Outcomes of Hepatic Venoplasty in Budd-Chiari Syndrome

Adnan Salim[1], Khurram Shafique[2], Sadia Jabbar[3], Zia Ur Rehman[1] and Kashif Malik[1]

[1]*Department of Gastroenterology and Hepatology, Shaikh Zayed Hospital, Lahore, Pakistan*
[2]*Department of Radiology, Shaikh Zayed Medical Complex, Lahore, Pakistan*
[3]*Department of Gastroenterology, Lahore General Hospital, Lahore, Pakistan*

ABSTRACT

Objective: To observe medium to long-term outcomes of hepatic venoplasty in patients with Budd-Chiari syndrome (BCS).

Study Design: An observational study.

Place and Duration of the Study: Department of Gastroenterology and Radiology, Shaikh Zayed Hospital, Lahore, Pakistan, from February 2021 to December 2022.

Methodology: After obtaining ethical approval, adult patients presenting to the authors' Institute with BCS and candidates for venoplasty were recruited. After the procedure, they were followed up for twelve months.

Results: Twenty-five patients were recruited including 18 (72%) males and 7 (28%) females. Vascular occlusion was due to identified hypercoagulable state in 11 (44%) and idiopathic in 14 (56%). Eight (32%) patients had liver cirrhosis. Ascites was present in 24 (96%) patients prior to venoplasty. Twenty-one (84%) patients had varices. Stage of liver disease was child A in 5 (20%), B in 16 (64%), and C in 4 (16%). Vascular blockage involved all hepatic veins in nearly a quarter of patients. Vascular stenting post-venoplasty was done in 18 (72%). All patients received warfarin (target INR 2-2.5). Twelve months after venoplasty, 4 (16%) patients had undergone liver transplantation, 4 (16%) lost to follow up, and 6 (24%) had expired due to cirrhosis-related complications. In the remaining 11 (44%), hepatic veins were patent in 10 (40%) and blocked in 1 (4%). One (4%) patient had significant ascites, whereas significant-sized varices were noted in 2 (8%).

Conclusion: Venoplasty results in good long-term outcomes in patients with BCS if they have not developed advanced cirrhosis. In patients with cirrhosis, outcomes are suboptimal and liver transplantation remains the only curative option. This highlights the importance of early detection and specialist centre referral before cirrhosis develops.

Key Words: *Budd-Chiari Syndrome, Venoplasty, Cirrhosis*.

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INTRODUCTION

Budd-Chiari Syndrome (BCS) is defined as obstruction of hepatic venous outflow. The obstruction may be due to thrombosis or non-thrombotic causes. The site of obstruction can be at any location from the hepatic venules to the junction of the inferior vena cava (IVC) with the right atrium.[1] This results in an increase in hepatic sinusoidal pressure with resultant portal hypertension.[2] There are various known aetiological factors for the development of BCS and as such, BCS is divided into primary and secondary BCS based on aetiology. Patients with vascular obstruction occurring due to underlying thrombophilic disorders are classified as having primary BCS.[1] Patients having hepatic venous compression or invasion from an external source, such as neoplasm, are characterised as having secondary BCS.[3]

Correspondence to: Dr. Adnan Salim, Department of Gastroenterology and Hepatology, Shaikh Zayed Hospital, Lahore, Pakistan
E-mail: adnansalim1147@gmail.com
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The most common causes of primary BCS are myeloproliferative diseases, accounting for almost 50% of cases.[4] Other causes include factor V Leiden mutation, protein C, protein S and antithrombin III deficiencies, antiphospholipid syndrome, pregnancy, and oral contraceptives.[5] The cause of BCS varies in Western and Eastern populations. In the West, it is mainly due to prothrombotic disorders. In the East, the most common causes include membranous obstruction of the vena cava (MOVC) and primary IVC thrombosis.[5]

The spectrum of clinical manifestation of BCS is wide and may vary from asymptomatic to fulminant. Patients may present with acute, subacute, chronic, or fulminant hepatic failure. Acute BCS is rare and usually develops within one month.[6] Subacute presentation is the most common clinical form in prothrombotic patients.[5] Chronic BCS is characterised by portal hypertension and its complications.

