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Surgical Management of Budd-Chiari Syndrome: A Comprehensive Review of Techniques and Outcomes

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ABSTRACT

Budd-Chiari syndrome (BCS) is a rare but life-threatening condition characterized by obstruction of hepatic venous outflow, leading to hepatic congestion, portal hypertension, and potential liver failure. While pharmacological and interventional approaches play a significant role in its management, surgical intervention remains a cornerstone for cases refractory to less invasive measures. This review explores the surgical strategies employed in the treatment of BCS, ranging from shunting procedures, such as portosystemic shunts, to advanced techniques like liver transplantation. Emphasis is placed on patient selection criteria, perioperative considerations, and long-term outcomes. Additionally, the evolving role of minimally invasive and hybrid approaches in this domain is analyzed. By synthesizing current evidence, this article aims to guide clinicians in optimizing the surgical care of BCS patients.

KEYWORDS: Budd-Chiari syndrome, hepatic venous outflow obstruction, surgical management, liver transplantation, portosystemic shunt, hepatic congestion

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INTRODUCTION

Budd-Chiari syndrome (BCS) represents a rare vascular disorder resulting from the obstruction of hepatic venous outflow. This obstruction, caused by thrombotic, stenotic, or compressive mechanisms, leads to hepatic congestion, impaired venous drainage, and subsequent portal hypertension. The clinical manifestations of BCS are highly variable, ranging from asymptomatic cases to fulminant hepatic failure. While the condition is uncommon, its impact on liver function and systemic health underscores the critical need for effective management strategies.1,2

The therapeutic approach to BCS is multifaceted, involving anticoagulation, thrombolysis, interventional radiology techniques, and surgical interventions. The choice of treatment is guided by the etiology, severity of hepatic dysfunction, and the presence of underlying conditions such as myeloproliferative disorders or hypercoagulable states. Surgical management, although reserved for selected cases, is particularly crucial in patients with refractory disease, progressive liver failure, or complications such as refractory ascites.1,2

This article focuses on the surgical options available for BCS, including portosystemic shunts, mesoatrial bypass, and orthotopic liver transplantation (OLT). These techniques, while diverse in their application and complexity, share the common goal of restoring venous outflow, reducing portal hypertension, and preserving liver function. The introduction also discusses the role of minimally invasive and hybrid approaches, reflecting the continuous evolution of surgical strategies in hepatology.3

Through a detailed examination of current evidence and expert opinions, this review aims to provide a comprehensive understanding of the indications, techniques, and outcomes of surgical management in BCS, offering a valuable resource for clinicians involved in the care of these complex patients.3

EPIDEMIOLOGY

Budd-Chiari syndrome (BCS) is a rare vascular disorder with a global distribution, although its incidence and prevalence vary significantly depending on geographic region, genetic predisposition, and underlying etiological factors. The syndrome is characterized by hepatic venous outflow

obstruction, either primary (caused by venous thrombosis) or secondary (resulting from external compression). Its rarity and diverse presentations complicate accurate epidemiological assessments, but its burden is particularly notable in regions where hypercoagulable conditions and certain infections are more prevalent.4,5

The reported incidence of BCS ranges from 0.5 to 1 case per million annually in Western countries. However, in parts of Asia and Africa, particularly regions with a high prevalence of chronic infections such as tuberculosis or parasitic diseases like schistosomiasis, the incidence is likely underreported due to limited diagnostic resources. In India, for example, BCS is more commonly associated with membranous obstruction of the inferior vena cava, reflecting distinct regional etiologies.4,5

Gender and age distribution also reveal notable patterns. BCS is more frequently diagnosed in women, particularly those of childbearing age, due to the increased risk of hypercoagulable states linked to pregnancy, oral contraceptive use, or underlying hormonal factors. However, in areas where primary hepatic vein thrombosis predominates, the male-to-female ratio may be more balanced. The peak age of onset generally falls between 20 and 40 years, although cases have been reported in children and elderly individuals.4,5

Etiologically, hypercoagulable states are the leading contributors to BCS, with a significant proportion of cases associated with myeloproliferative disorders such as polycythemia vera or essential thrombocythemia. Genetic mutations, particularly those involving JAK2 (V617F mutation), are frequently identified in these patients. Other contributing conditions include paroxysmal nocturnal hemoglobinuria, antiphospholipid syndrome, and inherited thrombophilias such as factor V Leiden mutation or prothrombin G20210A mutation. In regions where BCS is associated with membranous obstruction, the pathophysiology may involve a combination of genetic susceptibility and environmental exposures.4,5

