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A Prospective Study on Etiology Based Prevalence, Clinical Spectrum and Outcome of Budd Chiari Syndrome in Southern India

Authors
Arnab Dey[1*], K. Narayanasamy[2], K. Premkumar[3], A. Santhi Selvi[4]
[1]DM Hepatology Postgraduate, Institute of Hepatobiliary Sciences, Madras Medical College, Chennai, Tamil Nadu
[2]Director and Professor, Institute of Hepatobiliary Sciences, Madras Medical College, Chennai, Tamil Nadu
[3]Associate Professor, Institute of Hepatobiliary Sciences, Madras Medical College, Chennai, Tamil Nadu
[4]Assistant Professor, Institute of Hepatobiliary Sciences, Madras Medical College, Chennai, Tamil Nadu
*Corresponding Author
Dr Arnab Dey

Abstract

Aim: *Budd–Chiari syndrome, or Hepatic venous outflow obstruction (HVOO) is defined as the obstruction of hepatic venous outflow regardless of its causative mechanism or level of obstruction. Indian studies on BCS date back to the 1970s. It was suggested that in contrast to western world where hepatic vein (HV) obstruction is a commoner IVC obstruction is a commoner in the far east. This study was done to evaluate the aetiology-based prevalence, clinical spectrum with pattern of obstruction, outcome and non-surgical treatment in patients with BCS from southern part of India.*

Methods: *All the consecutive cases of Budd Chiari syndrome (BCS) or Hepatic Vein Outflow Obstruction (HVOO) in the study period from September 2018 to July 2021 were prospectively evaluated. Diagnosis was based on angiographic evidence of HVOTO (i.e., obstruction of IVC and/or HV). All the patients with BCS were subjected to tests available for hypercoagulable state before starting any treatment and if required was confirmed in the first degree relative for any hypercoagulable state. After the confirmation of the diagnosis patients were taken up for appropriate radiological intervention after discussion with the intervention radiology and anaesthesia team.*

Results: *Seventy-eight patients, forty-three female and thirty-five males had radiologically confirmed BCS. The mean age at presentation was 35.86 years (range 10–65 years). Twenty-seven patients had acute presentation and fifty-one had chronic illness. Abdominal pain and jaundice were more common in patients with acute disease than the chronic patients but distended veins were more commonly seen in the later. Right upper quadrant pain or epigastric pain (65.38%), distension of the abdomen (75.64%), pedal oedema (41%), UGIB (24.35%) were the primary symptoms and distended veins (66.67%), hepatomegaly (69.2%) were the most common presenting signs. Hypercoagulable state was the etiology in 14 (18%) patients. 48 (61.53%) patients had both IVC and HV obstruction. Associated portal venous thrombosis was seen in 8 (10.25%) patients. Out of the 27 patients that presented with the acute onset of the disease, 2 (7.4%) patients underwent balloon angioplasty of the IVC obstruction.3 (11.1%) patients underwent angioplasty with stenting and they had pure hepatic vein obstruction. 5 (18.5%) patients underwent TIPS procedure. Out of the 51 patients that presented with chronic disease, two third of the patients had*

radiological features suggestive of established cirrhosis. 12 (23.5%) patients with combined IVC and hepatic vein obstruction underwent angioplasty with stenting.

Conclusion: *Combined obstruction is more common in India in contrast to the west. Hypercoagulable states are a common cause of etiology in Indian patients which was reported earlier in very few studies. Combined IVC and HV obstruction can be treated with angioplasty with stenting as demonstrated in our study which is usually difficult to treat. Significant mortality seen with surgical procedures is not seen in patients treated with non-surgical methods. TIPS has been useful in patients with complete HV obstruction with IVC obstruction.*

Keywords: *Budd Chiari syndrome; HVOO; TIPS in BCS; Angioplasty in BCS; Acute BCS; Chronic BCS.*

Introduction

Budd–Chiari syndrome, or Hepatic venous outflow obstruction (HVOO) is defined as the obstruction of hepatic venous outflow regardless of its causative mechanism or level of obstruction[(1)].This obstruction can be traced to the small hepatic venules up to the entrance of the inferior vein cava (IVC) into the right atrium. The obstruction can extend from the small hepatic venules up to the entrance of the inferior vein cava (IVC) into the right atrium. BCS can be classified as primary when the obstruction originates in the vein and thrombosis is the main cause, or secondary when the vein is externally compressed (abscess, tumour). HVOO is frequently known to cause portal hypertension[(2,3)].