The fulminant form presents with acute liver failure. Asymptomatic forms may occur in 15-20% of patients. The classical triad of abdominal pain, ascites, and hepatomegaly is found in most patients. At first presentation to a clinician, over 60% patients with BCS patients have abdominal pain and ascites. Only about 20% patients have painless ascites. Other, less frequent, features include jaundice, splenomegaly, pedal oedema, portosystemic encephalopathy, and gastrointestinal bleeding.[5-7]

Radiologic imaging is the mainstay for the diagnosis of BCS. Amongst imaging techniques, Doppler ultrasound and MRI remain the gold standard for diagnosis.[6] However, contrast-enhanced triphasic computed tomography can be used because of the limited availability of MRI in many centres. Radiologic features include direct signs such as blockage or compression of hepatic veins and/or the IVC and venous collaterals. Indirect signs include morphological changes in liver with caudate lobe hypertrophy and delayed nodule formation.[6] Other laboratory tests such as albumin, PT, INR, ALT, creatinine, and bilirubin levels have limited value in diagnosis. These are mainly used for the assessment of disease severity and are of prognostic value.[8]

Treatment options for BCS include medical management with anticoagulants, recanalisation therapies, surgical shunting, transjugular intrahepatic portosystemic shunt (TIPS), and orthotopic liver transplantation. Recanalisation therapies include balloon angioplasty, stenting, and thrombolytic therapy. Compared to other, more prevalent, causes of liver disease such as liver cirrhosis and its complications including vascular thrombosis, patients with BCS are far lesser in number in Pakistan. Combined with the high cost of interventional radiological procedures with their requisite highly trained operators, it is difficult to conduct clinical trials comparing different treatment modalities. This is further complicated by the fact that the recanalisation procedure is limited to a handful of tertiary care centres in Pakistan. Hence, most recommendations are based on expert opinions, case reports, retrospective studies, or those examining short-term outcomes.[2] Anticoagulation therapy is usually the first-line treatment option. Recanalisation with or without TIPS or TIPS alone is done if the disease progresses despite anticoagulation therapy.[1,8] Percutaneous interventions have been practised for some time with the earliest reported cases from the mid to late 1980s.[9,10] Recanalisation therapy is used in selected patients after assessment of site and length of obstruction and clinical condition of patients.[11] Surgical shunt procedures are less often used these days because of technical difficulties and high perioperative mortality.[8] Orthotropic liver transplantation is reserved for patients with fulminant hepatic failure and when all other treatment options fail, including patients with advanced cirrhosis.[1,8] As there is very limited data on prognostic implications of recanalisation therapies in Pakistan as compared to international data, the aim of this study was to determine the short-to medium-term outcomes of hepatic venoplasty in patients with BCS.

METHODOLOGY

BCS was defined as the obstruction of hepatic venous outflow anywhere from small hepatic veins to the suprahepatic inferior vena cava.

Hepatic venoplasty was defined as balloon dilatation with or without stenting of obstructed hepatic veins. The primary outcome was the patency of stent ensuring hepatic venous outflow after 12 months of venoplasty. The secondary outcomes were improvement in ascites and oesophageal varices after 12 months of venoplasty as a marker of improvement in portal hypertension.

It was an observational study, initiated after obtaining approval from the Ethical Review Committee of the Department of Gastroenterology and Hepatology, Shaikh Zayed Hospital, Lahore, Pakistan, from February 2021 to December 2022.

All adults aged 18 and above presenting to the authors' institution with BCS who were confirmed to have hepatic venous obstruction by undergoing Doppler abdominal ultrasound and contrast-enhanced CT and were candidates for hepatic venoplasty were recruited. Patients who had at least one hepatic vein with some degree of patency proximal to the central occlusion/stenosis were candidates for venoplasty/stenting. Sampling was of the non-probability consecutive type and temporal i.e. time-based, recruiting all cases presenting from February 2021 to December 2021. Patients were excluded if they were already listed for liver transplantation within the following 2-4 weeks, had IVC or hepatic vein stenosis due to malignancy, had previously undergone surgical shunt procedure or liver transplantation, and those who had inaccessible veins with excessive occlusion in the intrahepatic portion with no patency of lumen who could not undergo vascular intervention.

The patients were counselled and explained the details of the procedure and the purpose of the study. Written informed consent and detailed history were taken from each patient. Data on variables such as age, gender, child class, cause of disease, intervention details, and data regarding outcomes were collected through a specifically designed proforma.

After the standard preparation, a micro-puncture needle set was used to gain access to the hepatic venous system followed by contrast injection to assess the level of obstruction of the hepatic vein. A stiff shaft-glide wire with a multipurpose catheter was then passed across the obstruction. Once the obstruction was crossed, the stiff shaft-glide wire was exchanged with a super stiff wire. The obstruction site was successively dilated with 6 mm, 8 mm, 10 mm, and 12 mm balloons. Following this, repeated imaging of the dilated vein was performed. If images showed good patency, the procedure was concluded. If elastic recoil of the dilated vessel wall was observed, a 12 mm x 40 mm self-expanding metal stent was placed. All patients were placed on warfarin post-procedure with a target INR of 2-3. Patients were followed up for 12 months to check for survival, patency of hepatic veins, degree of ascites, and size of varices.

