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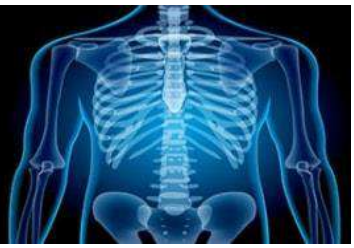

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Role of non-invasive imaging in budd-chiari syndrome

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Abstract
Background and Objectives: Symptoms of Budd-Chiari syndrome (BCS) are often nonspecific or even absent; as a result radiographic imaging plays a critical role in diagnosis. The diagnosis is profoundly dependent on modern radiological imaging techniques, depending on the detection of thrombosis and/or stenosis of hepatic veins or the upper portion of the inferior vena cava. This study was hence conducted to evaluate the role of imaging in BCS.
Materials and Methods: This prospective study was conducted in 40 patients diagnosed with BCS over a period of three years. Patients with history of frequent abdominal pain, ascites, hepatomegaly, fever, hepatic encephalopathy and variceal bleeding were included. All patients underwent imaging tests like ultrasonography, or computed tomography scan, and magnetic resonance imaging (MRI).
Results: A total of 40 patients, with an average age of 35.13 ± 15.19 years, were included in our study. There was no significant difference in the size of liver and size of spleen estimated by the imaging techniques; however, the type of liver margin was better identified by the MRI. There was no significant difference in the caudate lobe-to-right lobe ratio, distribution of enhancement patterns, intrahepatic collaterals, inferior vena cava parameters, and ultrasonography spectral findings of the three hepatic veins between the modalities.MR venography was found to be better at identifying the different patterns in the right, middle and left hepatic veins.
Conclusions*:* Not only does MRI identify BCS stages and potential complications, these findings are diagnosed with an added advantage of no radiation exposure and reproducibility of findings.

Keywords: Budd-Chiari Syndrome, magnetic resonance imaging, ultrasonography

Introduction
Budd–Chiari syndrome (BCS), a rare clinical entity, is defined as hepatic venous outflow obstruction at any level ranging from small hepatic veins (HV) to junction of the inferior vena cava (IVC) and the right atrium, irrespective of the cause of obstruction [1]. Laboratory investigations can identify the degree of liver injury, with decreased levels of serum albumin, hepatic cytolysis, altered coagulation, and cholestasis, whereas imaging studies may detect venous obstruction [2]. Since symptoms are often nonspecific or even absent, radiographic imaging plays a critical role in diagnosis [3]. Confirmation of the diagnosis is established with the help of unequivocal radiological confirmation of hepatic venous outflow obstruction [1].
Findings of imaging modalities can be categorized as direct and indirect signs. Direct signs are the hallmarks of BCS and consist of visualization of the upper portion of the IVC or obstructive lesions of the HV. (Figure 1, figure 3a and 3b) Indirect signs, secondary to venous obstruction, relate to signs of portal hypertension, dysmorphy, intra- and extrahepatic collateral circulation and perfusion abnormalities [4, 5]. (Figure 2). Understanding of these findings is vital for early diagnosis and appropriate treatment [6].
Abdominal ultrasonography (USG) is the first step in BCS imaging assessment [6], with an accuracy of up to 85% [7]. Patients with BCS manifest stigmata of portal hypertension, like dilatation of the spleeno-portal venous axis, abdominal varices, and splenic enlargement, despite the normal contour, size, and echotexture of the liver [6]. Absent or reversed flow in the HV and/or flat flow in the HV along with reversed flow in the inferior vena cava may be considered diagnostic for BCS [7]. Compared to other imaging techniques, a significant advantage of the Doppler ultrasound is its ability to determine the flow direction as a sign of an obstruction or thrombosis [8]. In the initial stage, venous congestion predominates, with ultrasonography findings convening a hyperechoic appearance of the liver, appearing

heterogeneous and hypoattenuating on computed tomography (CT), while magnetic resonance imaging (MRI) shows a variable appearance of the liver; a constant finding includes a striking difference in signal intensity between the peripheral and central zones [4, 9].