Indian studies on BCS date back to the 1970s[(4–7)] following which various studies were done concentrating on clinical spectrum, aetiology, diagnostic modalities and various treatment strategies.[(2,3,8–12)]. It was suggested that in contrast to western world where hepatic vein (HV) obstruction is a commoner IVC obstruction is a commoner in the far east[(13)].In 8 recent reports from India the commonest pattern obstruction in BCS was combined hepatic vein and IVC obstruction (54%- 64%) in 4 studies[(2,14–16)], isolated hepatic vein obstruction (42%-59%) in 2 studies[(17,18)] and isolated IVC obstruction in 2 studies (54%-82%)[(19,20)]. Tumours, pregnancy, OCP and infections were proposed as predominant underlying causes, hypercoagulable blood dyscrasias were uncommon, and idiopathic cases were the most common[(2,3,7,8,10,21)]. In the studies done during the last 10 years, various hypercoagulable states are being increasingly described as aetiologies of BCS[(15,22–27)].

Majority of patients with HVOO present with a chronic course, while only a small number of patients present with acute or fulminant forms. Abdominal pain and distension are the most common symptoms in patients with HVOO, some also present with upper gastrointestinal bleed (UGIB) and jaundice[(2,3,28)]. The most common signs are tender hepatomegaly, ascites, visible front and back veins and pedal oedema[(2,3,28)]. Mostly the disease is encountered in the adult population and considered uncommon in children; but seen in children, the clinical presentation is somewhat similar[(2)].

BCS should be suspected or discarded in any patient with acute or chronic form of liver disease, because of its heterogeneous clinical presentation especially when the aetiology of the liver disease is unknown and/or if there is an underlying prothrombotic condition[(29)]. Non-invasive imaging techniques (Doppler ultrasound, CT or MRI) forms the mainstay of diagnosis. Doppler USG, performed by an experienced operator, has a sensitivity of >75% and should be the first-choice option[(30)]. Medical management in the form of anti-thrombotic drugs, anticoagulants and diuretics form the mainstay in the treatment of acute and chronic BCS. Radiological intervention in the form of percutaneous balloon angioplasty has also been used successfully in the treatment of BCS. Although TIPS has gained popularity in the treatment of BCS, very few studies have reported its use in these patients[(27,31)]. In this study we evaluated the aetiology-based prevalence, clinical spectrum with pattern of obstruction, outcome and

non-surgical treatment in patients with BCS from southern part of India.

What is already known?

- IVC obstruction was the commonest pattern of BCS in India
- Combined obstruction was reported in few Indian studies with small sample size.
- Ascites, hepatomegaly and abdominal pain – most frequent clinical manifestations
- Acute or sub-acute cases are more common in the west whereas chronic presentation is common in the east.
- Etiology of the obstruction can be identified in minority of the patients in the Indian context and majority are labelled as idiopathic.
- USG with doppler is the first line of investigation.
- MR imaging and CT evaluation are done for diagnostic confirmation.
- Treatment is a stepwise approach consisting of medical management (anticoagulants, diuretics), angioplasty/stenting, TIPS and Liver Transplant.

What is new in this study?

- Combined IVC and Hepatic vein obstruction is the most common pattern in this study.
- Overall Female sex is predominant in the present group of patients and females were more commonly seen in the acute group.
- Acute group of patients were younger than the chronic group probably due to acquired hypercoagulable state in the presence or absence of other autoimmune diseases.
- Pain abdomen, jaundice, high SAAG high protein asciteswas more common in the acute group in the present study.
- Visible veins, esophageal varices, UGIB, Hepatic encephalopathy, pedal edema, splenomegaly and serum albumin < 3 g/dl - more in the chronic group.
- Etiology of the obstruction identified in 40% of the patients, hypercoagulable state is the most common etiology in the present study.
- Antiphospholipid syndrome was most common followed by factor V Leiden mutation in the present study.
- Overlap of HVOTO with PVT seen in 26% of the patients in the acute group in the present study, mostly associated with hypercoagulable state.
- Angioplasty ± stenting is an important modality of treatment in patients presenting with acute clinical manifestations.
- TIPS helpful in complete HV obstruction with IVC obstruction as seen in the present study with promising primary patency rates.

