BJR

https://doi.org/10.1259/bjr.20180441

Received: 16 May 2018 | Revised: 13 June 2018 | Accepted: 09 July 2018

Cite this article as:
Bansal V, Gupta P, Sinha S, Dhaka N, Kalra N, Vijayvergiya R, et al. Budd-Chiari syndrome: imaging review. *Br J Radiol* 2018; **91**: 20180441.

PICTORIAL REVIEW

Budd-Chiari syndrome: imaging review

[1]VARUN BANSAL, MD, [2]PANKAJ GUPTA, MD, [2]SAROJ SINHA, MD, DM, [2]NARENDER DHAKA, MD, DM, [1]NAVEEN KALRA, MD, [3]RAJESH VIJAYVERGIYA, MD, DM, [2]USHA DUTTA, MD, DM and [2]RAKESH KOCHHAR, MD, DM

[1]Department of Radiodiagnosis and Imaging, Postgraduate Institute ofMedical Imaging and Research (PGIMER), Chandigarh, India
[2]Deaapartment of Gastroenterology, Postgraduate Institue of Medical Education and Research, Chandigarh, India
[3]Department of Cardiology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Address correspondence to: Dr Pankaj Gupta
E-mail: *pankajgupta959@gmail.com*

The authors Varun Bansal and Pankaj Gupta contributed equally to the work.

ABSTRACT

Budd-Chiari syndrome (BCS), also known as hepatic venous outflow tract obstruction includes a group of conditions characterized by obstruction to the outflow of blood from the liver secondary to involvement of one or more hepatic veins (HVs), inferior vena cava (IVC) or the right atrium. There are a number of conditions that lead to BCS-ranging from hypercoagulable states to malignancies. In up to 25% patients, no underlying disorder is identified. Diagnosis of BCS is based on a combination of clinical and imaging features. A major part of the literature in BCS has been devoted to interventions; however, a detailed description of various imaging manifestations of BCS is lacking. In this review, we highlight the importance of various imaging modalities in the diagnosis of BCS.

INTRODUCTION

Budd-Chiari syndrome (BCS) comprises a heterogeneous group of conditions characterized by partial or complete hepatic venous outflow obstruction.[1–3] There is an increase in hepatic sinusoidal pressure secondary to hepatic venous outflow obstruction. This results in portal hypertension and liver congestion. The latter leads to hypoxia and hepatocyte dysfunction.[2,3] If the obstruction is severe and not corrected timely, hepatocyte necrosis sets in. This finally progresses to hepatic fibrosis and cirrhosis.[2,3]

There are several causes of BCS. The most common condition is an underlying hypercoagulable or a prothrombotic state.[4–6] These include various congenital conditions (including protein C deficiency, protein S deficiency, antithrombin deficiency, factor V Leiden mutation), myeloproliferative disorders, paroxysmal nocturnal hemoglobinuria, pregnancy, oral contraceptive intake, antiphospholipid antibody, malignancy, and few other conditions.[4–6] Secondary BCS comprises of the conditions like the extension of thrombus from the renal cell carcinoma or hepatocellular carcinoma as well as a direct mass effect by liver lesions including liver tumors and abscesses. No underlying condition is found in up to 25% patients.[6] However, recent literature reports an identifiable cause in 90% patients due to the availability of testing for a greater number of gene mutations.[4]

The clinical presentation ranges from mild symptoms to fulminant liver failure with a majority of patients manifesting as chronic liver disease.[7,8] The type of clinical presentation is determined by the extent and acuteness of venous obstruction and the adequacy of the patent primary and collateral venous outflow pathways.[9,10] Imaging evaluations play an important role in the diagnosis and classification of the disease.[11] This is particularly important as the presentation in some patients may be non-specific and imaging may provide the first clue to the underlying condition. The management in BCS is heavily dependent on the imaging findings.[11] Besides, imaging plays a very crucial role of follow up in patients following shunt procedures as well as those on medical management.[12]

CLASSIFICATION OF BUDD-CHIARI SYNDROME

BCS is classified based on the etiology, the site of obstruction as well as the duration of disease. According to etiology, it is classified into primary (obstruction due to endoluminal venous lesion-thrombosis, webs, endophlebitis) and secondary (lesion outside venous system-tumor, abscess, cyst). Two classifications have been proposed based on the site of obstruction.[7,9]

Several investigators have classified BCS into fulminant, acute, subacute, and chronic; however, others tend to divide

Table 1. Three types of BCS based on the level of obstruction[3]

Type	Level of obstruction
I	Obstruction of IVC with or without secondary hepatic vein occlusion
II	Obstruction of major hepatic veins
III	Obstruction of the small centrilobular venules (considered by some as veno-occlusive disease)

BCS, Budd-Chiari syndrome; IVC, inferior vena cava.

them into acute, subacute, and chronic.[7,9] Differentiation of acute, subacute, and chronic is mainly based on clinical features.

Tables 1–3 highlight the types of BCS.

Imaging features of BCS

Imaging features are mainly determined by the duration of disease, the number of hepatic veins (HVs) involved, and the degree of obstruction.[13–17]

The imaging features of different stages of BCS are illustrated in Figures 1–8.

Acute BCS

In the acute form of BCS, patients present with a rapid development of upper abdominal pain, abdominal distension secondary to ascites and liver enlargement, renal failure, and jaundice.[8,18] The imaging features of acute BCS are discussed below and highlighted in Table 4.

Parenchymal changes

As a result of acute obstruction of hepatic venous outflow (attenuation/ thrombosis of HVs and/or IVC), there is hepatomegaly and ascites. Venous thrombosis leads to an elevation of sinusoidal pressure and delay or reversal of portal venous blood inflow. In the acute form, venous collaterals have not developed and hepatocellular necrosis occurs rapidly.[11,13–17] The liver becomes non-homogeneous with delayed enhancement of periphery. The peripheral zones of the liver usually appear hypodense on contrast enhanced CT and hypointense on contrast-enhanced MRI in the arterial phase, which results from the increased post-sinusoidal pressure produced by hepatic venous obstruction. Caudate lobe demonstrates increased contrast enhancement compared with the remainder of the liver.[18–20] The sparing of caudate lobe is explained by its separate venous drainage. In the portal venous phase, a "flip-flop" pattern ensues, with low attenuation of the central part of the liver because of washout, while attenuation in the peripheral part of the liver gradually increases with the accumulation of contrast material from capsular veins.[14,15] The caudate lobe enlargement is seen in 80–91% of all cases.[16,17]

Table 2. Four types of BCS based on the level of obstruction[4]

Type	Level of obstruction
I	Hepatic vein obstruction or thrombosis without IVC obstruction or compression
II	Hepatic vein obstruction or thrombosis with IVC obstruction or thrombosis
III	Isolated hepatic venous webs
IV	Isolated IVC webs

BCS, Budd-Chiari syndrome; IVC, inferior vena cava.

Vascular changes

A recent thrombus is hypoechoic on ultrasound and leads to expansion of the lumen of the involved HVs.[17] It produces signal void on Doppler ultrasound. Over a few days, the thrombus becomes echogenic with retraction of the lumen of the involved hepatic veins.[17,21] On CT and MRI, filling defect is seen within the HVs seen as hypodensity (on CT) or hypointensity (on MRI).[21–23] Stenosis is best detected with Doppler ultrasound as it allows interrogation of the hemodynamic effects of the stenosis in the form of color aliasing and a local increase in flow velocity.[17] Directional alterations in flow may also be detected with Doppler. These changes are difficult to detect on CT or MRI.[17] The vascular changes in acute BCS are discussed in detail in Table 5.

Subacute BCS

It is the most common clinical type of BCS. This form has an insidious onset and may take up to 3 months to become symptomatic.[5,6,24,25]

Imaging features are characterized by volume redistribution as well as the development of collateral vessels.[10–14] These changes overlap with those of chronic BCS and distinction between the two may not be possible based solely on imaging and clinical presentation needs consideration. These changes are discussed in detail in the section below and Table 6.