There is a need to bridge the lacuna in existing knowledge about the subject and improve patient care. The present study was hence conducted to evaluate the role of USG, CT and MRI in Budd-Chiari syndrome.

Materials and Methods

This prospective study was conducted between May 2017 and May 2020 at a tertiary hospital in Western India. The study was approved by the Institutional Ethics Committee and complied with local practice guidelines.

Patients

Patients attending the emergency or out-patient departments with history of frequent abdominal pain, ascites, hepatomegaly, fever, hepatic encephalopathy and variceal bleeding and clinically suspected for BCS and diagnosed using screening imaging investigations/ biochemical parameters suggestive of cirrhosis were included. While patients with renal dysfunction or end-stage renal disease (ESRD), claustrophobia, pregnant females, end stage renal failure (for contrast-enhanced CT [CECT]), known history of contrast allergy or those with any electrically, magnetically or mechanically activated implants (e.g. pacemaker, cochlear implant for MRI) were excluded. Only patients willing to provide written informed consent were included in this study.

A set departmental protocol for investigation of patients referred with chronic liver parenchymal disease and/or suspected cases of BCS was used. All patients were assessed for demographics, history, clinical examination and laboratory test findings. After placement of intravenous cannula, patients were prepared for radiological procedures.

Imaging

The USG was carried out using either Samsung HS50, GE LOGIC V3 series or Toshiba Xario Xg prime. With a proper supine position and the patient's abdomen was scanned with use of anterior, lateral decubitus and subcoastal views. Ultrasonography parameters used were gray scale mode, colour and power Doppler, spectral Doppler and contrast ultrasound with curvilinear and linear probe, wherever available.

After the USG, patients underwent a CECT triple phase scan using a Phillips Brilliance 64 slice CT scanner. Patient were kept orally nil 4 hours prior to the scan to avoid contrast complications. Routine anteroposterior topogram of the abdomen was initially taken, after which axial plain sections of 5 mm thickness were taken from the lung base to the ischial tuberosities. A plain scan was followed by an intravenous contrast scan. The scan was divided into five phases *viz*. plain (0 sec), arterial (22 sec), portal (45 sec), venous phase (60 sec) and a delayed scan at the end of 7 mins. All scans were performed at 120 KVP, 250 mAS and collimation of 64 x 0.625 with rotation time of 0.75 sec and scan FOV of 400. Post-study reconstructions were done at 2.5 mm; sagittal and coronal reconstructions were made wherever necessary. Multislice CT included curved planar reformatting, volume rendering, Maximum and Minimum Intensity Projections, as and when necessary. The magnification mode was commonly employed, and the scans were reviewed on a direct display console at multiple window settings (i.e. abdomen window at 320/40; Lung window 1400/-600; Bone window of 2400/200).

All patients were then planned for MRI abdomen on the consecutive day, and was carried out on a Philips Achieva 3.0T MRI scanner. Scans were done with the patient in supine position following the MRI abdomen protocol. Breath-holding was required in few sequences. Gadolinium based-contrast (Gadobenate) at 0.1 mmol/kg was injected at the rate of 2.5 ml/sec followed by saline flush. The MRI sequences performed were Survey in three orthogonal planes, T2W TSE (axial and coronal), T2W TSE SPAIR (axial and coronal), T1W TFE axial, Dual FFE breath hold (axial), etc.

Statistical analysis

Qualitative data were represented as percentages, and quantitative data using mean ± SD. Spearman or Pearson's correlation coefficient was performed to assess correlation between parameters; p-value <0.05 was considered statistically significant. MS Office, SPSS (v 9.4) and GraphPad software's were used for the analysis.

Results

Based on the study selection criteria, we included a total of 40 patients. The baseline characteristics of the study patients are summarised in Table 1. None of our patients had a family history of BCS.

