harm to the fetus. Treatment of the disorder is usually with anticoagulation. Low molecular weight heparin is preferred for this as it is safer in pregnancy. Warfarin is not recommended in the first

Table 2 Comparison and limitations of prognostic scoring systems in Budd-Chiari syndrome

Feature	New Clichy Score	BCS-TIPS Index	Zeitoun Score	PELD Score
Population	Primarily validated in adult patients with BCS	Specifically designed for adult patients with BCS who have undergone a TIPS procedure	Developed for adult patients with BCS	Designed for pediatric patients (< 12 years old) with end-stage liver disease awaiting LT
Predictors	Ascites score, Child-Pugh score, age, serum creatinine, clinicopathological form III (acute on chronic)	Age, serum total bilirubin, INR	Age, response of ascites to diuretics, Child-Pugh score, serum creatinine	Age (< 1 year), albumin, bilirubin (total), INR, growth failure
Cutoff	≥ 3.75 for predicting 1-year outcomes	< 7 indicates excellent 1-year survival (96%); ≥ 7 indicates high-risk	≤ 5.4 indicates good prognosis; > 5.4 indicates poor prognosis	≥ 15 indicates high risk of death without LT in chronic liver disease
Limitations	Primarily validated in a specific Egyptian cohort; generalizability needs confirmation. Overall survival prediction	Predicts survival specifically post-TIPS but not overall BCS survival. Generalizability may vary; did not predict 1-year shunt patency in one study	Focused on overall survival; impact of specific interventions like shunts uncertain in the original study	Primarily predicts pre transplant mortality for children < 12 years. Accuracy influenced by unmeasured factors. Not typically used for BCS prognosis

Comparison and limitations of prognostic scoring systems in Budd-Chiari syndrome adapted from[12-17]. BCS: Budd-Chiari syndrome; TIPS: Transjugular intrahepatic portosystemic shunt; PELD Score: Pediatric End-stage Liver Disease Score; INR: International normalized ratio; LT: Liver transplantation.

trimester and near term because it may produce fetal deformities and cause bleeding. Hepatologist, hematologist, and maternal-fetal medicine specialist consulting is essential. Females with varicose veins or clotting problems may need a C-section[29].

Oral contraceptives may increase thrombotic risk by increasing the synthesis of procoagulant factors and reducing antithrombin activity in females. Studies indicate that using this product increases your chances of developing hepatic vein thrombosis by 2.3 times higher than the average person, particularly in the first year or two of using it. In contraceptive-using females with thrombotic events of unknown origin, screening for thrombophilia is required[30,31].

BCS in the pediatric population

BCS in children is exceptionally rare but often presents with more severe clinical manifestations due to delayed diagnosis. In pediatric cases the cause is different than adults, and inherited thrombophilic conditions are prevalent. Each year the incidence varies geographically between 0.2-4.1 million. Pediatric BCS is more common in parts of Asia. The main causes is inherited thrombophilias such as factor V Leiden and protein C deficiency and congenital vascular anomalies may contribute[32]. The infant or child may exhibit hepatosplenomegaly with failure to thrive or vague gastrointestinal symptoms. Older children are similar to adult presentations with ascites and abdominal pain. Early diagnosis *via* Doppler ultrasound or MRI is critical. The first line therapy is anticoagulation with TIPS or LT used in refractory or advanced cases. Eventually, early treatment with early and aggressive intervention lead to a good prognosis[32,33].

Malignancy-associated BCS

BCS can be caused by malignancies through direct obstruction or thrust, mechanical obstruction, or tumor-induced hypercoagulability. There are many events that can induce malignancy-associated BCS, like invasion or compression in HCC, renal cell carcinoma, and adrenal tumors that can directly invade or compress hepatic veins or the inferior vena cava. Furthermore, cancer causes a state that triggers blood clots or keeps blood clots in control. Tumor thrombi may also contribute to venous obstruction. Eventually, iatrogenic factors including chemotherapy, central venous catheters, and hematological malignancy increase the risk of thrombosis. Treatment usually involves two focuses: Anticoagulation to treat thrombotic components and oncological therapies to manage tumor burden. In some cases endovascular interventions (*e.g.*, stenting) and/or surgical resection is needed to relieve compression. The prognosis depends on the tumor biology and the hepatic reserve. Selected patients for LT are those with localized HCC and good liver function[34].