Chronic BCS

Parenchymal changes

In the chronic BCS there is bridging fibrosis from HV to portal tracts.[26] In the chronic stage of BCS, there are features of chronic liver parenchymal disease. Parenchymal edema which was a prominent feature in the acute and subacute stage is not a prominent feature of chronic BCS.[19,25] Differential attenuation (on CT)/signal intensities (on MRI) observed in patients with acute and subacute stages of disease are more subtle in chronic stage. Differential enhancement pattern between center and periphery of the liver is also more subtle. Caudate lobe is markedly enlarged in chronic disease. Ascites, and collateral vessels are other ancillary findings (may also be seen in subacute stage) of chronic BCS.[16,17]

Another typical feature is the development of regenerative nodules. These are usually multiple and range in diameter from 0.5 to 4 cm. Pathogenesis involves insufficient blood supply to the hepatocytes leading to atrophy. Compensatory nodular hyperplasia occurs in areas where blood supply is adequate.[27] These regenerative nodules are predominantly supplied by arteries and hence show marked arterial hyperenhacement and may be sometimes difficult to differentiate from adenoma.[28] Most of the lesions are likely to be reactive than neoplastic.[29] Hepatocellular carcinoma can develop in patients with chronic BCS and accounts for 0.7% of all hepatocellular carcinoma.[30]

Table 3. Classification of BCS according to the duration of disease[4,5]

Type	Duration
Fulminant	Present with hepatic encephalopathy within 8 weeks of development of jaundice
Acute	Short duration (<1 month), ascites, hepatic necrosis without formation of venous collaterals
Subacute	Insidious onset (1–6 months), ascites, minimal hepatic necrosis, and portal and hepatic venous collaterals
Chronic	(>6 months) Complications of cirrhosis in addition to findings in the subacute form

BCS, Budd-Chiari syndrome.

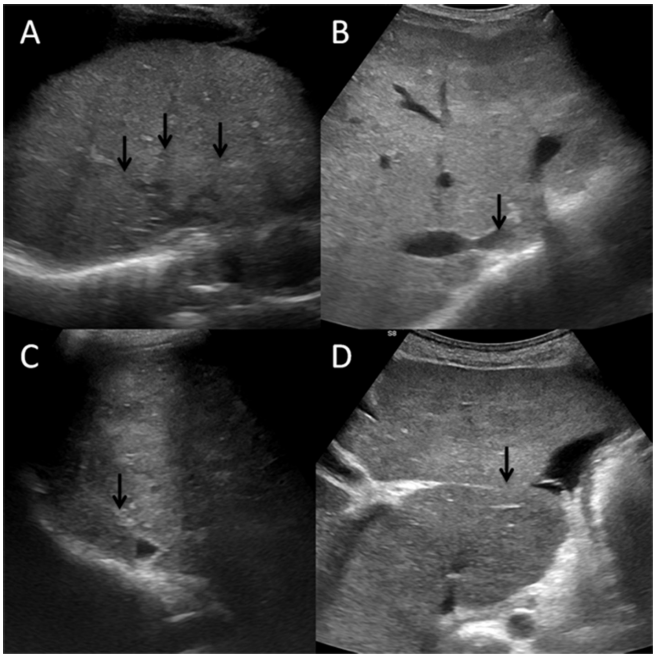
The parenchymal changes of chronic BCS are described in Table 7.

Vascular changes

Direct evidence of venous thrombosis is not much appreciated in the chronic stage. There is the development of small, bridging intrahepatic and capsular hepatic venous collaterals. These are well developed in chronic, not seen in acute, and may be present to a lesser extent in subacute forms.[16,17] Table 8 highlights the vascular changes in chronic BCS.

Features of chronic liver disease and portal hypertension may develop. Portal vein and splenic veins are dilated. The paraumbilical vein is dilated. Multiple collaterals are seen in periportal, peripancreatic, near splenic hilum region.[11] Esophageal and rectal varices may be seen. Besides, the collaterals that develop secondary to portal hypertension, the extrahepatic collaterals are a part of chronic BCS. The pattern of these extrahepatic collaterals is characteristic of BCS.[17] These are discussed in detail in the section below.

Figure 1. Ultrasound changes in BCS: Abnormal hepatic veins that are not seen along their entire course and show slightly echogenic lumen (arrows, A). Short segment stenosis of the right hepatic vein (arrow, B). Echogenic right hepatic vein (arrow, C). Enlarged caudate lobe (arrow, D). BCS, Budd-Chiari syndrome.



ROLE OF INDIVIDUAL IMAGING MODALITIES

Doppler ultrasound

Doppler ultrasound is the first line imaging study for evaluation of patients with BCS.[17] The advantages of Doppler ultrasound are easy availability, cost-effectiveness, and lack of ionizing radiations. It has a sensitivity and specificity of 85% for the diagnosis of BCS.[31] It allows accurate evaluation of the vascular changes including the cause, site, and the extent of the block (Figure 1). It also allows depiction of intrahepatic venovenous collaterals, a highly specific finding for BCS (Figure 2). Besides, it provides information about the blood flow direction and flow pattern.[17] It is also fairly accurate in the demonstration of morphological

Figure 2. Doppler findings in BCS: Dilated caudate lobe vein is seen in a patient with chronic BCS (arrow, A). Doppler ultrasound in another patient with chronic BCS shows marked extrinsic compression of IVC (arrow, B). Doppler ultrasound image in the same patient shows intrahepatic comma-shaped veno-venous collateral between the hepatic veins (arrow, C). Monophasic flow in right hepatic vein that shows normal color flow (arrow, D). BCS, Budd-Chiari syndrome; IVC, inferior vena cava.

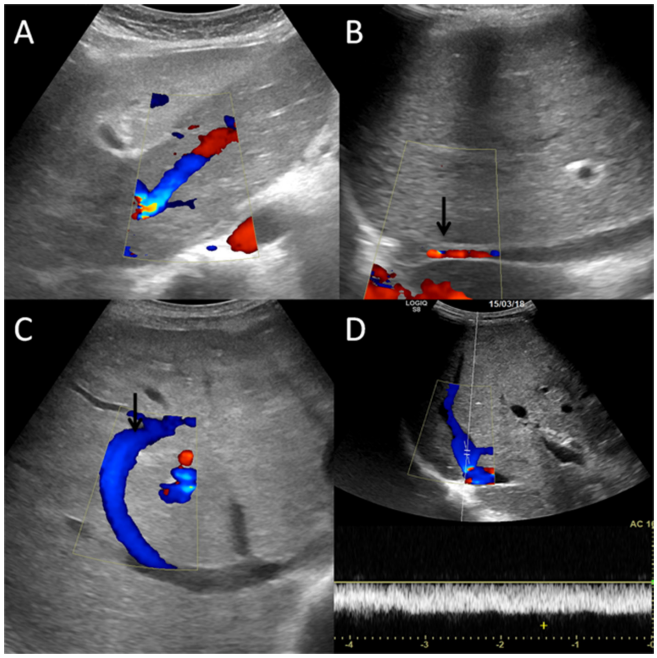


Figure 3. CT findings in BCS: Axial CT image in a patient with subacute BCS shows non-opacified hepatic veins (arrows, A). In another patient with chronic BCS, the abnormal confluence of hepatic veins with IVC is seen on axial CT image (arrow, B). Venovenous and perihepatic collaterals are seen on axial CT in a patient with chronic BCS (arrows, C). Sagittal CT image shows short segment stenosis of IVC with azygous-lumbovetebral collaterals (arrows, D). BCS, Budd-Chiari syndrome; IVC, inferior vena cava.