Liver

The size of the liver was compared using the three different modalities. The USG and colour Doppler showed a size of 13.78 ± 2.67 cm (range: 8-17.5 cm), while the triple phase CECT showed 14.02 ± 2.72 cm (range: 8.4-19 cm), and the MR venography (MRV) showed 14.17 ± 2.70 cm (range: 8.8 – 19 cm). There was no statistically significant difference in the mean size of liver among the three techniques. We found a statistically significant difference in the findings of the margin of liver, while there were no significant differences in focal lesions in liver, intrahepatic collaterals and IVC parameters using the different imaging modalities (Table 2).

There was no statistically significant difference in the mean CRL ratio (0.73 ± 0.12; range, 0.48-1.11) between the triple phase CECT and MRV. The enhancement pattern using the triple phase CECT was heterogeneous in 82.5% patients, flip flop in 10% patients, mottled in 2.5% patients and transient hepatic attenuation differences (THAD) in 5% patients, while using the MRV it was heterogeneous in 85% patients, flip flop in 10% patients, and mottled and THAD in 2.5% patients each, with no significant difference compared to CECT. The caudate lobe was found to be hypertrophic in all patients using the USG Doppler and the MRV.

Findings of the different hepatic veins, as visualised by the different imaging techniques, were found to be statistically significant (Table 3).

The USG spectral findings of the right HV showed triphasic waveform in 17.5% patients, loss of triphasic waveform in 12.5% patients, and thrombosis in 70% patients. In the middle HV, triphasic waveform was noted in 15% patients, loss of triphasic waveform in 25% patients, and thrombosis in 60% patients. In the left HV, triphasic waveform was

noted in 20% patients, loss of triphasic waveform in 15% patients, and thrombosis in 65% patients. The portal vein showed no statistically significant difference in identifying normal (82.5%) and thrombosed (17.5%) veins using the three modalities.

Spleen

There was no statistically significant difference in the mean size of spleen using the USG (14.86 ± 2.33 cm; range: 11-20 cm), the triple phase CECT (15.04 ± 2.31 cm; range: 10.5-20.3 cm), and the MRV (15.08 ± 2.31 cm, range: 10.5–20.5 cm). Findings relating to different parameters related to the spleen using the different modalities are summarised in Table 4.

Discussion

Budd Chiari Syndrome can be diagnosed by ultrasound angiography to look for hepatic sinusoids and MRI of liver with hepatic veins and IVC [10]. The primary diagnostic finding on Doppler ultrasound is the detection of lack of blood flow or thrombus within the hepatic veins. Imaging using the CT forms the backbone of the radiological diagnosis of BCS. Nonetheless, the features of BCS on CT imaging vary based on the severity of the condition. Though relatively invasive, venography is still considered the "gold standard" imaging modality in confirming the diagnosis of BCS [9].

The mean age of patients with BCS have been variable in similar studies and ranged from 29.5 to 46 years [11, 12, 13]. We noted 50% of males to have BCS, however, most researchers have noted a higher prevalence of BCS among males [11, 12]. Dilawari JB *et al.* reported abdominal distension, epigastric or hypochondriac pain, presence of distended abdominal veins, upper gastrointestinal bleeding, jaundice, and hepatic encephalopathy as the most common complaints of BCS [11].

We found MRV to be better at identifying the margin of liver than the USG and the triple phase CECT. (figure 4a and 4b). There was no significant difference in the different enhancement patterns identified by MRV or CECT. In terms of focal lesions in the liver, we found 20% regenerative nodules, 7.5% confluent hepatic fibrosis, 5% hepatocellular carcinoma and 5% other lesions using MRV. Dilawari JB *et al*. recorded a total of 5.08% patients with hepatocellular carcinoma [11].

