

CLINICAL SPECTRUM AND OUTCOMES OF ENDOVASCULAR INTERVENTION IN CHRONIC BUDD–CHIARI SYNDROME: EXPERIENCE FROM A TERTIARY CARE CENTRE IN SOUTH INDIA

Hepatology

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ABSTRACT

Objective & Aim: This study aimed to evaluate the clinical spectrum and outcomes of Endovascular interventions in patients with Chronic Budd–Chiari Syndrome (CSBS) treated at a tertiary care centre in South India. **Methods:** A total of 45 consecutive patients with CBCS were prospectively evaluated. Patients presenting with abdominal pain, ascites, jaundice, upper gastrointestinal bleeding or worsening liver function were included. Based on the site of venous obstruction- Hepatic vein (HV), Inferior vena cava (IVC), or Combined IVC and HV-patients underwent endovascular interventions with anticoagulation therapy. Technical success, clinical success, reintervention rate, mortality rate and need for liver transplantation were analysed for a period of 6 months. **Results:** Of the 45 patients, 10 (22.2%) had isolated IVC obstruction, 15 (33.3%) had HV obstruction, and 20 (44.4%) had combined IVC and HV obstruction. Interventions included IVC angioplasty (n=9), IVC angioplasty with stenting (n=1), HV angioplasty (n=5), HV stenting (n=7), combined IVC and HV angioplasty (n=9), combined stenting (n=2), and Direct Intrahepatic Portosystemic Shunt (DIPS) (n=12). Technical success was achieved in 44 patients (97.8%), and clinical success in 37 (82.2%). Complications occurred in 9 patients (19.9%), and Reintervention was required in 5 patients (11%). Two patients (4.4%) required liver transplantation and overall mortality was 1 (2.2%) within 6 months. **Conclusion:** Combined IVC and HV obstruction was the predominant pattern of obstruction in this study. Endovascular intervention demonstrated high technical and clinical success with low morbidity and mortality, supporting its role as an effective definitive treatment and bridge to liver transplantation in chronic Budd–Chiari syndrome.

KEYWORDS

Chronic Budd–Chiari Syndrome; Endovascular Intervention; Inferior Vena Cava; Hepatic Vein; Direct Intrahepatic Portosystemic Shunt; Liver Transplantation

INTRODUCTION:

Budd Chiari syndrome is defined as obstruction of hepatic venous outflow that can be located from the small hepatic venule up to the entrance of the IVC into the right atrium.¹ Budd Chiari syndrome is a diverse disease with regard to the site of obstruction, the predisposing thrombophilic disorders and clinical presentation across the Asia-Pacific.² Geographical variations in disease pattern of Budd Chiari syndrome exist, which may reflect differing predisposing factors. While earlier studies from India reported isolated inferior vena cava (IVC) obstruction as the commonest disease type, this is a minority in more recent reports where a combination of IVC and hepatic vein obstruction is the commonest type. Poverty, malnutrition, recurrent bacterial infections and filariasis have been previously suggested as predisposing factors for IVC obstruction. Improvement in hygiene and sanitation may partly explain the recent change in disease profile of Budd Chiari syndrome in India.³ BCS should be considered in any patient with liver disease (recent or chronic) of unknown etiology.⁴ The pathophysiological consequences include obstruction which leads to sinusoidal congestion, ischemia, and finally hepatocellular necrosis. They can result in centrilobular fibrosis, nodular regenerative hyperplasia and/or cirrhosis.⁵ Imaging plays an important role both in establishing the diagnosis of Budd–Chiari syndrome as well as evaluating for underlying causes and complications such as portal hypertension.⁶ In a large cohort of patients with BCS recruited over a short period, a step-wise treatment approach provides good long-term survival.⁷ A progressive “step up” therapeutic strategy according to the clinical response from less to more invasive therapies is recommended for HVT/BCS. Liver Transplantation is reserved for HVT/BCS patients in whom the medical and vascular interventional approaches fail. TIPS/DIPS should be considered while waiting for transplant because it may improve liver function and potentially avoid the need for transplantation.⁴ The present study aimed to describe the clinical spectrum and evaluate outcomes of endovascular interventions in chronic Budd–Chiari syndrome at a tertiary hepatobiliary centre in South India.