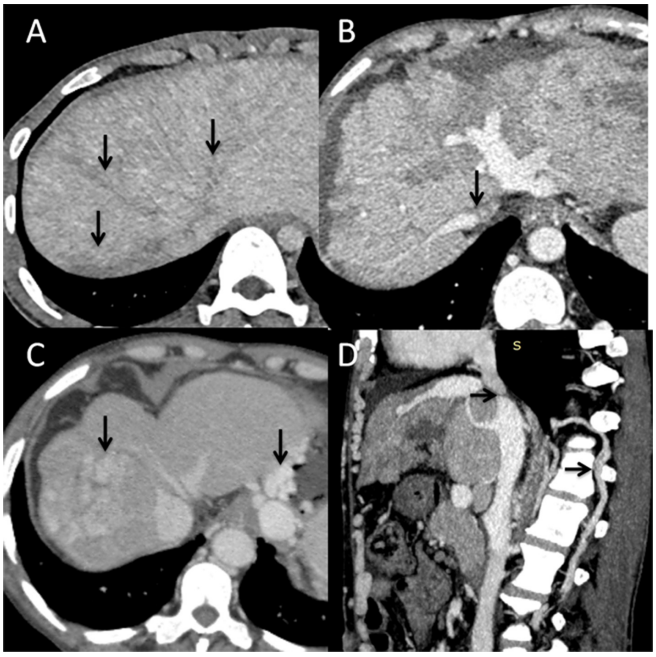
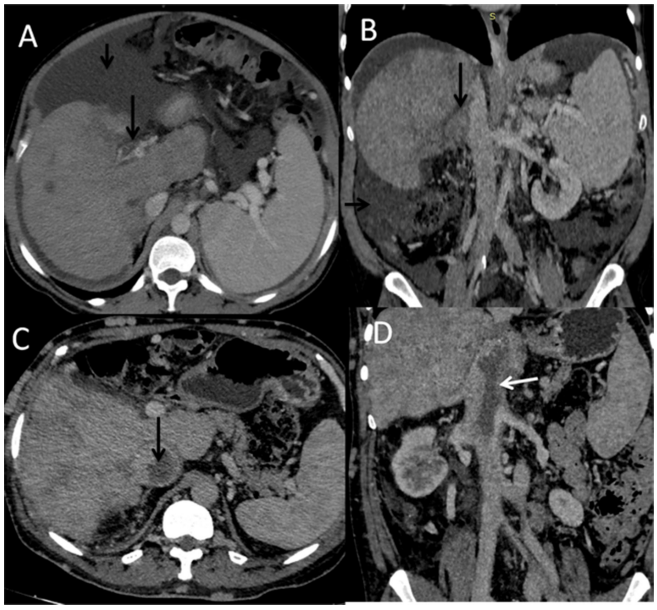


Figure 4. CT findings in BCS: (A) Axial CT image in a patient with subacute BCS shows thrombosis of main portal vein seen as filling defect (arrow, A). Coronal CT image in a patient with acute BCS shows filling defect in the middle HV (arrow, B). Ascites is also seen (short arrows, A and B). Axial and coronal CT images in a patient with thrombosis of IVC seen as filling defect causing expansion of the lumen (arrows, C and D). BCS, Budd-Chiari syndrome; HV, hepatic veins; IVC, inferior vena cava.



changes in the liver as well as the demonstration of changes of portal hypertension. It is less accurate in mapping the extrahepatic collateral pathways and characterization of focal liver lesions in the setting of BCS.[11] The limitations of Doppler ultrasound include operator dependence, technical difficulty posed by obesity and bowel gas, and inability to provide a reproducible roadmap for various endovascular and surgical interventions.[32] It remains the imaging test of choice in follow up of patients with BCS.

CT

CT is one of the most widely performed imaging tests in patients with suspected BCS. It delineates the vascular abnormality, evaluates the hepatic morphological changes as well as allows mapping of the vascular anatomy prior to endovascular interventions or surgery.[32] While CT is accurate in depicting the indirect signs of BCS, the venous abnormality (direct signs) may not be consistently and accurately depicted.[17]

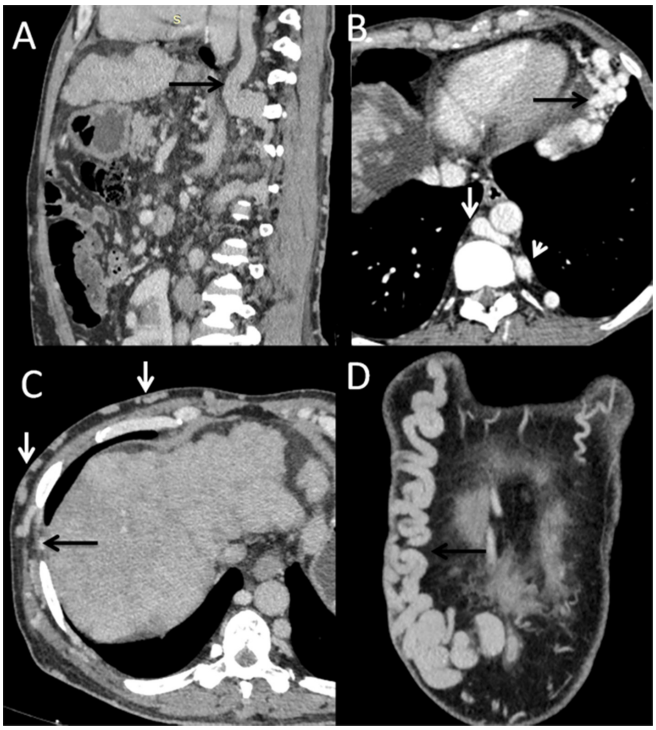
Direct sign

The most common sign of BCS on CT is non-visualized HVs (Figure 3A).[17] While undetectable HVs on CT are indicative of BCS, false positive or indeterminate results have been reported in 50% of cases.[17] Acute HV thrombosis is seen as a filling defect within the HV, which is hypodense on CT (Figure 4).[22] Thrombosis is more easily demonstrated during the acute phase because the involved HVs are distended. Direct identification of ostial stenosis is difficult on CT. Delayed hepatic parenchymal enhancement with HV dilatation may point towards ostial stenosis (Figure 3B).[17] Concomitant portal vein thrombosis may be seen in up to 15% patients (Figure 4A).[11]

The initial studies reported CT to be a less sensitive modality in evaluation of IVC involvement in BCS. Lim et al compared the findings of ultrasound, CT, and catheter venography in the evaluation of patients with suspected membranous obstruction of IVC.[33] 15 patients were evaluated and CT was found to be less sensitive in detecting obliteration of intrahepatic veins and collateral vessels. Ultrasound was superior to CT in demonstrating the IVC membranes and in the depiction of the pathological hepatic venous anatomy.[33] In another study, CT failed to detect IVC membrane in 23 patients.[34] Zhang et al compared the performance of ultrasound and CT in the evaluation of BCS using surgical exploration as the reference standard.[35] 70 patients with BCS were evaluated. Ultrasound was significantly more accurate than CT in detecting membranous lesions of the inferior vena cava, hepatic vein openings, and short-segment lesions (thrombosis and fibrous tissue without the membrane). There was no significant difference between ultrasound and CT in the detection of long segment lesions of the hepatic veins.[35]

However, recent studies report good diagnostic performance of CT in BCS. Zhou et al compared image quality and anatomy delineation by CT venography under fixed or flexible delayed scan times in BCS patients with IVC obstruction.[36] A total of

Figure 5. Collateral pathways in BCS: Coronal CT image in a patient with chronic BCS shows dilated azygous vein draining the lumbovertebral veins (arrow, A). In another patient with chronic BCS, pericardiophrenic venous collaterals is seen on axial CT image (arrow, B). Also seen are dilated azygous (short arrow, B) and hemiazygous veins (arrow head, B). Abdominal wall venous collaterals (short arrows, C) draining the hepatic capsular veins (arrow, C) and markedly dilated tortuous epigastric collaterals (arrow, D) are seen in a patient with chronic BCS. BCS, Budd-Chiari syndrome.