In the pediatric population, BCS is exceptionally rare but poses unique diagnostic and therapeutic challenges. Pediatric cases are often linked to congenital anomalies such as inferior vena cava atresia or syndromic conditions affecting vascular development. In contrast, in the elderly, secondary causes like malignant invasion of hepatic veins by hepatocellular carcinoma or other hepatic tumors may predominate.6

The global burden of BCS highlights disparities in access to diagnostic modalities and advanced therapeutic options. In high-resource settings, early diagnosis is supported by imaging techniques such as Doppler ultrasonography, CT, or MRI. This enables timely intervention, often involving transjugular intrahepatic portosystemic shunts (TIPS) or liver transplantation. Conversely, in low-resource environments, delayed diagnosis often results in advanced disease presentation, with more patients requiring surgical interventions for life-threatening complications such as refractory ascites or acute liver failure.7

Despite its low overall incidence, BCS represents a significant health burden due to its potential for severe morbidity and mortality if left untreated. Understanding the epidemiological nuances of BCS, including regional differences in etiology and presentation, is essential for developing targeted management strategies, optimizing surgical outcomes, and reducing disease burden worldwide.7

CLINICAL CONSIDERATIONS

The clinical management of Budd-Chiari syndrome (BCS) requires a comprehensive understanding of its diverse presentations, underlying etiologies, and the multifaceted pathophysiological processes involved in hepatic venous outflow obstruction. Surgical intervention is typically reserved for patients with advanced disease or those who fail to respond to medical and interventional therapies, making careful clinical evaluation a cornerstone of effective treatment planning.7

CLINICAL PRESENTATION

The clinical manifestations of BCS vary widely, ranging from asymptomatic cases detected incidentally to fulminant hepatic failure. BCS is commonly categorized into acute, subacute, and chronic forms, each with distinct clinical and pathological characteristics:8

1. **Acute BCS**: Presents with sudden onset of abdominal pain, hepatomegaly, ascites, and jaundice. These patients may rapidly progress to hepatic failure and require urgent intervention.8
2. **Subacute BCS**: The most common form, characterized by insidious onset of symptoms, including abdominal discomfort, progressive ascites, and mild to moderate liver dysfunction.8
3. **Chronic BCS**: May manifest with complications of portal hypertension, such as variceal bleeding, refractory ascites, and splenomegaly, often accompanied by cirrhosis and progressive liver failure.8

Diagnostic Evaluation

A thorough diagnostic workup is essential to identify the extent of hepatic venous obstruction, assess liver function, and determine the presence of comorbid conditions or complications that may influence surgical management. Key elements include:8

- **Imaging Studies**: Doppler ultrasonography is the first-line modality, offering non-invasive evaluation of hepatic venous flow. Cross-sectional imaging with CT or MRI provides detailed anatomical information, including the location and extent of venous obstruction, collateral formation, and liver parenchymal changes.8
- **Liver Function Tests**: Assessment of hepatic synthetic and excretory functions, including bilirubin, albumin, and coagulation parameters, to stratify the severity of hepatic dysfunction.9

- **Hemodynamic Studies**: Invasive assessment of portal pressures may be warranted to guide surgical decision-making, particularly in cases requiring portosystemic shunting.
- **Etiological Evaluation**: Identification of underlying prothrombotic states, such as myeloproliferative disorders or inherited thrombophilias, through laboratory and genetic testing.9

Preoperative Optimization

Patients with BCS frequently present with significant hepatic and systemic dysfunction, necessitating careful preoperative optimization to improve surgical outcomes:9

- **Management of Ascites**: Refractory ascites is a common complication requiring aggressive medical management, including diuretics and large-volume paracentesis. Albumin infusion and consideration of pharmacological agents such as midodrine may be employed to support intravascular volume.10
- **Coagulation Management**: Balancing anticoagulation therapy is critical, particularly in patients with a history of thrombotic events or ongoing hypercoagulability. Perioperative bridging strategies should be tailored to individual thrombotic and bleeding risks.
- **Nutritional Support**: Malnutrition is prevalent in advanced BCS and should be addressed with dietary optimization and supplementation.1'

Patient Selection for Surgery

Surgical interventions are indicated for patients with refractory symptoms or complications not amenable to medical or less invasive treatments. Patient selection must consider several factors:10

- **Severity of Liver Dysfunction**: Advanced liver failure (e.g., MELD score ≥15) may favor liver transplantation over shunting procedures.10
- **Portal Hypertension**: Significant portal hypertension may necessitate portosystemic shunting to reduce pressure and alleviate symptoms.11
- **Comorbid Conditions**: Patients with systemic disease or malignancy require a multidisciplinary approach to evaluate surgical feasibility and prognosis.11