MATERIAL AND METHOD:

Study Design And Patient Selection:

All consecutive chronic Budd Chiari syndrome patient who underwent Endovascular interventions at Madras medical college in Chennai

were prospectively analysed for 6 month (January 2025 to July 2025). Informed consent was taken prior to procedure. This study was approved by Ethical committee of Madras medical college, Chennai (No. IEC – MMC/Approval/18012025). Patient's details collected were Age at onset of disease, Gender and symptoms at presentation: Abdominal pain, Ascites, Jaundice, Gastrointestinal bleed, Pedal edema, Veins on the abdomen and back, and Hepatic encephalopathy. Laboratory data included Complete blood test, Liver function test, Renal function test, Prothrombin time, International normalized ratio (INR), Viral serology. The diagnosis of BCS was established by imaging with Doppler ultrasound abdomen, CT Abdomen Angiogram. Upper Gastrointestinal endoscopy, Chest X-ray, electrocardiogram, two-dimensional echocardiography was also done.

Endovascular Intervention:

Once Chronic BCS was confirmed, patients were counselled on various therapeutic options and subjected to interventional radiological procedure after obtaining informed consent. Based on site of obstruction, length of obstruction, feasibility of endovascular procedure underwent IVC angioplasty ± stenting, HV angioplasty ± HV stenting, combined angioplasty or combined stenting or DIPS. Post-procedure patients were observed in the intensive care unit and started on anticoagulation Heparin injection at a dose of 5000 IU four times a day for 72 hours with overlapping oral anticoagulation Warfarin, to maintain INR between 2 and 3. Follow up Doppler ultrasound was performed on Day 3, Day 7 and Day 30 then at 3month, 6 month after the procedure. In each visit, complete hemogram, liver function tests, prothrombin time, INR, and a Doppler ultrasound were done. Technical success was defined as successful completion of the intended procedure with satisfactory restoration of flow across the obstruction, Clinical success was defined as symptomatic improvement with documented post-procedural patency. The response to Intervention, requirement for Reintervention, procedure-related complications, and mortality were systematically evaluated.

Reintervention And Mortality:

In case of technical failure or clinical failure occurs, if feasible planned for Reintervention such as Angioplasty or Stenting or DIPS or DIPS Stent plasty. If not feasible for reintervention, then registered for liver transplantation with continuation of anticoagulation. If patient died

during procedure or during follow up period was recorded.

Statistical Analysis:

The collected data were entered in the Microsoft Excel 2016 and analysed with IBM SPSS Statistics for Windows, Version 29.0.(Armonk, NY: IBM Corp). To describe about the data descriptive statistics frequency analysis, percentage analysis was used for categorical variables and the mean & S.D were used for continuous variables. The Kaplan Meier survival curve was used for the survival analysis.

RESULTS:

A total of 45 patients were included in the study, with a mean age of 34.6 ± 12.7 years, ranging from 15 to 70 years. Among them, 28 (62.2%) were female and 17 (37.8%) were male, yielding a female-to-male ratio of approximately 1.6:1, indicating a higher prevalence among females. The most common presenting symptom was Abdominal distension observed in 36 patients (80%), followed by Abdominal pain in 8 patients (17.8%) and Jaundice in 1 patient (2.2%).The majority had combined inferior vena cava (IVC) and hepatic vein (HV) obstruction (44.4%), followed by isolated hepatic vein obstruction (33.3%) and isolated IVC obstruction (22.2%) (Figure 1)

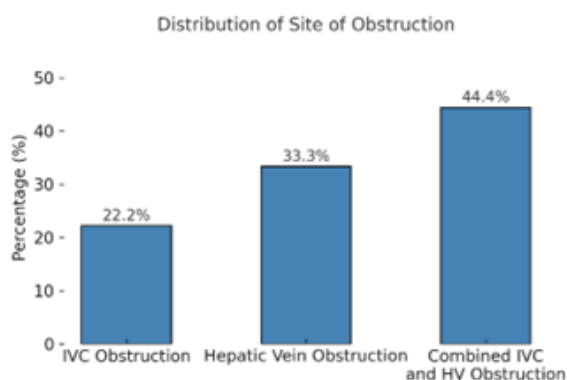


Figure 1. Distribution of site of venous obstruction among patients with chronic Budd–Chiari syndrome.