RISK STRATIFICATION AND MANAGEMENT OF BCS

The effective management of BCS requires a personalized risk stratification together with a dynamic and stepwise approach. Clinical heterogeneity and rarity of BCS lead to limited randomized controlled trials. Therefore, the majority of the current treatment framework is guided by expert opinion and observational data. As a result clinical judgment remains at the heart of therapy choice and treatment, especially when patient responses deviate from typical patterns or when comorbidities complicate matters[21].

Risk stratification

The process of determining the risk of BCS requires evaluating various clinical characteristics, biochemical parameters, and imaging. Different prognostic indices like the New Clichy, BCS-TIPS, Zeitoun, and PELD Scores may provide some

assistance, but they are not validated for individualized practice. The restricted discriminative power of these limitations and the absence of dynamic reactivity warrant a more thorough clinical evaluation[21,34].

Key clinical factors that can influence the prognosis are patient age, presence and severity of ascites, level of hepatic encephalopathy, renal dysfunction, and evidence of chronic liver disease. Biochemical markers like serum bilirubin, creatinine, albumin, and INR are the mainstay for evaluating hepatic synthetic function and coagulopathy. However, it must be noted that INR can be confounded by anticoagulation and may not always reliably indicate liver function[20]. Imaging techniques like Doppler ultrasonography, CT, and MR venography play an integral role in assessing the extent of hepatic venous outflow obstruction, collateral formation, and evidence of portal hypertension or hepatic congestion. In children prognosis is further complicated by different pathophysiology and development of the disease. Pediatric-specific tools have emerged such as the PELD Score and Zeitoun index. These tools have the potential use in stratifying risk and guiding early intervention. However, further validation in large, multicenter cohorts is required[22].

Management strategy

The modern treatment approach for BCS follows a stepwise and response-adapted strategy. It refers to the escalation of therapy based on clinical deterioration or failure to achieve therapeutic targets. The progressive nature of liver diseases and the need to intervene before irreversible hepatic decompensation is represented in this hierarchy. All management would initially include anticoagulation therapy, which is the mainstay of treatment. Prompt initiation of full-dose anticoagulation, usually involving low molecular weight heparin followed by vitamin K antagonists or direct oral anticoagulants is recommended at diagnosis. It is necessary to avoid the spread of a thrombus and assist the spontaneous recanalization in select cases. In most patients anticoagulation is continued on a long-term basis because of a high risk of recurrence that is often associated with prothrombotic disorders[21,35].

For those with constant symptoms who are suffering from complicating factors like worsening liver function and resistant ascites despite adequate anticoagulation, radiological treatment is warranted. Doctors might insert a stent in the hepatic vein in the case of focal hepatic vein stenosis. Moreover, this might enhance the condition of the health of the patient quite significantly. In patients with diffuse obstruction or failure of angioplasty, TIPS represents the preferred approach. TIPS helps the portal system by using the portal vein drainage. This creates communication with the systemic venous circulation, helping in reducing portal hypertension. Early TIPS procedure in selected patients has shown to improve transplant-free survival mainly in patients with high-risk features and for non-responders to medical therapy[21,36].

Orthotopic LT is reserved for patients with end-stage liver disease or hepatic failure or those with persisting disease who are refractory to prior interventions. An LT not only cures BCS but also any underlying chronic liver disease. LT outcomes are generally good, but patient selection is crucial. Long-term immunosuppressive therapy requires careful management. To treat BCS better a timely diagnosis, risk stratification, and personalized and stepwise treatment plan, commensurate with the severity of hepatic venous outflow obstruction and clinical course of the patient, is needed. Future directions should focus on the improved individual prediction of the risk of developing decompensation with prognostic tools, pediatric-specific algorithms, and prospective studies on the role of early TIPS in high-risk subgroups[21,37].

Therapy escalation and surveillance.

In BCS with a poor clinical situation or deeming the treatment ineffective, removal of the etiological factor is done categorically. Though the New Clichy, BCS-TIPS, and any other scores provide similar information, decisions depend on clinical judgement. Persistent symptoms like refractory ascites, encephalopathy, variceal bleeding, or an increase in jaundice necessitate moving towards advanced therapies such as TIPS or LT[21].