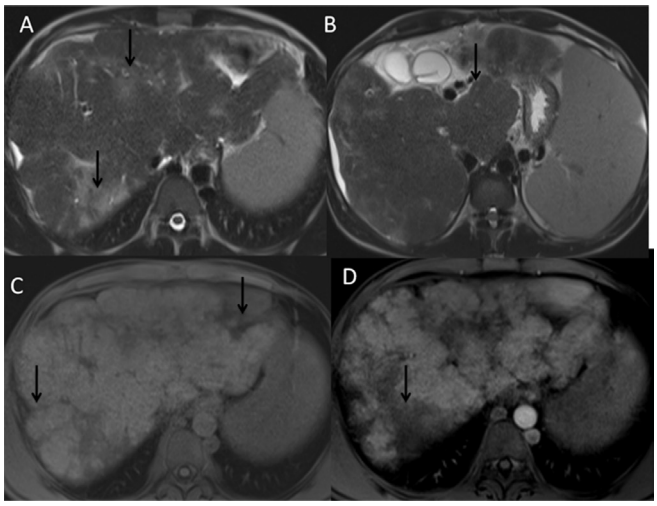


209 patients with BCS with IVC obstruction underwent either a CT venography study with a fixed delayed (180 s; n = 87) or a flexible delayed scan time according to IVC blood flow in color Doppler ultrasonography (n = 122). The flexible delayed scan time of CT venography allowed better detection and characterization of IVC obstruction compared to a fixed delayed scan time.[36] High diagnostic accuracy has also been reported by Liu et al in 108 BCS patients with partial IVC obstruction who underwent CT venography.[32] Virmani et al also reported an excellent correlation between CT venography and catheter venography in predicting the presence as well as grading the degree and length of IVC stenosis in 25 patients with BCS.[37] IVC membranes were accurately depicted in all the patients (n = 4). CT has also been shown to be highly accurate in detection of HV abnormalities. In a study by Faraoun et al 171 patients with BCS with HV abnormalities with or without IVC abnormalities were evaluated using Doppler ultrasound, CT, and MRI.[38] Perfect agreement amongst the three imaging modalities was reported for detecting long-standing HV thrombus. Slight to moderate agreement was reported for revealing the type of HV abnormality.

Indirect signs

CT is highly accurate in demonstrating the indirect signs of BCS. These include abnormal parenchymal enhancement, morphological alterations, abnormal perfusion, and intrahepatic and extrahepatic collaterals.[17]

Figure 6. MR findings in BCS: Axial T2W in a patient with subacute BCS shows heterogeneous signal intensity of the liver (arrow, A). Axial T2W image in the same patient at a different level shows caudate lobe enlargement (arrow, B). Axial T1W image shows nodular liver outline (arrows, C). Post-gadolinium image in the arterial phase shows heterogeneous enhancement of liver (arrow, D). BCS, Budd-Chiari syndrome; T1W, T_1 weighted; T2W, T_2 weighted.



Abnormal parenchymal enhancement

During the acute stage, the liver appears hypoattenuating and heterogeneous due to the predominance of venous congestion.[17] There is a difference in the degree and timing of enhancement of central and peripheral liver. During the subacute and chronic

Figure 7. MR findings in BCS: Axial contrast enhanced MR image in a patient with chronic BCS shows non-opacified hepatic veins (arrows, A). Axial and coronal contrast-enhanced MR images show filling defect in IVC (arrows, B and C). Non-opacified hepatic veins are seen in another patient with chronic BCS (arrows, D). BCS, Budd-Chiari syndrome; IVC, inferior vena cava.

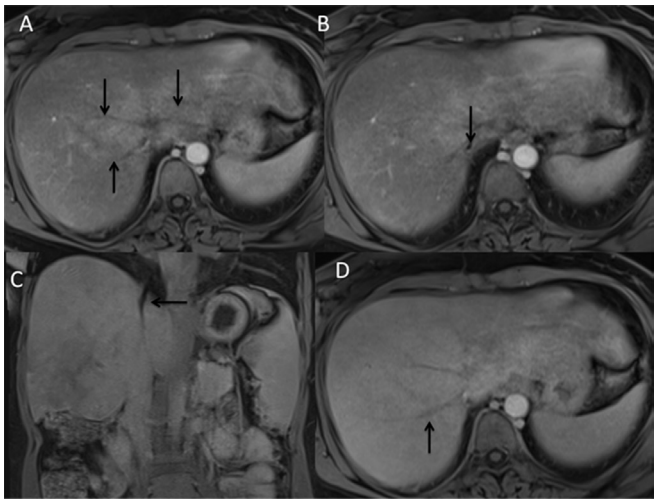
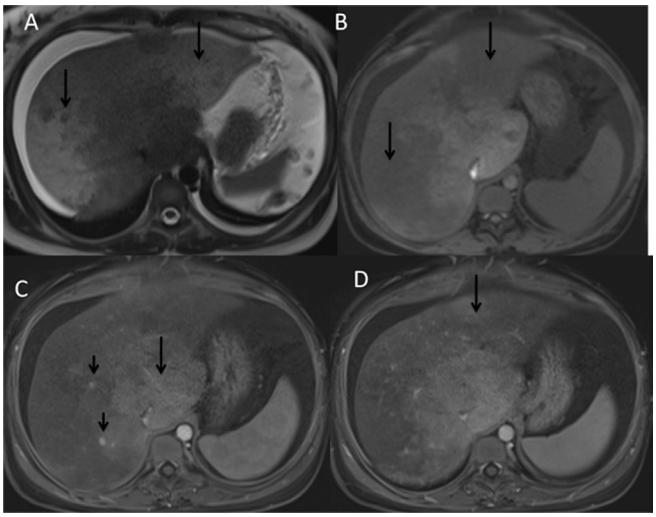


Figure 8. MR findings in BCS: Heterogeneous signal intensity of the liver is seen on axial T2W and T1W images in a patient with subacute BCS (arrow, A and B). Enhancement of the caudate lobe with non-enhancing periphery is seen in the arterial phase axial MR image in the same patient (arrow, B). Also note tiny hyper-enhancing nodules in right lobe (short arrows, C) suggestive of nodular regenerative hyperplasia. In the venous phase in the same patient, there is some enhancement of the liver periphery (arrows, D). BCS, Budd-Chiari syndrome; T1W, T_1 weighted; T2W, T_2 weighted.



stages of BCS, fibrosis sets in and leads to changes of cirrhosis. The heterogeneity persists and is more uniformly distributed while differences in the pattern of enhancement are more subtle.

Morphological alterations

During the acute stage, uniform hepatomegaly with a smooth liver outline is present.[16] During the subacute and chronic stages, morphological alterations are typified by nodular outline and volume redistribution with atrophic and hypertrophic segments. Caudate lobe is markedly enlarged.

Perfusion abnormalities

Perfusion abnormalities are produced by elevation of sinusoidal pressure. These are visible during the portal venous phase in the acute stage as diffuse hyperattenuating areas or mosaic areas.[39] The diffuse hyperattenuating areas are initially seen around the portal vessels. Later, these extend to the surrounding liver parenchyma. As the intrahepatic collateral vessels develop during the course of long-standing BCS, there is a hemodynamic balance between pre- and post-sinusoidal pressures and these hyperattenuating areas are no longer seen.[17]

Intrahepatic collaterals

This is a highly specific sign of BCS.[17] The collaterals form in the chronic stage in an attempt to decongest the liver parenchyma. The veno-venous collaterals may form between two or more HVs, HVs and pericapsular veins, HVs and IVC, and HVs and veins of caudate lobe.[40] CT is accurate in the depiction of veno-venous collaterals that are seen during the portal venous phase as comma-shaped enhancing channels bridging the vascular structures (Figure 3C). Sometimes, these channels may remain unenhanced due to the marked alteration in blood flow dynamics.

Extrahepatic collaterals

The pattern of extrahepatic collaterals in the setting of BCS is different from that seen in portal hypertension from other causes. The venous channels that drain the kidneys, diaphragm, vertebral plexus, and deep pelvic network form the extrahepatic collateral pathways in BCS (Figure 3C,D).[41] They can be divided into three deep and one superficial venous pathway. The former allows the communication between IVC and the superior vena cava via the left renal vein (to hemiazygos vein), the lumbovertebral venous plexus (to azygos vein), and the

Table 4. Parenchymal changes on imaging in acute BCS

Modality	Findings
Ultrasound (Figure 1)	Liver is large and bulbous
	Altered regional echogenicity (due to hemorrhagic infarction)
	Sparing of caudate lobe (emissary veins drain directly into IVC) with caudate hypertrophy
	Ascites
CT (Figure 3)	Patchy, decreased peripheral enhancement caused by portal and sinusoidal stasis and stronger enhancement of the central portion of liver parenchyma
	Caudate size ranges from normal to moderately increased
	Ascites
MRI (Figures 4–6)	T1W-decreased signal intensity within liver periphery and preservation of more normal, higher SI within the central liver and caudate lobe
	T2W-heterogeneously increased signal intensity in the peripheral portion of the liver and caudate lobe.
	Post-gadolinium images-increased enhancement within caudate lobe on arterial phase which persists on delayed phase images. Enhancement of liver periphery is heterogeneously decreased throughout early and late post-contrast sequences which persists on delayed phase images.