ENDOASCULAR INTERVENTION:

IVC ANGIOPLASTY±STENTING:

Among 45 patients, 9 patients(20%) underwent IVC angioplasty, while 1 patient (2.2%) underwent angioplasty with stenting. Two IVC angioplasty patients developed re-thrombosis with recurrence of ascites, both subsequently underwent IVC angioplasty with stent placement, achieving technical and clinical success.

HV ANGIOPLASTY±STENTING:

In this study 5 patients (11.1%) underwent balloon angioplasty of the hepatic vein and 7 patients (15.6%) underwent stent placement for HV obstruction. Stent thrombosis occurred in 1 HV stenting patient so underwent Direct Intrahepatic Portosystemic Shunt (DIPS) with better outcome on follow up.

COMBINED IVC AND HV ANGIOPLASTY±STENTING:

Combined IVC and HV balloon angioplasty was done in 9 patients (20%) and stent placement for combined lesions was performed in 2 patient(4.4%). Technical and clinical success was not achieved in 1 patient who underwent combined IVC and HV angioplasty he also worsened with Acute kidney injury development and died in 1 month.

DIRECT INTRAHEPATIC PORTOSYSTEMIC SHUNT:

Direct Intrahepatic portosystemic shunting was performed in 12 patients (26.7%). Technical and clinical success was achieved in all patients. But DIPS stent thrombosis occurred during follow up in Four patients (8.9%). Among them, two patient underwent DIPS stent plasty following stent thrombosis of a prior shunt. Technical and clinical success was achieved in that 2 patients. In remaining 2 patient as DIPS stent plasty was not feasible registered and underwent liver transplantation.

In this study, technical success was achieved in 44 patients (97.8%), while clinical success was achieved in 37 patients (82.2%). Procedure-related complications included Re-thrombosis or Stent thrombosis,

Acute kidney injury and Anticoagulation-induced coagulopathy were observed in 9 patients (19.9%). One patient developed a significant intrahepatic hematoma, which resolved with conservative management. Reintervention was required in 5 patients (11%), which included IVC stenting in 2 (4.4%), DIPS in 1 (2.2%), and DIPS stent plasty in 2 (4.4%). Technical success was achieved in all Reintervention cases; However, two patients (4.4%) required liver transplantation within 6 months.

The Kaplan–Meier survival analysis demonstrates high cumulative survival among patients following endovascular intervention. The survival probability remained above 95% throughout the 6 months follow-up period. Only one mortality event was observed, indicating good outcomes after successful endovascular management. (Figure 2)

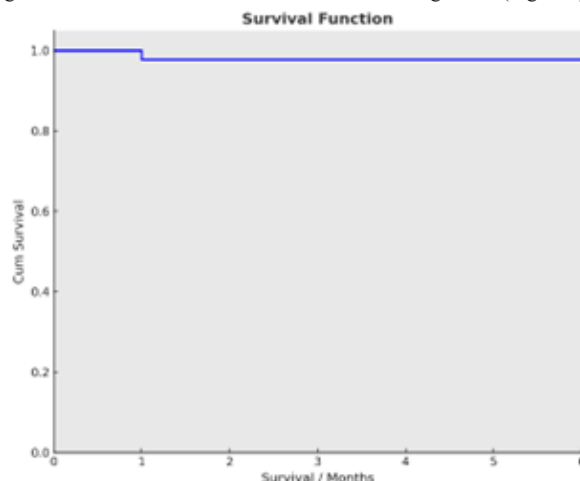


Figure 2. Kaplan–Meier survival curve showing cumulative 6 months survival following Endovascular intervention.