Using the different imaging modalities, there was no statistically significant difference in the presence of intrahepatic collaterals, or in the presence of thrombi in the IVC, IVC narrowing and normal IVC in our study. Dilawari JB *et al*. recorded IVC block in 75.32% patients [11]. Among the 162 patients analysed by Cheng D *et al*., 34% had IVC stenosis while the majority 66% had IVC occlusion. Cheng D *et al*. recorded right HV patency in 27.2% patients, right HV stenosis in 11.1% patients, right HV occlusion in 47.5% patients, middle HV patency in 19.8% patients, middle HV stenosis in 6.8% patients, middle HV occlusion in 42.6% patients, left HV patency in 21.6% patients, left HV stenosis in 9.3% patients, and left HV occlusion in 38.9% patients. Cheng D *et al*. reported 60.5% patients with venous collaterals formed between the hepatic veins [13].

Radiologic features of HV block noted by Dilawari JB *et al.* included partial obliteration, ostial narrowing and complete non-visualization [11]. In the study by Dridi M *et al*., sonographic features of HV involvement included partial or complete inability to see the hepatic veins, stenosis with proximal dilatation, intraluminal echogenicity, thickened walls, and thrombosis. Abdominal CT showed an enlarged segment I, and heterogeneous enhancement mosaic in certain patients; segment I was typically enhanced in all patients [12]. Dilawari JB *et al*. visualized the portal vein in 98.87% of their study patients [11].

The spleen in BCS gets massively enlarged at portal pressures comparable to other disorders of portal hypertension. Large spleen may display multiple Gamna–Gandy bodies on CT or MRI [6]. Among the patients studied by Dilawari JB *et al*., the spleen was enlarged in 46% patients. Dilawari JB and associates observed the presence of ascites in 74.01% of their study population [11]. while Cheng D *et al*. noted around 55.4% patients to have some amount of ascites, 28.9% had small amounts, 12.1% with moderate amount, and 15.2% with large amounts of ascites [13].

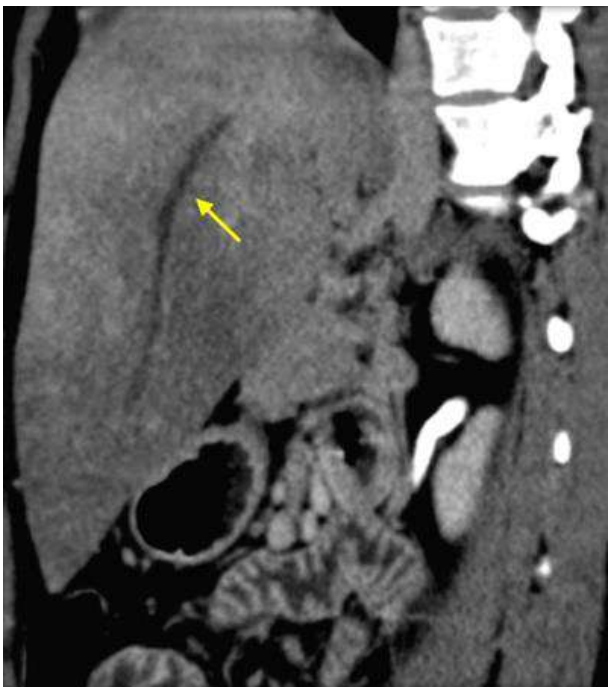

Fig 1: Sagittal CECT abdomen showing thrombosis of right hepatic vein. (As shown by yellow arrow).

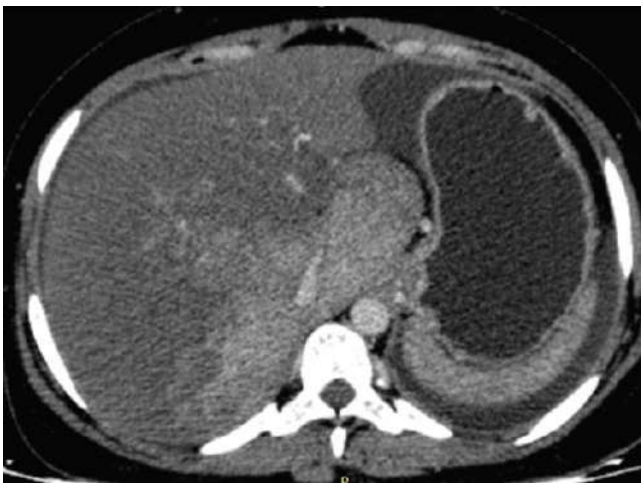

Fig 2: Axial CECT abdomen image showing hypoattenuation of the peripheral liver parenchyma and inhomogeneous enhancement of the central part of liver.