BCS, Budd-Chiari syndrome; IVC, inferior vena cava; SI, signal intensity; T1W, T_1 weighted; T2W, T_2 weighted.

Table 5. Vascular changes on imaging in acute BCS

Modality	Hepatic veins	IVC	Additional findings
Ultrasound (Figure 1)	Partial or complete inability to see hepatic veins	Narrowing due to compression by the enlarged caudate lobe	Caudate vein ≥3 mm in diameter strongly suggests the diagnosis in appropriate clinical setting.[13]
	Stenosis with proximal dilatation	Long segment narrowing without associated caudate lobe enlargement	
	Intraluminal echogenicity	Localized, marked narrowing consistent with a web	
	Thickened walls	Focal localized thrombus	
		Membranous web is seen as echogenic areas or focal obliteration of the lumen	
		An aneurysm may be seen in IVC	
Color Doppler (Figure 2)	No flow in part or all hepatic veins	Partial IVC obstruction or extrinsic IVC compression leads to pseudoportal flow	Portal flow may be affected, is dampened or reversed.
	Flow in the abnormal direction in part or all hepatic veins		
	High flow velocity is seen at the site of stenosis		
	Change in flow from phasic to absent, reversed, turbulent or continuous.		
	Continuous flow called "*pseudoportal Doppler*" signal		
CT (Figure 3)	Thrombosed hepatic veins/IVC seen as filling defect (partial/complete)	IVC compressed by enlarged caudate lobe	
MRI (Figures 4–6)	Abnormal appearance of hepatic veins on T2W images	Extrinsic compression of the IVC	
	Non-visualisation of hepatic veins	Filling defect	
	Non-opacification of one or more hepatic veins	Non-opacification	
	Filling defects within one or more hepatic veins		
	Focal stenosis of hepatic veins		

BCS, Budd-Chiari syndrome; IVC, inferior vena cava.

phrenic vein (to pericardiophrenic vein) (Figure 5A,B).[41] The superficial drainage pathway allows drainage of intrahepatic collaterals to veins of the chest and abdominal wall via hepatic capsular veins (Figure 5C,D). These channels include superior and inferior epigastric veins. CT allows adequate visualization of all the extrahepatic draining veins and in cases where the diagnosis of BCS is uncertain, the characteristic pattern of extrahepatic collaterals may suggest this diagnosis.

Table 6. Parenchymal changes on imaging in subacute BCS

Modality	Findings
Ultrasonography (Figure 1)	Enlargement of the caudate lobe with liver surface nodularity with hetero-echoic pattern of the peripheral liver parenchyma
	Ascites
CT (Figure 3)	Liver volume redistribution with a nodular surface
	Heterogeneous enhancement of the peripheral liver in the early contrast enhanced phase with homogeneous attenuation in delayed phases
	Homogeneous enhancement of the enlarged caudate lobe
MRI (Figures 4–6)	Preservation of normal signal intensity in the caudate lobe on all sequences Heterogeneously decreased SI within the peripheral liver on T_1 weighted images and heterogeneously increased SI within the peripheral liver on T_2 weighted images.
	Post-contrast enhancement of caudate lobe is less prominent than the heterogeneously increased enhancement observed in the peripheral liver. Overall enhancement pattern becomes more homogeneous on more delayed imaging sequence.

BCS, Budd-Chiari syndrome; SI, signal intensity.

Post-contrast enhancement of caudate lobe is less prominent than the heterogeneously increased enhancement observed in the peripheral liver. Overall enhancement pattern becomes more homogeneous on more delayed imaging sequence.

Table 7. Parenchymal changes on imaging in chronic BCS

Modality	Finding
Ultrasound (Figure 1)	Features of cirrhosis NRH nodules are iso-echoic and may be difficult to visualize
CT (Figure 3)	Regenerative nodules are multiple, ranging in size from 0.5 to 4 cm.
	Multiphasic CT – nodules are homogeneous. Show marked arterial phase enhancement. Enhancement persists in portovenous phase.[17]
MRI (Figures 4–6)	Changes of fibrosis and chronic liver parenchymal disease. Regenerative nodules: T_1 bright T_2 isointense to hypointense Post-contrast – hypervascular in arterial phase, no washout in the venous phase

BCS, Budd-Chiari syndrome; NRH, nodular regnerative hyperplasia.

MRI

In some centers, MR venography is preferred over CT venography for the initial evaluation of patients suspected to have BCS. Besides, MR venography may be utilized as an adjunct to Doppler ultrasound in the assessment of patients with BCS. MRI is an excellent modality for the depiction of all the aspects of BCS including vascular changes, morphological changes, mapping of intrahepatic and extrahepatic collaterals, as well as characterization of liver nodules (Figures 6–8).[17,42] It is also useful in follow up of patients following endovascular or surgical interventions.

Catheter venography

Venography is useful in the assessment of the extent of outflow obstruction and also allows for pressure measurements.[1] Also, endovascular interventions can be performed at the same sitting. It is however invasive, time-consuming, and needs expertise.[43] It is usually not used for diagnosis alone except in situations where the results of other studies are equivocal or conflicting.

The diagnostic performance of various imaging techniques in BCS is highlighted in Table 9.

The dilemma in imaging of BCS (conditions mimicking diagnosis of BCS or posing potential problems in diagnosis of BCS)

(1) Chronic liver parenchymal disease: In chronic liver parenchymal disease, there is IVC pseudostenosis which may mimic BCS.[44] There may be non-visualization of the hepatic vein near confluence which may lead to the false label of BCS.
(2) Obesity: one limiting factor in these patients is inadequate insonation of ultrasound waves.[32] So there may be a diagnostic dilemma in these patients, particularly when the patient is being worked up for cryptogenic cirrhosis and imaging needs to rule out the possibility of BCS. These patients may undergo MR venography. However, MR venography may have its own limitations including claustrophobia, patients with aneurysmal clips, inability to hold breath which leads to degraded quality of images.

Table 8. Vascular changes on imaging in chronic BCS

Modality	Findings
Ultrasound Doppler (Figure 2)	Intrahepatic veno-venous collaterals – specific diagnosis. Seen as comma-shaped channels showing color flow
	Intrahepatic collaterals – communicate with systemic vessels via subcapsular collaterals
	Collaterals from an occluded hepatic vein to a non-occluded hepatic vein
	Collaterals form occluded hepatic vein to caudate lobe veins
	Rarely from hepatic veins to suprahepatic IVC close to the right atrium.
CT venography (Figure 3)	The diameter of the hepatic artery is enlarged compared to the splenic artery.
	Multiple collateral channels as described above may be seen though less confidently
MR venography (Figures 4–6)	Venous collateralization may be seen with the same or higher degree of sensitivity and specificity as the ultrasound Doppler
Venography	Spiderweb pattern of collateral vessels
	Long segment compression of IVC caused by caudate lobe hypertrophy
	Thrombus within hepatic veins seen as filling defect
	Dilated hepatic artery with arterioportal shunts
	Thrombus in IVC

BCS, Budd-Chiari syndrome; IVC, inferior vena cava.

Table 9. Diagnostic performance of various imaging modalities in the diagnosis of BCS

Modality	Sensitivity	Specificity	PPV	NPV
Ultrasound Doppler[31]	87.5%	85%	–	–
CT venography[32a]	86.1%	97.3%	93.9%	93.5%
MR venography[42a]	100%	66.7%	94%	100%

BCS, Budd-Chiari syndrome; NPV, negative predictive value; PPV, positive predictive value.
[a]For IVC obstruction.