Future clinical and research implications of the study findings

- A multicenter study on a greater number of patients to identify reasons behind female sex predilection and young age patients for acute presentation is required.
- Bone marrow examination in patients with idiopathic causes may generate substantial information regarding the etiology.
- Lastly the role of Liver Transplantation in BCS patients needs to be evaluated further.

Materials and Methods

All the consecutive cases of Budd Chiari syndrome (BCS) or Hepatic Vein Outflow Obstruction (HVOO) in the study period from September 2018 to July 2021 were prospectively evaluated. Diagnosis was based on angiographic evidence of HVOTO (i.e., obstruction of IVC and/or HV). BCS was suspected in the following clinical and biochemical situations: patients with high serum-ascitic fluid albumin gradient (SAAG) (> 1.1) ascites with low cell count (< 250 cells/cm2); patients with ascites and/or visible back veins with or without hepatomegaly and/or

right upper quadrant pain; patients with refractory ascites with unknown aetiology for liver disease , patients with acute liver failure with hepatomegaly and/or ascites; patients with a known hypercoagulable state with evidence of liver involvement (on clinical examination or biochemistry or imaging); and patients with unexplained chronic liver disease (when work-up for alcohol related, drug-induced, viral, metabolic or autoimmune liver disease were ruled out). All these patients were subjected to USG abdomen with doppler study for the status of HV, IVC and splanchnic venous system. If there is evidence of any outflow obstruction then the site and extent of the obstruction was confirmed with CT or MR angiography. All patients with suspected BCS were subjected to chest x-ray, electrocardiogram and 2-dimensional echo of the heart to rule out cardiac aetiology. Liver histology was performed to rule out BCS due to small intrahepatic vein obstruction in the presence of strong clinical suspicion but preserved large veins on imaging. Upper gastrointestinal endoscopy was performed in all cases of BCS to determine the presence of varices.

All the patients with BCS were subjected to tests available for hypercoagulable state before starting any treatment and if required was confirmed in the first degree relative for any hypercoagulable state. All the female patients were tested for pregnancy and were questioned regarding use of oral contraceptive pills.

All the patients were subjected to all baseline blood tests for CBC, renal function, liver function and coagulation parameters. After the confirmation of the diagnosis patients were taken up for appropriate radiological intervention after discussion with the intervention radiology and anaesthesia team. All the patients who underwent radiological intervention were subjected to anticoagulation to maintain the INR within 2-3 in addition to symptomatic therapy. Asymptomatic or mildly symptomatic patients, patients with diuretic-responsive ascites, patients with stable liver function or patients who were not willing to undergo radiological intervention were subjected to anticoagulation to keep INR 2-3 in addition to symptomatic therapy (including low-salt diet, diuretic therapy and/or beta-blockers as needed). All other patients who were not fit for radiological intervention or with worsening liver or renal or coagulation parameters with cirrhosis of liver were referred for liver transplantation after initial stabilisation in the hospital.

Follow-up was done on the 3rd day by the team, at 1 week, at the end of the 1st month, every 3rd month for the 1st year and every 6th month thereafter. During each subsequent follow-up visit, clinical, biochemical and USG with Doppler evaluations were done. Patients with TIPS dysfunctions or post angioplasty re-stenosis or stent occlusion were managed by repeat radiological intervention.

Results

Seventy-eight patients, forty-three female and thirty-five males had radiologically confirmed BCS. The mean age at presentation was 35.86 years (range 10–65 years). Duration of symptoms varied from 15 days to 5 yrs. Twenty-seven patients had acute presentation and fifty-one had chronic illness. 4 patients (5.12%) developed symptoms before the age of 18 years and 1 patient developed the disease during pregnancy. Presenting symptoms were mainly abdominal distension, pain abdomen, pedal oedema and upper gastrointestinal bleeding. Distended veins, hepatomegaly, splenomegaly were the commonest signs.