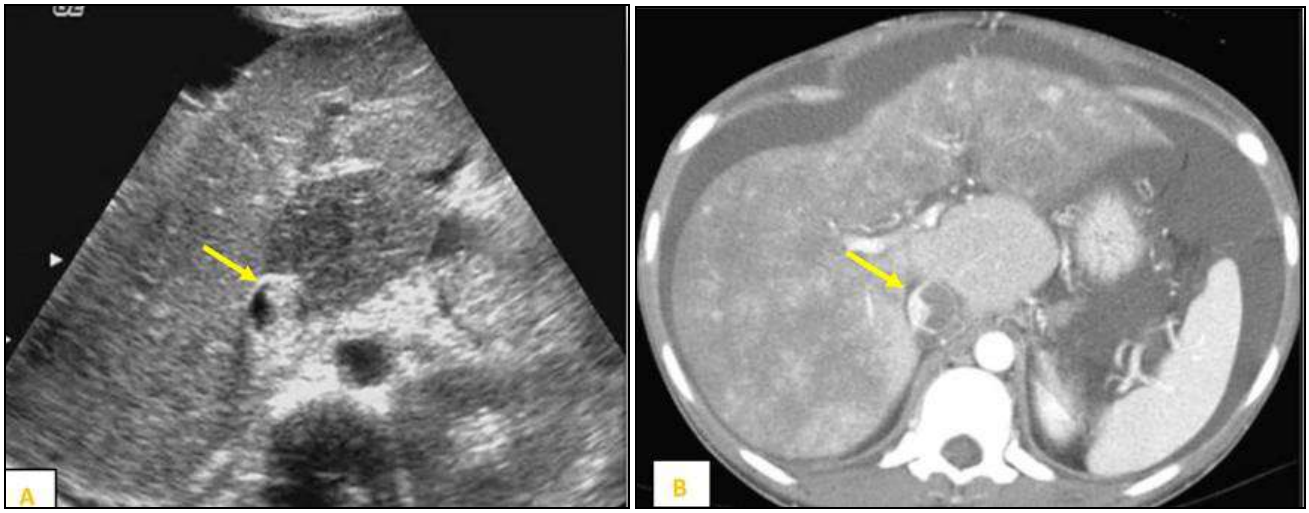

Fig 3ab: USG and CECT venous phase of a patient showing acute thrombosis of intrahepatic IVC with distal narrowing (yellow arrow) associated with caudate lobe hypertrophy (CL) and mottled post-contrast enhancement.