DISCUSSION:

In this prospective study of 45 patients with chronic Budd–Chiari syndrome, a clear female predominance (1.6:1) was observed, consistent with previous Indian and International studies. Abdominal distension was the most frequent presenting symptom, followed by Abdominal pain and Jaundice, aligning with findings from Singh et al., study reported abdominal pain, distension, jaundice, and upper gastrointestinal bleeding as the commonest clinical manifestations in BCS.⁸Regarding the site of venous obstruction, combined inferior vena cava (IVC) and hepatic vein (HV) involvement was the most prevalent pattern (44.4%), followed by isolated HV obstruction (33.3%) and isolated IVC obstruction (22.2%). This distribution mirrors the observations of Eapen et al., study noted that combined obstruction was the dominant anatomical pattern in Indian patients³ and is also consistent with Jagtap et al., who reported combined IVC–HV involvement in 14.8% of cases, isolated HV obstruction in 37.5%, and isolated IVC obstruction in 47.7% of their cohort⁹

Endovascular interventions formed the mainstay of management, with IVC angioplasty with or without stenting performed in 10 patients, HV angioplasty with or without stenting in 12 patients, combined IVC and HV angioplasty with or without stenting in 11 patients, and Direct Intrahepatic Portosystemic Shunt (DIPS) in 12 patients. These findings reflect a stepwise therapeutic approach similar to that described by Seijo et al., where progressive escalation from angioplasty to shunt creation achieved long-term survival rates of 69–89%.⁷ In our study following endovascular intervention, technical success was achieved in 97.8% and clinical success in 82.2% of patients, paralleling outcomes reported in large Asian studies and in Jagtap et al., who demonstrated 98.9% technical and 86.4% clinical success with a 1-year stent patency of 90.9%.⁹ Percutaneous recanalization yielded sustained patency and survival, supporting the conclusion of Chinese retrospective studies, where recanalization success exceeded 95% with combined angioplasty and stenting yielding better long-term outcomes than angioplasty alone.¹⁰ In our series, Direct Intrahepatic Portosystemic Shunt (DIPS) was performed in 12 patients (26.7%). During follow-up, stent thrombosis occurred in four patients (8.9%). Among these, two patients underwent successful DIPS stent plasty, achieving technical and clinical success. While in the remaining two patients, stent plasty was not feasible and both subsequently

underwent liver transplantation. Our results are in accordance with Mancuso et al. study. TIPS placement done in 14 of 15 CBCS patients, TIPS dysfunction occurred in four patients, managed successfully with balloon dilatation or re-stenting, and no patient required transplantation.¹¹ Similarly, a large multicentre European study involving 124 BCS patients treated with TIPS reported excellent 1- and 5-year orthotopic liver transplant (OLT)-free survival rates of 88% and 78%, respectively.¹²

Procedure-related complications occurred in approximately 20% of patients. Reintervention was required in 5 patients (11%), including IVC stenting in 2 (4.4%), DIPS in 1 (2.2%), and DIPS stent plasty in 2 (4.4%). Technical success was achieved in all reintervention cases; however, two patients (4.4%) required liver transplantation 6 months. The overall survival in our cohort exceeded 95% at six months, with only one mortality event. These findings are similar with the results of Ampurukar et al., study emphasizing the feasibility and durable benefit of DIPS and angioplasty in well-selected BCS patients.¹³ Overall, endovascular therapy demonstrated high efficacy, low morbidity, and acceptable Reintervention rates.

CONCLUSION:

Combined Inferior Vena Cava and Hepatic vein obstruction was the predominant site of obstruction. Patients with Chronic Budd–Chiari syndrome demonstrated favourable outcomes following Endovascular Intervention. In our study, patients who developed stent thrombosis were successfully managed with Reintervention. Even in cases of DIPS stent thrombosis, DIPS Stent plasty achieved good technical and clinical success. Liver transplantation was required only in patients in whom Reintervention was not feasible. Given its Efficacy and Accessibility, Endovascular Intervention was effective definitive treatment and bridge to liver transplantation in Chronic Budd–Chiari syndrome.

ABBREVIATIONS:

CBCS - Chronic Budd–Chiari Syndrome

HVT - Hepatic Vein Thrombosis

IVC - Inferior Vena Cava

HV - Hepatic Vein

DIPS - Direct Intrahepatic Portosystemic Shunt

TIPS - Trans jugular Intrahepatic Portosystemic Shunt

OLT - Orthotopic Liver Transplantation

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