Clinical Presentation and etiology

Patients presented to the hospital with symptoms of variable duration. 27 (34.61%) patients presented within 6 months of symptoms onset out of which 19 (70.37%) patients presented within 3 months of symptom onset. 51(65.38%) patients had symptom duration of more than 6 months out of which 23 (45.1%) had a symptom duration of more than 1 year and 9 (17.65%) were symptomatic for a period of more than 3 years.

4 (5.12%) patients had the disease before 18 years of age and 7 (8.97%) patients were older than 50 years. Majority of the patients, 68 (87.2%) were of the 18-50 age group.

The mean age of presentation in those with acute presentation (< 6months) was 29.74 years whereas it was 39.44 years in those with chronic (>6 months) disease.

Right upper quadrant pain or epigastric pain (65.38%), distension of the abdomen (75.64%), pedal oedema (41%), UGIB (24.35%) were the primary symptoms and visible veins (66.67%), hepatomegaly (69.2%) were the most common presenting signs (**Table 1**)

Table 1: Clinical Characteristics of the 78 patients of BCS

	Acute (<6months) (n=27)	Chronic (>6months) (n=51)	Total (n=78)
Male:Female	1:2	1.04:1	1:1.2
Mean age ± SD (years)	29.74±21.7	39.44±22.01	35.86±10.72
Age range (years)	10-65	22-60	10-65
Mean duration of symptoms (months)	2.37±1.25	17.87±12.67	12.86±13.46
Duration of symptoms range (months)	0.5-5	6-60	0.5-60
Abdominal pain	25 (92.6%)	26 (50.98%)	51(65.38%)
Distension of abdomen	19 (70.37%)	40 (78.53%)	59 (75.64%)
Jaundice	9 (33.33%)	7 (13.72%)	16 (20.51%)
Oesophageal varices	3 (11.1%)	25 (49%)	28 (36%)
UGIB	0	5 (9.8%)	5 (9.8%)
Fever	9 (33.3%)	8 (15.7%)	17 (21.8%)
Distended veins	10 (37.03%)	42 (82.35%)	52 (66.67%)
Hepatic encephalopathy	1 (3.7%)	15 (29.4%)	16 (20.5%)
Hepatomegaly	21 (77.8%)	33 (64.7%)	54 (69.2%)
Splenomegaly	7 (33.3%)	21 (41.2%)	35 (68.62%)
Oedema	6 (28.7%)	26 (51%)	32 (41%)

Various etiological factors were involved as outlined in **table 2.** IVC membrane was seen in 11 patients, tumour related in 4 and pregnancy related in 2patients. Completethrombophilic workup was done for 44 patients except bone marrow examination. Hypercoagulable state was the etiology in 14 (18%) patients out of which Antiphospholipid antibody syndrome was identified in 5 patients, factor V leiden mutation in 3 patients, 2 patients had Polycythemia vera (JAK 2 positive), Protein C deficiency in 2, protein S deficiency in 1 patient, and 1patient was diagnosed to have hyperhomocysteinemia.

Table 2: Etiology of BCS in relation to duration

Duration(months)	<1 month	1-3 months	4-6 months	7-12 months	13 – 60 months	Total
Cause						
IVC membrane	0	0	3	3	5	11 (14.1%)
Pregnancy	0	2	0	0	0	2 (2.56%)
Tumours	0	1	3	0	0	4 (5.13%)
Hypercoagulable state	3	5	5	1	0	14(18%)
Idiopathic	0	11	1	14	21	47 (60.25%)

11 (14.1%) patients had IVC web or membrane and 2(2.56%) patients had pregnancy related disease. 4(5.13%) patients had HCC related disease and in 60% of the patients the cause of the outflow obstruction could not be identified. Out of 11 patients who had IVC membrane 10 had coexisting HV thrombosis and 1 patient had IVC thrombosis below the membrane.

Site of Obstruction

Table 3 summarises the site of obstruction in the 78 patients. Pure Hepatic vein obstruction was

seen in 11 (14.10%) patients whereas pure IVC obstruction was seen in 19 (24.35%) patients. 48 (61.53%) patients had both IVC and HV obstruction. Associated portal venous thrombosis was seen in 8 (10.25%) patients.