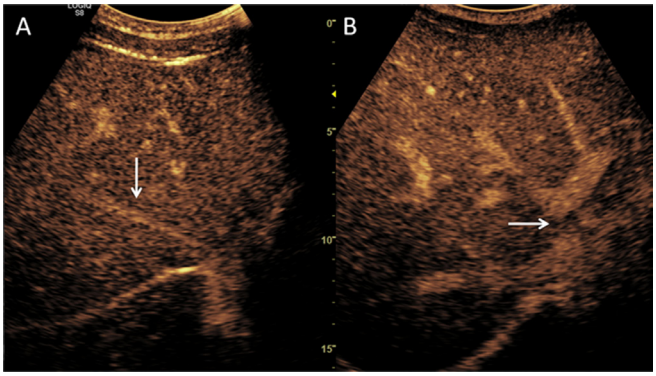
(3) The angle of insonation in Doppler: in author experience, 0° angle can be achieved only with the right hepatic vein. Angle can be achieved to a satisfactory level in middle hepatic vein. But due to the inherent angle of the left hepatic vein, appropriate Doppler angle may not be achieved which may lead to erroneous results and false interpretation.
(4) Unstable patients (fulminant/ acute BCS): Inability to hold breath which leads to diagnostic difficulties both in ultrasound Doppler and MR venography.

In author experience, contrast-enhanced ultrasound (Figure 9) is an important problem-solving tool in patients who are suspected to be having BCS clinically but there are issues with other imaging techniques as discussed above.

Imaging-guided therapeutic maneuvers

Endovascular treatment is the most important therapeutic option in patients with BCS.[45] Endovascular interventions include angioplasty, stenting, catheter-directed thrombolysis, and creation of transjugular intrahepatic portosystemic shunts (TIPSS).[16,45] Cross-sectional imaging including CT and MR venography play an important role in patient selection and treatment planning.[11] If all HVs are occluded, angioplasty or stenting is not feasible while TIPSS may be considered. Short segment stenosis or occlusion are predictors of the success of angioplasty and stenting. Additional information sought on imaging prior to treatment planning includes the presence and size of intrahepatic venous collaterals, permeability of the IVC and intra and extrahepatic portal veins as well as the size of the caudate lobe vein and accessory right hepatic veins.[11]

Figure 9. Contrast-enhanced ultrasound: Normal hepatic veins are seen as enhancing structures (arrows, A). Stenosis is seen at the junction of the middle-left hepatic vein and IVC in a patient with subacute BCS (arrow, B). BCS, Budd-Chiari syndrome; IVC, inferior vena cava.



Acute BCS

Catheter-directed thrombolysis

In acute BCS, radiological interventions allow thrombolysis of the involved HV.[45] This can be achieved by using pharmacological agents. Mechanical thrombolysis may also be utilized to restore deteriorating liver function. Thrombolysis technique is determined by the age of the thrombus; pharmacological agents alone may be effective for a hyperacute thrombus while older thrombus may additionally require mechanical thrombolysis.[45]

TIPSS

TIPSS has been extensively studied and found to be effective in patients with chronic BCS.[46,47] Few studies have described its favorable results in the acute BCS. He et al reported the results of TIPSS in 91 patients including 14 with acute BCS. TIPSS was

Figure 10. Inferior vena cava angioplasty and stenting: Venogram performed via transfemoral route shows lack of opacification of IVC. There is filling of multiple collaterals (arrows, A). Following balloon angioplasty, there is significant residual stenosis and stent is placed (arrows, B). Stent is seen *in situ* (arrows, C). Post-angioplasty and stenting venogram reveals normal opacification of the IVC (arrow, D). IVC, inferior vena cava.

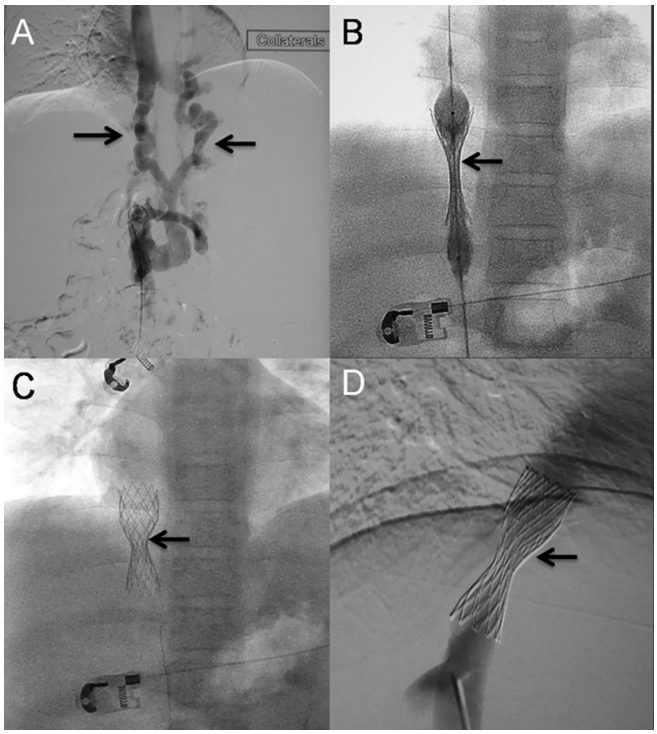
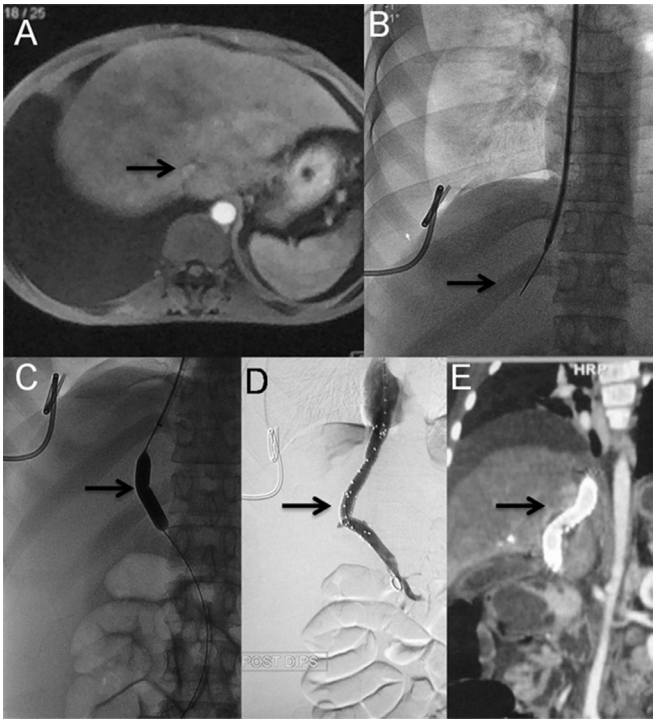


Figure 11. TIPSS in a patient with subacute BCS: Axial CT image shows lack of opacification of the hepatic veins; IVC is normally opacified (arrow, A). Right portal vein is accessed via the right transjugular approach (arrow, B). A parenchymal tract is created (arrows, C). Venogram reveals the TIPSS tract (arrow, D). Post-procedure CT shows normal opacification of the TIPSS stent (arrow, E). BCS, Budd-Chiari syndrome; IVC, inferior vena cava; TIPSS, transjugular intrahepatic portosystemic shunts.

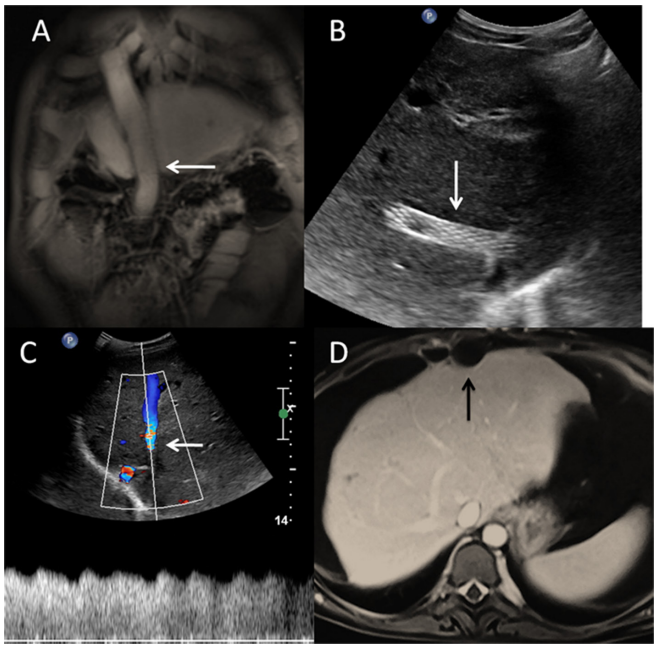


technically successful in 12 patients with a reduction of portosystemic pressure gradient to normal and significant improvement in the liver function and serum bilirubin.[48] In another study by He et al comprising 37 patients with acute BCS, TIPSS was performed in 21 patients with 100% technical success.[49] No major procedure-related complications were reported. There was a significant reduction in the portosystemic pressure gradient as well as serum bilirubin. However, in the study by Mancuso et al, out of the eight patients with acute BCS and hepatic failure who underwent TIPSS, deaths were reported in four patients soon after the TIPSS.[50]