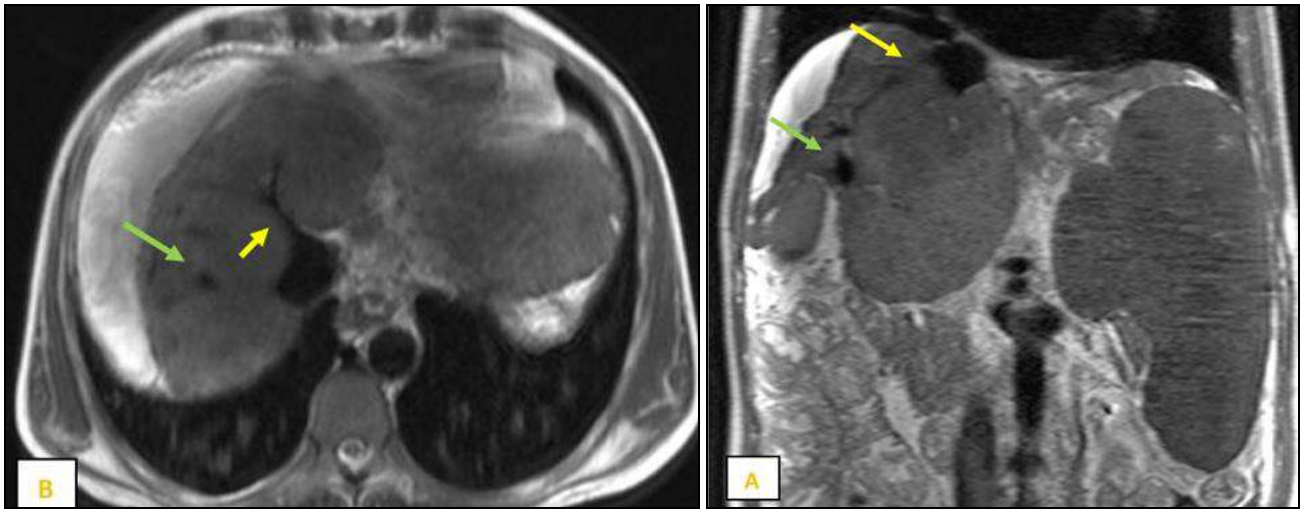

Fig 4ab: T2WI coronal and axial MRI images showing ostial stenosis of right hepatic vein (Yellow arrow), common channel of middle hepatic vein and left hepatic vein and intrahepatic collateral (Green arrow) and splenomegaly is also noted.

Table 1: Baseline characteristics of the study population

Baseline characteristics	Value
Age, mean ± SD (years)	35.13 ± 15.19
***Gender*, n (%)**	
Males	20 (50%)
Females	20 (50%)
Medical history, n (%)	
Abdominal distension	40 (100%)
Abdominal pain	40 (100%)
Upper gastrointestinal bleeding	18 (45%)
Jaundice	32 (80%)
Dilated vein over abdomen	28 (70%)
Hepatic encephalopathy	1 (2.5%)
Positive HbsAg	3 (7.5%)
HCV	3 (7.5%)
JAK-STAT mutation	1 (2.5%)
MTHFR mutation	2 (5%)

Abbreviations: HbsAg, Hepatitis B surface antigen; HCV, Hepatitis C virus; JAK-STAT, Janus kinase–signal transducer and activator of transcription; MTHFR, Methylenetetrahydrofolate reductase; SD, standard deviation

Table 2: Findings relating to different liver parameters using the different modalities.

Parameters	USG & colour Doppler	CECT	MRI
Margin of liver			
Irregular	2 (5%)	12 (30%)	18 (45%)
Irregular, nodular	38 (95%)	28 (70%)	22 (55%)
Focal lesions in liver			
Regenerative nodules	6 (15%)	7 (17.5%)	8 (20%)
Hepatocellular carcinoma	2 (5%)	2 (5%)	2 (5%)
Confluent hepatic fibrosis	0	2 (5%)	3 (7.5%)
Others	2 (5%)	4 (10%)	5 (12.5%)
Intrahepatic collaterals			
Present	24 (60%)	29 (72.5%)	29 (72.5%)
Absent	16 (40%)	11 (27.5%)	11 (27.5%)
IVC parameters			
Thrombus	3 (7.5%)	3 (7.5%)	4 (10%)
Narrowing	28 (70%)	24 (60%)	24 (60%)
Normal	11 (27.5%)	13 (32.5%)	13 (32.5%)

Abbreviations: CECT, contrast-enhanced computed tomography; MRI, magnetic resonance imaging; USG, ultrasonography

Table 3: Findings of the different hepatic veins using the different techniques.