Table 3: Sites of blockage in 78 patients with Budd–Chiari syndrome

Site of obstruction	Acute (N=27)	Chronic (N=51)	Total (N=78)
IVC	2	17	19(24.35%)
HV	8	3	11 (14.10%)
Both IVC & HV	17	31	48 (61.53%)
PVT	7	1	8 (10.25%)

Clinical differences between the Acute and the Chronic cases:

On comparison of the patients with acute and chronic BCS it was seen that most of the patients with the acute disease were below 30 years of age whereas majority of the chronic patients belonged to more than 35 years age group. Abdominal pain and jaundice were more common in patients with acute disease than the chronic patients but distended veins were more commonly seen in the later. Upper GI bleed and hepatic encephalopathy were more commonly seen in the chronic patients. Hepatomegaly was seen with equal frequency in both the group of the patients but splenomegaly and oedema predominated in the chronic patients. Pregnancy was more commonly associated with acute presentation and these patients had combined hepatic vein an IVC obstruction. Hepatic vein and IVC obstruction were found in both the groups with equal frequency. Hypercoagulable state was the predominant etiology in the acute patients whereas tumours and idiopathic cases were prevalent in the chronic patients.

Table 4: Acute and chronic Budd chiari syndrome: clinical and etiologic differences

		Acute (<6m) N= 27	Chronic (>6m) N=51
Male:Female		9:18	26:25
Mean age	p<0.0005	29.74	39.44
Abdominal pain	p<0.005	25 (92.6%)	26 (50.98%)
Distension	p=0.43(NS)	19 (70.37%)	40 (78.53%)
Jaundice	p<0.05	9 (33.33%)	7 (13.72%)
Oesophageal varices	p<0.05	3 (11.1%)	25 (49%)
UGIB (variceal)	p<0.05	0	5 (9.8%)
Fever	p=0.07 (NS)	9 (33.3%)	8 (15.7%)
Visible veins	p<0.005	10 (37.03%)	42 (82.35%)
Hepatic encephalopathy	p <0.05	1 (3.7%)	15 (29.4%)
Hepatomegaly	p =0.23 (NS)	21 (77.8%)	33 (64.7%)
Splenomegaly	p <0.005	7 (33.3%)	35 (68.62%)
Oedema	p <0.05	6 (28.7%)	26 (51%)
S. albumin (<3g/dl)	p= 0.6 (NS)	17 (62.9%)	35 (68.62%)
High SAAG high protein ascites	p<0.005	15 (55.6%)	5(9.8%)
HV block	p=0.09 (NS)	17 (62.9%)	41 (80.39%)
IVC block	p= 0.13 (NS)	21(77.7%)	46 (90.2%)
Pregnancy related	p<0.05	2 (7.4%)	0
Hypercoagulable state	p <0.005	13 (48.14%)	1 (1.96%)
IVC membrane	p=0.5 (NS)	3 (11.1%)	8 (15.69%)
Tumours	p< 0.005	4(14.8%)	0
Idiopathic	p<0.05	12 (44.4%)	35 (68.6%)
Portal venous thrombosis	p<0.005	7 (26%)	1 (2%)

NS = Not significant, SAAG = Serum ascites albumin gradient, UGIB = Upper Gastrointestinal Bleed.

Radiology

Obstruction of the veins was seen in about 90% of the patients with the initial doppler USG although all the patients underwent either CT or MR angio tocorrectly assess the siteand the extent of the obstruction and also to ascertain any associated portal vein thrombosis.

Radiologic features of HV obstruction commonly included visualisation of the occluded veins, endoluminal thrombus, partial narrowing and complete non-visualisation of the veins. Features of IVC obstruction that were commonly seen included IVC membranes or webs, filling defect within the IVC suggesting thrombus, partial/segmental narrowing and complete non-visualisation of the intra or suprahepatic IVC. Intrahepatic comma shaped collaterals were the most common finding in both the groups followed by caudate lobe hypertrophy which was more predominant in the chronic group of patients.