Chronic BCS

Patients are categorized into two groups for planning interventions. Patients having membranous or short segment occlusion of HV/IVC are the ideal candidates for angioplasty with or without stenting (Figure 10).[45] In general, balloon angioplasty is sufficient for short segment stenosis or occlusion. If there is residual stenosis following balloon angioplasty, stenting may be required.[45] The patients in whom all the HV are replaced by intrahepatic collaterals are not suitable for angioplasty and require the creation of TIPSS (Figure 11).[16]

Figure 12. Post-treatment imaging: Normal opacification of the mesocaval shunt (arrow, A) is seen on coronal MR venography image. Middle hepatic vein stent (arrow, B) on ultrasound. It shows aliasing (arrow, C) and increased flow velocity on Doppler ultrasound evaluation suggestive of the stenosis. Non-opacified meso-caval shunt in another patient on axial post-contrast MR image (arrow, D).



Post-treatment imaging

Follow-up imaging test of choice is ultrasound Doppler (Figure 12A–C). Morphologically, response to treatment is manifest in the form of a reduction in ascites and the size of caudate lobe.[51] The demonstration of color flow along the entire length of stent/ shunt is critical.[51] The whole length of the shunt may be difficult to visualize on Doppler because of body habitus and overlying bowel gas. In these cases, MR venography allows reliable assessment[52] . The blockage may be seen as stenosis or filling defect (Figure 12D). Reversal of blood flow in portal vein serves as an indirect evidence of shunt patency.

REFERENCES

1. Janssen HL, Garcia-Pagan JC, Elias E, Mentha G, Hadengue A, Valla DC, et al. Budd-Chiari syndrome: a review by an expert panel. *J Hepatol* 2003; **38**: 364–71. doi: https://doi.org/10.1016/S0168-8278(02)00434-8
2. Grus T, Lambert L, Grusová G, Banerjee R, Burgetová A, Syndrome B-C. Budd-Chiari syndrome. *Prague Med Rep* 2017; **118**: 69–80. doi: https://doi.org/10.14712/23362936.2017.6
3. Goel RM, Johnston EL, Patel KV, Wong T. Budd-Chiari syndrome: investigation, treatment and outcomes. *Postgrad Med J*

2015; **91**: 692–7. doi: https://doi.org/10.1136/postgradmedj-2015-133402
4. Aydinli M, Bayraktar Y. Budd-Chiari syndrome: etiology, pathogenesis and diagnosis. *World J Gastroenterol* 2007; **13**: 2693–6. doi: https://doi.org/10.3748/wjg.v13.i19.2693
5. Martens P, Nevens F. Budd-Chiari syndrome. *United European Gastroenterol J* 2015; **3**: 489–500. doi: https://doi.org/10.1177/2050640615582293
6. Valla DC. Budd-Chiari syndrome/hepatic venous outflow tract obstruction. *Hepatol Int* 2018; **12**(Suppl 1): 168–80. doi: https://doi.org/10.1007/s12072-017-9810-5
7. Ludwig J, Hashimoto E, McGill DB, van Heerden JA. Classification of hepatic venous outflow obstruction: ambiguous terminology of the Budd-Chiari syndrome. *Mayo Clin Proc* 1990; **65**: 51–5. doi: https://doi.org/10.1016/S0025-6196(12)62109-0
8. Loomes DE, Chang A, Webber D, Scudamore CH, Yoshida EM. Acute Budd-Chiari syndrome. *Can J Gastroenterol* 2011; **25**: 302–3. doi: https://doi.org/10.1155/2011/756425
9. Senzolo M, Cholongitas EC, Patch D, Burroughs AK. Update on the classification, assessment of prognosis and therapy of Budd-Chiari syndrome. *Nat Clin Pract Gastroenterol Hepatol* 2005; **2**: 182–90. doi: https://doi.org/10.1038/ncpgasthep0143
10. Noone TC, Semelka RC, Siegelman ES, Balci NC, Hussain SM, Kim PN, et al. Budd-Chiari syndrome: spectrum of appearances of acute, subacute, and chronic disease with magnetic resonance imaging. *J Magn Reson Imaging* 2000; **11**: 44–50. doi: https://doi.org/10.1002/(SICI)1522-2586(200001)11:1<44::AID-JMRI6>3.0.CO;2-O
11. Van Wettere M, Bruno O, Rautou P-E, Vilgrain V, Ronot M. Diagnosis of Budd–Chiari syndrome. *Abdom Radiol* (. 2017; **13**. doi: https://doi.org/10.1007/s00261-017-1447-2
12. Copelan A, Remer EM, Sands M, Nghiem H, Kapoor B. Diagnosis and management of Budd Chiari syndrome: an update. *Cardiovasc Intervent Radiol* 2015; **38**: 1–12. doi: https://doi.org/10.1007/s00270-014-0919-9
13. Buckley O, O' Brien J, Snow A, Stunell H, Lyburn I, Munk PL, et al. Imaging of Budd-Chiari syndrome. *Eur Radiol* 2007; **17**: 2071–8. doi: https://doi.org/10.1007/s00330-006-0537-2
14. Brancatelli G, Vilgrain V, Federle MP, Hakime A, Lagalla R, Iannaccone R, et al. Budd-Chiari syndrome: spectrum of imaging findings. *AJR Am J Roentgenol* 2007; **188**: W168–W176. doi: https://doi.org/10.2214/AJR.05.0168
15. Erden A. Budd-Chiari syndrome: a review of imaging findings. *Eur J Radiol* 2007; **61**: 44–56. doi: https://doi.org/10.1016/j.ejrad.2006.11.004
16. Das CJ, Soneja M, Tayal S, Chahal A, Srivastava S, Kumar A, et al. Role of radiological imaging and interventions in management of Budd-Chiari syndrome. *Clin Radiol* 2018; **73**: 610–24. doi: https://doi.org/10.1016/j.crad.2018.02.003
17. Faraoun SA, Boudjella MA, Debzi N, Benidir N, Afredj N, Guerrache Y, et al. Budd-Chiari syndrome: an update on imaging features. *Clin Imaging* 2016; **40**: 637–46. doi: https://doi.org/10.1016/j.clinimag.2016.01.006
18. Plessier A, Valla DC, Syndrome B-C. Budd-Chiari syndrome. *Semin Liver Dis* 2008; **28**: 259–69. doi: https://doi.org/10.1055/s-0028-1085094
19. Soyer P, Rabenandrasana A, Barge J, Laissy JP, Zeitoun G, Hay JM, et al. MRI of Budd-Chiari syndrome. *Abdom Imaging* 1994; **19**: 325–9. doi: https://doi.org/10.1007/BF00198189
20. Valla DC. Primary Budd-Chiari syndrome. *J Hepatol* 2009; **50**: 195–203. doi: https://doi.org/10.1016/j.jhep.2008.10.007
21. Torabi M, Hosseinzadeh K, Federle MP. CT of nonneoplastic hepatic vascular and perfusion disorders. *Radiographics* 2008; **28**: 1967–82. doi: https://doi.org/10.1148/rg.287085067
22. Yilmaz S, Yekeler E, Agayev A, Pinarbasi B, Bakkaloglu H, Acunas B. A rapidly increasing abdominal girth in a young patient: MDCT findings of Budd-Chiari syndrome. *Surgery* 2008; **144**: 101–2. doi: https://doi.org/10.1016/j.surg.2007.06.013
23. Bargalló X, Gilabert R, Nicolau C, García-Pagán JC, Ayuso JR, Brú C. Sonography of Budd-Chiari syndrome. *AJR Am J Roentgenol* 2006; **187**: W33–W41. doi: https://doi.org/10.2214/AJR.04.0918
24. Ferral H, Behrens G, Lopera J. Budd-Chiari syndrome. *AJR Am J Roentgenol* 2012; **199**: 737–45. doi: https://doi.org/10.2214/AJR.12.9098
25. Stark DD, Hahn PF, Trey C, Clouse ME, Ferrucci JT. MRI of the Budd-Chiari syndrome. *AJR Am J Roentgenol* 1986; **146**: 1141–8. doi: https://doi.org/10.2214/ajr.146.6.1141
26. p. Klatskin GCH. *Histopathology of the liver*. New York: Oxford University Press; 1993. pp. 123–4.
27. Brancatelli G, Federle MP, Grazioli L, Golfieri R, Lencioni R. Benign regenerative nodules in Budd-Chiari syndrome and other vascular disorders of the liver: radiologic-pathologic and clinical correlation. *Radiographics* 2002; **22**: 847–62. doi: https://doi.org/10.1148/radiographics.22.4.g02jl17847
28. de Sousa JM, Portmann B, Williams R. Nodular regenerative hyperplasia of the liver and the Budd-Chiari syndrome. Case report, review of the literature and reappraisal of pathogenesis. *J Hepatol* 1991; **12**: 28–35.
29. Tanaka M, Wanless IR. Pathology of the liver in Budd-Chiari syndrome: portal vein thrombosis and the histogenesis of veno-centric cirrhosis, veno-portal cirrhosis, and large regenerative nodules. *Hepatology* 1998; **27**: 488–96. doi: https://doi.org/10.1002/hep.510270224
30. Takayasu K, Muramatsu Y, Moriyama N, Wakao F, Makuuchi M, Takayama T, et al. Radiological study of idiopathic Budd-Chiari syndrome complicated by hepatocellular carcinoma. A report of four cases. *Am J Gastroenterol* 1994; **89**: 249–53.
31. Miller WJ, Federle MP, Straub WH, Davis PL. Budd-Chiari syndrome: imaging with pathologic correlation. *Abdom Imaging* 1993; **18**: e35–. doi: https://doi.org/10.1007/BF00201775
32. Liu SY, Xiao P, Cao HC, Jiang HS, Li TX. Accuracy of computed tomographic angiography in the diagnosis of patients with inferior vena cava partial obstruction in Budd-Chiari syndrome. *J Gastroenterol Hepatol* 2016; **31**: 1933–9. doi: https://doi.org/10.1111/jgh.13420
33. Lim JH, Park JH, Auh YH. Membranous obstruction of the inferior vena cava: comparison of findings at sonography, CT, and venography. *AJR Am J Roentgenol* 1992; **159**: 515–20. doi: https://doi.org/10.2214/ajr.159.3.1503015
34. Ciesek S, Rifai K, Bahr MJ, Boozari B, Steinmann E, Helfritz FA, et al. Membranous Budd-Chiari syndrome in Caucasians. *Scand J Gastroenterol* 2010; **45**: 226–34. doi: https://doi.org/10.3109/00365520903406719
35. Zhang LM, Zhang GY, Liu YL, Wu J, Cheng J, Wang Y. Ultrasonography and computed tomography diagnostic evaluation of Budd-Chiari syndrome based on radical resection exploration results. *Ultrasound Q* 2015; **31**: 124–9. doi: https://doi.org/10.1097/RUQ.0000000000000122
36. Zhou PL, Wu G, Han XW, Bi YH, Zhang WG, Wu ZY, YH B, ZY W. Detection and characterization of Budd-Chiari syndrome with inferior vena cava obstruction: comparison of fixed and flexible delayed scan time of computed tomography venography. *Eur J Radiol* 2017; **91**: 52–6. doi: https://doi.org/10.1016/j.ejrad.2017.03.021
37. Virmani V, Khandelwal N, Kang M, Gulati M, Chawla Y. MDCT venography in the evaluation of inferior vena cava in Budd-