Surgical Considerations

The choice of surgical technique depends on the underlying pathology, anatomical considerations, and the presence of complications:

1. **Portosystemic Shunts**: Indicated for patients with preserved liver function but significant portal hypertension. Options include mesoatrial, mesocaval, and splenorenal shunts. Technical success relies on creating durable anastomoses and maintaining patency.11
2. **Liver Transplantation**: The definitive treatment for patients with end-stage liver disease or significant hepatic dysfunction. Preoperative planning should address vascular anomalies and the potential for hypercoagulability in the post-transplant period.11
3. **Inferior Vena Cava Reconstruction**: In cases of IVC thrombosis or stenosis, surgical or endovascular reconstruction may be required to restore hepatic venous outflow.12

Postoperative Considerations

Postoperative care should focus on monitoring for complications, including thrombosis, infection, and graft dysfunction. Lifelong anticoagulation and surveillance imaging are often necessary to prevent recurrent venous obstruction and ensure the durability of the surgical intervention.12

By integrating a thorough understanding of clinical presentations, diagnostic findings, and patient-specific considerations, clinicians can optimize the surgical management of BCS and improve long-term outcomes for this complex and challenging condition.13

Treatment

The management of Budd-Chiari syndrome (BCS) involves a multidisciplinary approach that encompasses medical, interventional, and surgical strategies aimed at restoring hepatic venous outflow, alleviating portal hypertension, and preserving liver function. Surgical treatment is reserved for patients with advanced disease, refractory symptoms, or complications that are unresponsive to pharmacological and minimally invasive interventions. Surgical options vary based on the underlying etiology, the severity of hepatic dysfunction, and the presence of systemic or local complications.13

Indications for Surgical Management

Surgical intervention is typically considered in the following scenarios:

- **Refractory Ascites**: Persistent fluid accumulation despite optimal medical and interventional therapies.14
- **Severe Portal Hypertension**: Complications such as variceal bleeding or hepatic encephalopathy due to elevated portal pressures.14
- **Failed or Infeasible Endovascular Therapy**: In cases where angioplasty, stenting, or transjugular intrahepatic portosystemic shunt (TIPS) placement is contraindicated or unsuccessful.15
- **Advanced Hepatic Dysfunction**: Patients with progressive liver failure who are candidates for orthotopic liver transplantation.
- **Extensive Hepatic Venous Outflow Obstruction**: Cases involving complete thrombosis or stenosis of the hepatic veins and inferior vena cava (IVC).15

Surgical Techniques

The primary surgical interventions for BCS include portosystemic shunting, liver transplantation, and vascular reconstruction.15

1. **Portosystemic Shunting**
    - **Objective**: To decompress the portal venous system and reduce portal hypertension by creating a bypass between the portal and systemic venous systems.15
    - **Techniques**:
        - **Mesoatrial Shunt**: Connection between the superior mesenteric vein and the right atrium.15
        - **Mesocaval Shunt**: Anastomosis between the superior mesenteric vein and the inferior vena cava.15
        - **Splenorenal Shunt**: Directing blood flow from the splenic vein to the left renal vein.15
    - **Considerations**: Shunting is best suited for patients with preserved hepatic synthetic function. Technical precision is essential to prevent complications such as shunt thrombosis or excessive hepatic encephalopathy.15
2. **Inferior Vena Cava Reconstruction**
    - **Objective**: To restore venous drainage from the liver in cases of IVC stenosis or occlusion.15
    - **Techniques**:
        - Surgical thrombectomy to remove thrombotic material.
        - Patch angioplasty or graft placement to widen stenotic segments.
        - Bypass grafting for extensive occlusions.16
    - **Outcomes**: These procedures can improve venous return and alleviate hepatic congestion but require long-term anticoagulation to maintain patency.
3. **Orthotopic Liver Transplantation (OLT)**
    - **Indication**: The definitive treatment for end-stage liver disease or fulminant hepatic failure secondary to BCS.16
    - **Procedure**: Replacement of the diseased liver with a donor organ. Special considerations include the management of coexisting hypercoagulable states and ensuring adequate anastomosis of the hepatic veins and IVC.16
    - **Prognosis**: Long-term survival rates are favorable, with significant improvement in quality of life. Lifelong immunosuppression and vigilant monitoring for graft rejection are essential.16
4. **Hybrid and Minimally Invasive Approaches**
    - Advances in surgical and interventional radiology have enabled the integration of minimally invasive techniques with traditional surgery. Examples include combining endovascular stenting with open shunting procedures or using laparoscopic techniques for liver transplantation in select cases.16