Table 5: Various Radiologic features of obstruction on USG doppler and CT/MR angio (Patients can have more than one finding)

Radiologic features	Pure HV obstruction N=10	Pure IVC obstruction N=19	Both HV and IVC obstruction N=48
Intrahepatic collaterals	9 (90%)	17 (89%)	47 (97%)
Caudate lobe hypertrophy	5 (50%)	15 (79%)	46 (95.83%)
Occluded vein	2 (20%)	-	13 (27%)
Filling defect	2 (20%)	16 (84.2%)	9 (18.75%)
No flow on doppler USG	2 (20%)	-	16 (33.3%)
Membrane	-	1 (5.2%)	10 (20.8%)
Segmental narrowing or stenosis	4 (40%)	10 (52.6%)	35 (73%)
Complete non visualisation	3 (30%)	-	20 (41.7%)

Histopathology

Histopathological features in 29 patients that underwent biopsy (Table 6) revealed established cirrhosis in 16 patients and 9 patient had mixed features of congestion and cirrhosis.

Table 6: Liver Biopsy findings in 29 patients with BCS

	Acute (N=8)	Chronic (N=21)	p value
Necrosis with Centrizonal congestion	2 (25%)	2 (9.52%)	p=0.28 (NS)
Mixed features: congestive and cirrhotic changes	5 (62.5%)	4 (19.04%)	p< 0.05
Established cirrhotic changes	1 (12.5%)	15 (71.42%)	p<0.005

NS – Not significant

Treatment

Various treatment modalities were used for the patients. Diuretics were used for control of ascites. 15 (19.23%) patients underwent endoscopic variceal ligation out of which 5 (6.4%) patients had prior history of upper gastrointestinal bleed. After the initial routine haematological, radiological and etiological evaluation, patients were started on diuretics for control of ascites.

Treatment	Acute (N=27)			Chronic (N=51)		
	IVC	HV	IVC+HV	IVC	HV	IVC+HV
Balloon angioplasty	2 (7.4%)	-	-	14(27.5%)	-	-
Angioplasty with stenting	-	3(11.1%)	-	-	-	12(23.5%)
TIPS	-	-	5(18.5%)	-	-	2 (4%)
Medical management only	-	3(11.1%)	1(3.7%)	3(5.8%)	1(1.96%)	1(1.96%)

Out of the 27 patients that presented with the acute onset of the disease, 2 (7.4%) patients underwent balloon angioplasty of the IVC obstruction. 3 (11.1%) patients underwent angioplasty with stenting and they had pure hepatic vein obstruction. 5 (18.5%) patients underwent TIPS procedure as they had complete occlusion of the hepatic veins with partial narrowing of the IVC. 7 (26%) patients had coexisting portal vein thrombosis who were referred for liver transplantation. 4 (14.8%) patients, 3 (11.1%) with partial hepatic vein obstruction and 1 (3.7%) with combined IVC and hepatic vein obstruction, continued on anticoagulants, improved clinically and were not willing for any further radiological intervention. 6 (14.8%) patients, 4 (14.8%) with hepatocellular carcinoma with combined obstruction and 2 (7.4%) with combined obstruction died during the course of hospital stay due to chronic liver failure.

Out of the 51 patients that presented with chronic disease, two third of the patients had radiological features suggestive of established cirrhosis. 14 (27.5%) patients with pure IVC obstruction underwent balloon angioplasty. 12 (23.5%) patients with combined IVC and hepatic vein obstruction underwent angioplasty with stenting. 2 (4%) patients with combined obstruction underwent IVC angioplasty followed by TIPS. 9 (17.65%) patients who had combined obstruction died during the course of hospital stay due to chronic liver failure. 5(9.8%) patients with stable liver function were treated conservatively with medical management. 9 (17.65%) had cirrhotic liver with deranged liver function, coagulopathy, renal dysfunction was referred for liver transplantation.