All collected data were entered and analysed through SPSS version 22. Numerical variable i.e. age was presented by mean ± SD. Categorical variables i.e. gender, ascites, variceal size, and hepatic vein patency were presented as frequency and percentages at 6 and 12 months.

RESULTS

A total of 25 patients, comprising 18 (72%) males and 7 (28%) females, were recruited. The causes of vascular occlusion were protein C and S deficiency in 3 (12%), JAK II mutation in 3 (12%), protein C and antithrombin II in 2 (8%), protein C only in 2 (8%),

thrombosis secondary to ongoing prothrombotic inflammatory condition (in this case, ulcerative colitis) in 1 (4%), and idiopathic in 14 (56%) patients. Eight (32%) patients had liver cirrhosis evidenced by a coarse liver on ultrasound. Ascites was present in 24 (96%) patients prior to venoplasty, its severity being mild-to-moderate in 11 (44%) and severe in 13 (52%) patients. Twenty-one (84%) patients had varices on screening endoscopy. The stage of liver disease, as per Child Pugh Turcotte score, was Child A in 5 (20%), B in 16 (64%), and C in 4 (16%) patients.

Extent of vascular occlusion near IVC was in the middle hepatic vein, left hepatic vein, and right and left hepatic vein in one (4%) patient each; right and middle hepatic veins in 2 (8%) and all three hepatic veins in 18 (72%) patients. In the last group, the vein selected for venoplasty had some portion of patent lumen in its intrahepatic course. The IVC was thrombosed in 10 (40%) patients. A trans-hepatic venous access approach was utilised in all 25 (100%) patients. Self-expanding metal stents were passed in 18 (72%) patients. All patients received post-procedure anticoagulation with warfarin with a target INR of 2-2.5.

Six-month post-procedure, 3 (12%) patients were lost to follow up, 3 (12%) had undergone living donor liver transplantation, and 5 (20%) patients had expired. In the remaining 14 (56%) patients, Doppler ultrasound at six months showed patent hepatic veins in 10 (40%) patients. Hepatic veins were found to be blocked in 4 (16%). Ten (40%) patients had no or minimal ascites, while 4 (16%) had significant ascites. Nine (36%) had no or small sized varices, whereas medium to large varices were noted in 5 (20%).

Patients with blocked veins and those with significant ascites were referred for liver transplantation. Pending donor availability, they were managed with beta-blockers and diuretics (Table I).

Table I: Progress of clinical parameters at six months post-venoplasty (n = 14).

Clinical parameters	Six months
Ascites	
None / minimal	10 (40%)
Moderate / severe	4 (16%)
Varices	
None / small	9 (36%)
Medium / large	5 (20%)
Hepatic vein patency	
Patent	10 (40%)
Blocked	4 (16%)

Table II: Progress of clinical parameters at twelve months post-venoplasty (n = 11).

Clinical parameters	Twelve Months
Ascites	
None / minimal	10 (40%)
Moderate / severe	1 (4%)
Varices	
None / small	9 (36%)
Medium / large	2 (8%)
Hepatic vein patency	
Patent	10 (40%)
Blocked	1 (4%)

Twelve month post-procedure, one (4%) more patient had undergone liver transplantation. One (4%) was lost to follow up, and one (4%) more patient had expired. In the remaining 11 (44%) patients, Doppler ultrasound at 12 months showed patent hepatic veins in 10 (40%) patients. Hepatic veins were found to be blocked in one (4%) patient. Ten (40%) patients had no or minimal ascites, while one (4%) had significant ascites. Nine (36%) had no or small-sized varices whereas medium-sized varices were noted in two (8%) patients.

Patients who had not undergone transplant were managed with diuretics and beta blockers (Table II).

The causes of death among the 6 (24%) patients were hepatorenal syndrome and refractory ascites in 4 (16%), sepsis in 1 (4%), and hepatic decompensation post variceal bleeding in 1 (4%) patient. No procedure-related complications were observed.

DISCUSSION

This study shows various challenges faced in the management of BCS. These challenges are at every step of the disease manifestation and include late detection and late presentation, whereby cirrhosis has already set in. They include a scarcity of specialised centres and interventionists who are able to diagnose and treat the disease in time. Furthermore, high procedural costs limit access of a vast majority of patients. In addition, poor knowledge, attitude, and practices regarding follow-up after therapeutic procedures result in deterioration even after initial management has somewhat stabilised the patient. These challenges are representative of the general healthcare-related shortcomings whereby diseases remain undiagnosed for prolonged time periods due to late diagnosis and referral. This necessitates complex and expensive interventions and procedures such as liver transplantation by the time patients present to tertiary care centres. Despite repeated counselling, there remains a significant proportion of patients, who after having undergone high-end treatment, fail to follow up with their healthcare teams.