The long-term management of patients with BCS requires surveillance of the obstruction especially after apparent therapeutic success. Early recognition of any disease recurrence, shunt dysfunction, and evolution of complications, including HCC, is possible through regular monitoring. This is an acknowledged late sequelae, especially in those with underlying chronic liver disease or cirrhosis at diagnosis. Imaging techniques like dopamine ultrasound and contrast MRI or CT should be done at regular intervals to see the hepatic vasculature, TIPS patency, and parenchymal changes. Moreover, laboratory surveillance including liver function tests and serum alpha-fetoprotein tests may be useful for detecting hepatocarcinogenesis[22].

BCS has a heterogeneous long-term prognosis that is largely dependent on underlying cause, treatment response, and adherence to surveillance. Due to this level of risk, management is individualized with tailored follow-up plans. Furthermore, a multidisciplinary approach is optimal. Finally, consideration may be given to LT candidacy if clinical stability cannot be maintained.

PERSISTENT RESEARCH GAPS IN BCS: SPECIAL POPULATIONS, PROCEDURAL OUTCOMES, AND STANDARDIZED METRICS

Even with improvements in BCS diagnosis and stepwise management, there are many important research gaps that remain unanswered, particularly those related to subtle nuances, population-specific matters, and procedural insight. There are limits stopping evidence-based clinical decision-making. There is an urgent need for prognostic indices that are specific to children. Existing tools limit predictive ability. Current tools such as the PELD Score were originally designed for chronic liver disease and demonstrate limited predictive performance in dynamic settings like pediatric acute liver failure in which timely LT may be lifesaving. Static parameters used in these scores do not adequately capture the quick

clinical deterioration common to pediatric acute liver failure. Validation of newer dynamic models is inconsistent[35]. Moreover, in chronic liver diseases, like in children with BCS and in cases of ACLF, existing adult-based tools, like MELD, are not useful. While alternatives like the pediatric Chronic Liver Failure-Sequential Organ Failure Assessment score that integrates multi-organ dysfunction feature potential, a strong pediatric validation method is not in existence. The absence of accurate prognostic tools that can be used at the bedside delay interventions and negatively affect the clinical outcomes of children[38,39].

There are limited data about the long-term outcome of TIPS in patients with portal hypertension with age-specific challenges like shunt patency and HCC. The data on long-term surveillance is limited despite expanded polytetrafluoroethylene-covered stents having improved short-term to medium-term patency[40]. Numerous patients require numerous interventions or have imaging followed up closely, indicating a need for predictive markers of shunt durability. In addition the association of TIPS with subsequent HCC development in patients with BCS is not well established. The literature on post-TIPS HCC risk comes from cirrhosis studies not BCS studies, limiting extrapolation[41]. The role of TIPS in patients with pre-existing HCC is intricate and debated. Although it might be possible for the pressure of the portal vein to decrease or to use it as a bridge to oncological therapy, one still worries about it causing the spread of the tumor or altering the behavior of the tumor. Some studies may not show an increased risk, but we can hardly determine anything or optimize patient selection due to the lack of dedicated prospective studies[40-42].

A third domain that requires urgent attention is the lack of standard treatment response criteria in BCS. In both the clinical trials and in routine practice, there is no standardization for evaluating success of treatment, whether anticoagulation, TIPS, or surgical shunts[43]. Most guidelines provide a stepwise therapeutic algorithm but do not define clear and validated endpoints to measure treatment effect. Research indicates that the disappearance of the ascites or peripheral edema can also be used as markers. However, such definitions are not comprehensive and vary in their adoption. The comparison of therapeutic modalities and multicenter trials in neuromuscular disease has been difficult. The reason is the absence of consensus-based definitions that incorporate clinical, biochemical, radiological, and patient-reported outcomes. The absence of standardized methods to measure outcomes hampers systematic review and meta-analysis endeavors but is also seen to limit their clinical transferability[44,45]. Ultimately, filling these research gaps, which include developing pediatric-specific prognostic scoring tools, long-term assessment of the outcomes of TIPS, including HCC risk, and universally accepted definitions of treatment response, will be essential for fine-tuning the care pathway in BCS. The areas of study outlined here should represent key targets for future collaborations and prospective studies that may ultimately improve patient stratification, monitoring, and individualized management in diverse populations[43].