Some of the patients in the radiological intervention group suffered complications post procedure. Out of the 15 patients that underwent angioplasty with stenting 1 (6.67%) patient had occlusion of the stent at 5th day post-operative following which balloon dilatation was done. All other patients who underwent angioplasty with stenting (3 with HV and 11 with IVC+HV obstruction) had resolution of ascites and pain abdomen. 1 (6.25%) patient out of the 16 patients that underwent balloon dilatation had HIT (Heparin induced thrombocytopenia) with low molecular weight heparin which was then managed with directly acting oral anticoagulants. Out of the 7 patients that underwent covered TIPS, 1 (14%) patient had stent occlusion at 1 month follow-up which was probably due to the fact that the patient stopped taking the anticoagulants and was lost to follow-up. Balloon dilatation was tried was tried but was unsuccessful in this patient.

Discussion

The etiology and the prevalence of BCS varies in different parts of the world. In the west the site of occlusion is most commonly the hepatic veins[32,33] whereas in Asia it is the IVC or combined obstruction that predominates[34]. Our study is one of the largest studies that has ever been undertaken in southern part of India and is also distinct from other Indian studies. It has showed the predominance of combined IVC and Hepatic vein obstruction with the cause of obstruction identified in around 40% of the patients. We studied a set of 78 patients of hepatic venous outflow tract obstruction (HVOO) over the last 2 years (2019-21). The disease was classified as acute and chronic on the basis of the pattern followed in earlier studies[2,14].

43 (55.12%) females and 35 (44.8%) males consisted of the patients. Female patients predominated in our study as was seen in other studies(31,35,36). Female predominance in BCS patients in the west was primarily due to OCP use and pregnancy but none of our female patients had any history of OCP use and 2 (4.6%) had pregnancy related disease. Around 65% of the patients presented with chronic form of the disease in contrast to western population where majority of the patients present acutely or sub-acutely[28,37,38]. But chronic presentation was more common in Indian patients as was also seen in two previous studies[2,31]. The mean age of

presentation was 35.86±10.76 years, mean age at the acute group was 29.74 years whereas it was 39.44 years in the chronic group. The patients in the acute group were much younger probably because of the fact that these patients had some form of Primary BCS due to hypercoagulable state or pregnancy related. This relation of acute BCS with younger group of patients has not been mentioned in any of the previous studies.

The clinical features at presentation depend on a great extent on the site of obstruction and the rapidity of the evolution of the obstruction[2,3,28,39]. Around 62% of our patients had both IVC and HV obstruction followed by only IVC obstruction in 24% and only hepatic vein obstruction in 14%. Pure IVC obstruction is not commonly seen in the west but quite common in India, South Africa and Nepal[39]. Combined obstruction has been cited as the predominant site in very few Indian studies[8,15,16]. Abdominal pain, distension and pedal oedema were the most common symptoms whereas hepatomegaly, visible veins at the front and the back, splenomegaly were the most common signs all of which has been reported in studies in the past[14,28,38,40–42].

But there were certain clinically significant differences in the acute and chronic group, pain abdomen, jaundice, high SAAG high protein asciteswas more common in the acute group whereas distended veins, esophageal varices, UGIB, Hepatic encephalopathy, pedal edema, splenomegaly and serum albumin < 3 g/dl were more in the chronic group. A previous Indian study on 177 patients[2] cited similar differences but they had greater incidence of abdominal distension and hepatic encephalopathy in the acute group unlike our study.

The etiology of HVOO could be identified in 39.74% of our patients, other studies have reported 30%(14), 35%(2), 42%(8), 53%(43), 66%(4) and 75%(37).Causes in our study included IVC membrane in 11 (14.1%) patients, pregnancy related in 2 (2.56%), HCC related in 4 (5.13%), hypercoagulable state in 14 (18%). In two previous Indian studies[2,8] MOVC and tumor related etiology were more common unlike our study where hypercoagulable causes were more common (18%) followed by IVC membrane. In another study[31] hypercoagulable causes constituted 55% of the etiology. This difference in detection of hypercoagulable states was probably due to the fact that bone marrow examination for myeloproliferative disorders was not done in our patients with idiopathic cases and this is a limitation of our study. Anti-phospholipid antibody in 5 (35.71%), factor V leiden mutation in 3 (21.4%), polycythemia vera in 2 (14.2%), protein c in 2 (14.2%), protein S in 1 (7.14%) and hyperhomocysteinemia in 1 (7.14%) patient. Hypercoagulable state was most commonly seen in the acute group whereas IVC membrane in the chronic group. These findings were consistent with previous Indian study[2]. 26% patients in the acute group had associated non cirrhotic portal vein thrombosis which was reported in very few prior studies[31,44].