Chiari syndrome. *Indian J Gastroenterol* 2009; **28**: 17–23. doi: https://doi.org/10.1007/s12664-009-0004-5
38. Faraoun SA, Boudjella MA, Debzi N, Afredj N, Guerrache Y, Benidir N, et al. Budd-Chiari syndrome: a prospective analysis of hepatic vein obstruction on ultrasonography, multidetector-row computed tomography and MR imaging. *Abdom Imaging* 2015; **40**: 1500–9. doi: https://doi.org/10.1007/s00261-015-0380-5
39. Meng XC, Zhu KS, Qin J, Zhang JS, Wang XH, Zou Y, et al. Clinical significance of multislice spiral CT scans in hepatic veins occlusion in Budd-Chiari syndrome. *Chin Med J* 2007; **120**: 100–5.
40. Chandrasekaran S, Velliyappillil J, Kumar Muthusamy A, Joseph G, Venkataraman J. Alternate pathways in hepatic venous outflow obstruction by color Doppler imaging. *Ann Gastroenterol* 2007; **20**: 218–20.
41. Cho OK, Koo JH, Kim YS, Rhim HC, Koh BH, Seo HS. Collateral pathways in Budd-Chiari syndrome: CT and venographic correlation. *AJR Am J Roentgenol* 1996; **167**: 1163–7. doi: https://doi.org/10.2214/ajr.167.5.8911174
42. Lu X, Yang C, Xu K, Rong YT, Li SD, Li JS, et al. Magnetic resonance venography in the diagnosis of inferior vena cava obstruction in Budd-Chiari syndrome. *Eur Rev Med Pharmacol Sci* 2015; **19**: 256–64.
43. Valla DC. The diagnosis and management of the Budd-Chiari syndrome: consensus and controversies. *Hepatology* 2003; **38**: 793–803. doi: https://doi.org/10.1002/hep.1840380404
44. Rector WG. Pseudo-Budd-Chiari syndrome: extrinsic deformity of the intrahepatic inferior vena cava mimicking membranous obstruction. *J Clin Gastroenterol* 1989; **11**: 88–91.
45. Mukund A, Gamanagatti S. Imaging and interventions in Budd-Chiari syndrome. *World J Radiol* 2011; **3**: 169–77. doi: https://doi.org/10.4329/wjr.v3.i7.169
46. Fitsiori K, Tsitskari M, Kelekis A, Filippiadis D, Triantafyllou K, Brountzos E. Transjugular intrahepatic portosystemic shunt for the treatment of Budd-Chiari syndrome patients: results from a single center. *Cardiovasc Intervent Radiol* 2014; **37**: 691–7. doi: https://doi.org/10.1007/s00270-013-0697-9
47. Michl P, Bilzer M, Waggershauser T, Gülberg V, Rau HG, Reiser M, et al. Successful treatment of chronic Budd-Chiari syndrome with a transjugular intrahepatic portosystemic shunt. *J Hepatol* 2000; **32**: 516–20. doi: https://doi.org/10.1016/S0168-8278(00)80405-5
48. He F, Zhao H, Dai S, Wu Y, Wang L, Huang H, et al. Transjugular intrahepatic portosystemic shunt for Budd-Chiari syndrome with diffuse occlusion of hepatic veins. *Sci Rep* 2016; **6**: 36380. doi: https://doi.org/10.1038/srep36380
49. He FL, Wang L, Zhao HW, Fan ZH, Zhao MF, Dai S, et al. Transjugular intrahepatic portosystemic shunt for severe jaundice in patients with acute Budd-Chiari syndrome. *World J Gastroenterol* 2015; **21**: 2413–8. doi: https://doi.org/10.3748/wjg.v21.i8.2413
50. Mancuso A, Fung K, Mela M, Tibballs J, Watkinson A, Burroughs AK, et al. TIPS for acute and chronic Budd-Chiari syndrome: a single-centre experience. *J Hepatol* 2003; **38**: 751–4. doi: https://doi.org/10.1016/S0168-8278(03)00118-1
51. Raby N, Karani J, Meire H, Michell M, Howard E. Budd-Chiari syndrome: shunt selection and post-operative assessment. *Clin Radiol* 1989; **40**: 586–90. doi: https://doi.org/10.1016/S0009-9260(89)80312-5
52. Lupescu IG, Dobromir C, Popa GA, Gheorghe L, Georgescu SA. Spiral computed tomography and magnetic resonance angiography evaluation in Budd-Chiari syndrome. *J Gastrointestin Liver Dis* 2008; **17**: 223–6.