BCS remains a topic where in-country studies and research in Pakistan are limited. The data observed previously were mainly observational or short term after intervention.[1,12] This is the first study in the country to prospectively see the results of active intervention into this disease entity and observe medium-term clinical outcomes.

In this patient population, the frequency of a hypercoagulable state was seen in 44% of cases, which is less than observed elsewhere. A lesser number of patients having full-blown cirrhosis at the time of presentation was also observed. The number of patients having ascites, however, was similar to other studies. In regards to the degree of vascular involvement, the present study figures again echoed the nearly 50% occlusion of the IVC in patients in other studies.[1,12-14]

As stated above, vascular interventions for BCS started to come to light in the 1980s. Prior to that, surgery was the only known option, however, this does not mean that it is no longer used.[15] Vascular intervention is now deemed standard therapy, where

permissible, depending on the degree of vascular occlusion. It requires trained and experienced interventional radiologists. Local and international studies have confirmed the safety and efficacy of this procedure in both adults and children.[1,16-18]

Although TIPS is also used in the management of BCS, it was not performed in any of this study's patients, and data indicate that intervention on the hepatic veins alone without performing TIPS shows fewer complications related to the procedure. All the patients were placed on anticoagulation with warfarin, keeping a target INR of 2-3. This practice of combining anticoagulation with vascular intervention is also deemed superior to endovascular therapy alone.[19]

The authors noted a significant mortality in nearly a quarter of patients who died in the 12-month postvenoplasty observation period. The common condition among all those who died was advanced liver cirrhosis at initial recruitment. Thus, mortality among these was attributable to advanced liver disease, already existing at the time of the procedure. This highlights the importance of early diagnosis of disease before cirrhosis develops. This is also seen in other studies whereby liver function markers indicating more advanced disease, lead to worse outcomes following venoplasty.[20]

Unfortunately, a detailed ultrasound evaluation of the hepatic vessels for any anomalies is beyond the expertise of primary healthcare teams. This leads to late referral. In addition, the presence of ascites, negative viral markers (in a country/region where viral hepatitis is endemic and the most common cause of ascites), initial low serum albumin gradient ascites (as observed in early stages of BCS), and a low ascitic fluid cell count with slightly raised neutrophils also convinces many clinicians that they are dealing with abdominal tuberculosis, the latter again being common in this part of the world. This happens more often than reported and many patients are several weeks into their anti-tuberculous therapy when they come to a specialised tertiary centre where proper treatment is initiated.[21] The authors have also observed the same at their centre whereby a significant number of patients admitted with BCS were on ATT. This underlines the importance of trained radiologists closer to non-urban population centres, in *Tehsil* / District HQ hospitals. This also needs more outreach by specialist centres to conduct training workshops in order to better tune GPs and radiologists in peripheral areas and internists (later in large cities) for them to recognise pathologies outside the large realm of viral hepatitis.

The limitations of this study include the rarity of BCS, expensive treatment, and a limited number of centres able to treat the disease. The cumulative effect of all these factors inevitably leads to a small study population. In an ideal scenario, whereby affordable access to treatment is possible, a larger study may be conducted which may also allow a more comprehensive comparison of treatment response in patients with early-stage liver disease *versus* those with advanced cirrhosis and its complications.

CONCLUSION

It is important to diagnose BCS early before advanced cirrhosis develops and liver transplantation becomes the only curative option. Early detection, before the development of advanced liver disease, coupled with immediate referral to specialist centres allows timely endovascular treatment which results in excellent long-term outcomes.

ETHICAL APPROVAL:

This study was initiated after obtaining ethical approval from the Institutional Review Board of the Shaikh Zayed Federal Postgraduate Medical Institute, Lahore, Pakistan (IRB ID: SZMC/IRB/Internal/272/2021; Dated: 15 February 2021).

PATIENTS' CONSENT:

Informed consent was obtained from the patients to publish the data concerning this case.

COMPETING INTEREST:

The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

AS: Conception, design, acquisition, analysis, interpretation of data, drafting of the work, and revising the manuscript critically for important intellectual content.
KS, SJ: Conception, design, acquisition, analysis, and interpretation of data.
ZUR: Drafting of the work and revising the manuscript critically for important intellectual content.
KM: Final approval of the version to be published.
All authors approved the final version of the manuscript to be published.

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