Postoperative Management

The postoperative phase focuses on preventing complications and ensuring the durability of surgical interventions:

- **Anticoagulation**: Lifelong anticoagulation is typically required to prevent recurrence of thrombotic events, particularly in patients with underlying prothrombotic disorders.16
- **Immunosuppression**: For liver transplant recipients, immunosuppressive therapy is mandatory to prevent graft rejection.16
- **Surveillance**: Regular imaging studies, including Doppler ultrasound and CT or MRI, are performed to assess the patency of shunts or vascular reconstructions.16
- **Management of Complications**: Monitoring for postoperative infections, hepatic encephalopathy, or shunt dysfunction is critical.16

The outcomes of surgical management for BCS depend on multiple factors, including the timing of intervention, the severity of liver dysfunction, and the patient's overall health. Portosystemic shunting provides symptom relief in many cases, but progression to liver failure may necessitate transplantation. Liver transplantation offers a curative option with favorable survival rates, exceeding 75% at five years in experienced centers. Vascular reconstruction techniques continue to evolve, improving the prognosis for patients with complex venous obstruction.16

The surgical treatment of BCS remains a cornerstone for managing advanced or refractory cases. By tailoring interventions to the patient's clinical condition and incorporating multidisciplinary expertise, surgical strategies can significantly improve outcomes and enhance the quality of life for individuals affected by this rare but severe condition.16

CONCLUSION

The surgical management of Budd-Chiari syndrome (BCS) represents a cornerstone in the treatment of advanced cases characterized by hepatic venous outflow obstruction, refractory symptoms, and progressive liver dysfunction. This rare yet serious condition demands a nuanced approach that integrates a thorough understanding of its pathophysiology, clinical heterogeneity, and underlying etiologies. Surgical interventions, including portosystemic shunting, vascular

reconstruction, and orthotopic liver transplantation, offer life-saving options for patients who fail to respond to medical and interventional therapies.

The decision-making process in the surgical management of BCS must be guided by comprehensive clinical assessment and advanced diagnostic imaging to delineate the extent of venous obstruction, evaluate liver function, and identify coexisting systemic or thrombotic disorders. Patient selection for specific surgical techniques should consider factors such as the severity of portal hypertension, the degree of hepatic dysfunction, and the feasibility of less invasive interventions. Multidisciplinary collaboration, involving hepatologists, interventional radiologists, and transplant surgeons, is essential to optimize outcomes and tailor treatment to the individual patient's needs.

Portosystemic shunting, whether through mesoatrial, mesocaval, or splenorenal techniques, has proven effective in alleviating portal hypertension and improving venous drainage in patients with preserved liver function. However, these procedures carry the risk of complications such as shunt thrombosis and hepatic encephalopathy, necessitating meticulous surgical technique and rigorous postoperative monitoring. In cases of extensive venous thrombosis or IVC stenosis, vascular reconstruction provides an alternative means of restoring hepatic venous outflow.

For patients with end-stage liver disease or fulminant hepatic failure, orthotopic liver transplantation offers the definitive solution, addressing both the venous outflow obstruction and the sequelae of hepatic decompensation. Advances in surgical techniques and postoperative care have significantly improved long-term survival rates and quality of life for transplant recipients. Nevertheless, liver transplantation entails challenges such as lifelong immunosuppression and the potential for recurrence of thrombotic complications, particularly in patients with predisposing hypercoagulable states.

The success of surgical management for BCS hinges on careful postoperative care, including anticoagulation to prevent recurrent thrombotic events, surveillance imaging to ensure the durability of interventions, and ongoing management of complications such as hepatic encephalopathy or graft dysfunction. Moreover, addressing the underlying etiologies of BCS, such as myeloproliferative disorders or inherited thrombophilias, is critical to achieving durable remission and minimizing the risk of disease recurrence.

In conclusion, while the surgical management of BCS remains challenging, advances in diagnostic modalities, surgical techniques, and perioperative care have significantly improved outcomes for this complex condition. By adopting a personalized and multidisciplinary approach, clinicians can maximize the therapeutic benefits of surgery and offer patients the prospect of long-term survival and improved quality of life. Future research should continue to refine surgical strategies, explore minimally invasive alternatives, and investigate the molecular underpinnings of BCS to enhance treatment paradigms further. This commitment to innovation and individualized care will be pivotal in advancing the management of this rare but impactful disease.

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