Parameters	USG & colour Doppler	CECT	MRI	p-value*
Right hepatic vein				
Thrombus	7 (17.5%)	19 (47.5%)	22 (55%)	0.0092
Cord-like	15 (37.5%)	6 (15%)	5 (12.5%)	
Fibrosis	9 (22.5%)	3 (7.5%)	1 (2.5%)	
Narrowing	2 (5%)	4 (10%)	2 (5%)	
Ostial stenosis	4 (10%)	5 (12.5%)	5 (12.5%)	
Normal	3 (7.5%)	3 (7.5%)	5 (12.5%)	
Middle hepatic vein				
Thrombus	3 (7.5%)	15 (37.5%)	16 (40%)	0.0126
Cord-like	13 (32.5%)	6 (15%)	5 (12.5%)	
Fibrosis	11 (27.5%)	8 (20%)	4 (10%)	
Narrowing	4 (10%)	3 (7.5%)	5 (12.5%)	
Ostial stenosis	6 (15%)	4 (10%)	3 (7.5%)	
Normal	3 (7.5%)	4 (10%)	8 (20%)	
Left hepatic vein				
Thrombus	3 (7.5%)	15 (37.5%)	16 (40%)	0.0261
Cord-like	12 (30%)	6 (15%)	4 (10%)	
Fibrosis	12 (30%)	5 (12.5%)	2 (5%)	
Narrowing	1 (2.5%)	2 (5%)	5 (12.5%)	
Ostial stenosis	8 (20%)	6 (15%)	7 (17.5%)	
Normal	4 (10%)	6 (15%)	6 (15%)	

Abbreviations: CECT, contrast-enhanced computed tomography; MRI, magnetic resonance imaging; USG, ultrasonography

Table 4: Findings relating to different spleen parameters using the different modalities.

Parameters	USG & colour Doppler	CECT	MRI
Focal lesions in spleen			
Gamma gandy bodies	10 (25%)	12 (30%)	11 (27.5%)
Infarct	2 (5%)	8 (20%)	8 (20%)
Hemangioma	1 (2.5%)	1 (2.5%)	1 (2.5%)
Splenic vein			
Normal	37 (92.5%)	37 (92.5%)	37 (92.5%)
Thrombosed	3 (7.5%)	3 (7.5%)	3 (7.5%)
Ascitis			
Mild	28 (70%)	24 (60%)	26 (65%)
Moderate	10 (25%)	12 (30%)	11 (27.5%)
Severe	2 (5%)	4 (10%)	3 (7.5%)
Bowel loops			
Normal	31 (77.5%)	27 (67.5%)	29 (72.5%)
Colopathy	9 (22.5%)	13 (32.5%)	11 (27.5%)
Collaterals			
Present	38 (95%)	38 (95%)	38 (95%)
Absent	2 (5%)	2 (5%)	2 (5%)

Abbreviations: CECT, contrast-enhanced computed tomography; MRI, magnetic resonance imaging; USG, ultrasonography

Conclusions

Diagnosis of BCS is profoundly dependent on modern radiological imaging techniques, depending on the detection of thrombosis and/or stenosis of hepatic veins or the upper portion of the IVC. The multidetector computed tomography triple phase and MRI are ultimately needed to reach the final diagnosis as well as to plan the further management of these patients. CT helps in aiding the diagnosis as it shows focal liver lesions and help characterize the BCS into acute, subacute and chronic types. The additional findings like presence or absence of ascitis, collaterals and the status of spleen are easily assessed by CT increased sensitivity and specificity.

Before planning the final treatment plan, MRI has now investigation of choice for BCS as it is possible with MRI to scan non-invasively and acquire the minute details about the exact status of hepatic veins and IVC as well as indirect signs of BCS besides confirmation of the stage of this syndrome over the invasive modality. The underlying focal liver lesions can be picked up with higher sensitivity and specificity. Not only does MRI identify BCS stage and potential complications, these findings are diagnosed with an added advantage of no radiation exposure and reproducibility of findings.

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Conflicts of Interest: None.

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