Earlier scintigraphy, venography and histopathology were essential for the diagnosis of HVOO[4,43,45] but recently various studies have shown that doppler ultrasonography[36,46,47], CT[48] and MRI[49] are also useful. USG identified the obstruction in 90% of our patients which was confirmed by CT or MR angiography.

HVOO leads to characteristic histological changes in the liver. The acute form of the disease is characterized by centrizonal necrosis and congestive features with normal periportal area but with progression of the disease the chronic form is dominated by features of portal hypertension and histologically suggestive of cirrhosis[2]. Histological evidence of cirrhosis was seen in 71% of our chronic cases but overlapping features of congestion, necrosis and cirrhosis was more commonly seen in the acute patients, 62.5% and the difference was clinically significant. The findings are consistent with other studies[2,8,14] but characteristic feature of centrizonal necrosis and congestion for acute BCS was not seen in acute group of our patients unlike prior studies[2,14] and this was due to the fact that most of these patients

probably had prior asymptomatic or less troublesome features of HVOO.

Patients with HVOO have poor prognosis if untreated, two studies have reported mortality of 40% at 1 year and 90% at 5 years[37,50]. Few studies[51–53] have shown effectiveness of only medical management in a small group of HVOO patients with diuretic responsive ascites, stable renal and liver function and mildly symptomatic presentation.

Various studies have shown the poor results of surgery in these patients, low success rate with anatomical difficulties, high post-operative complication and mortality rates[54,55]. In the Indian context two studies[56,57] had shown fair results for only IVC obstruction but the results of surgery in HV obstruction were not encouraging[3,58]. This was the reason why we didn't opt for surgical shunts for our patients.

Studies have shown the success rate of radiological intervention for IVC and HV in the range of more than 90% and patency rates of more than 80% with reintervention also successful [13,59–61]. Previous studies from India have shown the efficacy of balloon angioplasty with or without stenting for IVC obstruction, and a restenosis rate of 17% to 29%[2,3,9,19,57,62]. Use Balloon angioplasty for HV obstruction throughout the world has shown variable results[54,60] but in India the experience with angioplasty for HV thrombosis is scarce[3,62]. Combined IVC-HV obstruction, a difficult to treat entity, were treated with balloon angioplasty followed by surgical portosystemic shunting in previous Indian studies, but it was associated prominent post-operative complications and mortality[3]. A two-stage process for treating combined obstruction was studied in China[62] where IVC stenting was done first followed by another session of HV stenting. In our group of patients balloon angioplasty of the IVC followed by TIPS or HV angioplasty with stenting was done in a single session. 40% of our patients underwent angioplasty ± stenting.

In the recent times TIPS for BCS patients has been widely described in the worldwide literature[63–71]. Since the 90s, surgical shunts have been replaced by the less-invasive TIPS in most places. TIPS has been demonstrated to be more effective in maintaining patency and is associated with lower morbidity and mortality than surgery in patients with failure on medical treatment or when recanalization has failed[30,72]. TIPS primary patency rate using PTFE-covered stents is 67% at 2-year follow-up[73]. Some studies have mention the success rate for TIPS to be around 75-100%[63,66].Complications related to TIPS occur in less than 20% of cases[74]. TIPS dysfunction is lower for covered TIPS (33% vs 87% for uncovered TIPS) and also has higher patency rate (67% vs 19% in uncovered group) [1]. In our study TIPS was the primary treatment for patients with complete HV obstruction with combined IVC obstruction. 7 patients underwent TIPS in our study and the stent re-occlusion was seen only in 1 (14%) patient. This patient was lost to follow-up briefly due to the covid pandemic and she was on irregular medications following the procedure. The primary patency rate of TIPS was 85% in our study.

Our study, although with a small group of patients, showed promising results with angioplasty ± stenting for combined IVC + partial/segmental HV obstruction and TIPS as the primary modality for combined IVC + complete HV obstruction in BCS patients.

Conflict of Interest

The authors who have taken part in this study declared that they do not have anything to disclose regarding funding or conflict of interest in respect to this manuscript.

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