FUTURE DIRECTIONS

Recent advancements in imaging diagnostics and minimally invasive therapies have reshaped the management of BCS. However, several unmet needs persist. Future research should aim for the design of adaptive models of real-time prognosis that lead to implementation but are not limited to clinical and biochemical signatures, radiological markers, genetic susceptibility, and inflammatory biomarkers. The use of artificial intelligence (AI) and machine learning (ML) algorithms can better individualize risk stratification, predict treatment response, and assist with escalation therapy than current scoring systems[46].

The early evaluation of hepatic vascular flow through Doppler methods, enhanced ultrasound, and flow quantification by MRI should be done to detect early hemodynamic changes to allow for timely intervention[47]. Similarly, advanced endovascular technologies like bioengineered covered stents, drug-eluting shunt systems, and image-guided navigational TIPS will greatly aid in improving procedural results and minimizing complications while also extending shunt patency. Parallel strategies to refine anticoagulation depending on thrombotic risk, bleeding propensity, and genotypes (*e.g.*, JAK2, factor V Leiden) will help in optimizing the safety and efficacy of the treatment[48-52]. Additionally, it is important to study treatments that enhance blood flow and endothelial capacity in future studies to prevent venous stasis and clotting. Ultimately, we do need better-standardized outcome definitions (clinical, biochemical, radiological, and patient-reported outcome measures) to harmonize data collection between studies to enhance comparability. It will be important to establish such metrics to allow robust prospective multicenter trials that could ultimately lead to evidence-based personalized treatment algorithms that improve long-term outcomes of this rare but complex vascular liver disorder[49].

AI and ML have the potential to transform the prediction and personalized treatment of BCS. Compared with traditional scores, which utilize only clinical and laboratory data, AI/ML models can leverage more complex and multidimensional data (*e.g.*, longitudinal labs, imaging features, genomic data) to uncover hidden signals and improve outcome prediction[51]. Techniques like supervised learning (random forests, boosting algorithms), which predict response to TIPS or risk of dying, or unsupervised clustering of patients into clinically relevant subgroups (*e.g.*, A FLIP, pregnancy-related BCS, MPN-related BCS) can help. Furthermore, it is postulated that radiomics and deep learning applied to CT, MRI or Doppler imaging could aid in noninvasive evaluation of hepatic vein patency and liver congestion (trials could help to establish or disprove this hypothesis, leading to dynamic tools monitoring). As the use of AI applications expand in cardiology and cardiovascular medicine, the natural language processing could extract structured data from clinical notes in real-time to support decisions. Although the application of AI/ML into BCS workflows is only at an early stage, it has the potential to help with improved risk stratification, treatment optimization, and prediction of long-term outcomes[51].

CONCLUSION

BCS is a rare but clinically relevant vascular liver disease that presents significant challenges for diagnosis and therapy. Although important progress has been made in explaining its pathophysiology and treatment options ranging from anticoagulation and endovascular treatments to TIPS and OLT, BCS still needs an individualized multidisciplinary approach. The heterogeneity of the disease, the variability of its presentation, and overlapping with systemic pro-thrombotic states hampers implementation of standardized management protocols. Special populations may complicate this considerably. These populations may include children, patients with malignancy-related BCS, and those with acute-on-chronic liver failure[1-5].

The existing prognostication models (*e.g.*, New Clichy Score, BCS-TIPS Index, MELD/PELD) are useful in risk stratification on a broad scale. However, they may not always offer precise or generalizable data. This is especially true in changing clinical circumstances and in the younger cohort. This brings to light an urgent necessity for improved prognostic tools that account for changing factors and clinical conditions[19,22].

Several critical research gaps persist. Absence of validated pediatric-specific scoring systems impedes identification of children that may have benefited from urgent interventions. The long-term effects of TIPS like stent patency and HCC risk have not been evaluated enough in BCS compared with other portal hypertension etiologies. Moreover, the lack of universally recognized, standard definitions of treatment response greatly limits the comparability of data across studies and clinical trials. We must close these knowledge gaps through rigorous multicenter prospective research to help advance our field. The use of standardized outcome measures, prognostic indices relevant to age, and long-term surveillance data will help refine management algorithms[44,45].

FOOTNOTES

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