Budd-Chiari syndrome is characterized by hepatic venous outflow obstruction, with or without involvement of the inferior vena cava (IVC), and can be due either to venous thrombosis or to membranous occlusion, often leading to chronic liver disease. It is a well known clinical entity in the Far East, the Middle East, India and Africa. The site, extent, and rapidity of hepatic venous occlusion can be highly variable. Information on these aspects of venous obstruction is important for planning appropriate treatment for patients with BCS. Thus, imaging of the hepatic veins and IVC plays a crucial role in the management of patients with BCS. Several non-invasive diagnostic imaging techniques like color Doppler (CD), multidetector computed tomography (MDCT) and, more recently, magnetic resonance imaging (MRI) have all been used in patients with BCS. Because of wide variations in clinical presentations and venous anatomy, and availability of a variety of imaging modalities, patients with BCS need an individualized imaging strategy to obtain a complete evaluation of venous abnormalities and deciding an appropriate treatment plan.

Since the clinical condition of patients with BCS can deteriorate rapidly, an early correct diagnosis and evaluation is important. Ultrasonography combined with CD has a diagnostic sensitivity of more than 75% to diagnose venous obstruction. It is also cheap, quick, non-invasive and widely available. Because of these advantages, it is usually the first line of investigation to diagnose hepatic venous outflow obstruction in patients with liver disease. Normal hepatic veins exhibit characteristic triphasic Doppler signal when patent. Classic findings that suggest involvement of hepatic vein(s) in BCS are non-visualization of the vein(s), replacement of the vein with a fibrous cord-like structure, presence of a thrombus in the vein lumen and venous stenosis at the confluence of the hepatic vein with the IVC. Demonstration of a caudate lobe vein equal to or larger than 3 mm in diameter is a specific sign of BCS, provided the patient does not have heart failure. However, CD evaluation of hepatic veins in a swollen liver in BCS is often difficult and inaccurate, because the compressed hepatic veins may not be visible even though these may be patent. In addition, it is sometimes difficult to detect occlusion of the left and middle hepatic veins using CD as cardiac pulsations are in close proximity with the left lobe of the liver. Thus, though CD appears to be a good screening tool in the initial evaluation of patients suspected of having BCS, it may not provide all the information needed for therapeutic decision-making.

MR evaluation of BCS is preferred as a second line of investigation, since it has a multiplanar capability which allows for a better anatomic assessment of the hepatic veins and the IVC. MR imaging also allows better characterization of benign regenerating nodules and perfusion defects. Furthermore, benign regenerating nodules and hepatocellular carcinoma, both of which are commonly seen in end-stage liver disease due to BCS, can be differentiated using MRI based on their characteristic signal intensities and enhancement patterns. MRI also permits easy visualization of ‘comma-shaped’ intrahepatic collaterals or varices. Thus, MRI may be useful for excluding BCS in patients with end stage liver disease.

On CT evaluation, patients with BCS show a broad spectrum of morphologic and attenuation changes of the liver, hepatic veins and the IVC. Nonvisualization of the hepatic veins, and fan-shaped enhancement of the caudate lobe and central part of the liver around the IVC on CT are diagnostic of BCS. However, in 50% of the cases, hepatic venous thrombosis may not be seen. The advantages of MDCT include superior spatial resolution, very fast image acquisition, and simultaneous evaluation of the liver, which may
be critical in patients with BCS. Isotropic data acquisition during MDCT allows excellent resolution in any plane, which is essential for endovascular or surgical planning. Conventional CT does have some limitations. For instance, it may not depict membranous occlusion of the IVC well. However, newer developments in technology have led to improved the capabilities of CT. Today, MDCT can show the morphology and thickness of the membrane occluding the intrahepatic IVC.

In this issue of the Journal, Virmani et al. have reported their experience with the use of MDCT in patients with BCS. These authors took the unconventional road less traveled and used MDCT venography as a front line investigation to diagnose BCS. They found good correlation between MDCT venography and DSV in the detection, extent, degree of stenosis and site of IVC obstruction. Intrahepatic and extrahepatic collaterals were better seen on MDCT rather than on DSV. However, despite their good results, one must remember that use of MDCT has some limitations in the diagnosis of BCS. This is primarily related to artifacts that occur secondary to flow-related phenomena and partial-volume averaging. These artifacts may mimic a thrombus in the IVC, leading to erroneous diagnosis of BCS. Furthermore, the use of MDCT to diagnose BCS carries the risks of exposure to ionizing radiation and contrast-related complications.

Despite the attractiveness of the relatively non-invasive imaging techniques discussed above, digital subtraction venography remains the gold standard for the evaluation of the IVC as well as the hepatic veins. Besides permitting a better assessment of venous anatomy, this technique also allows for direct measurements of venous pressures and hence gradients at various locations, and permits simultaneous endovascular therapeutic interventions to relieve venous obstruction.

BCS is an uncommon disorder with potentially dismal outcomes in a subset of patients, if left untreated. Therefore accurate diagnosis and successful therapeutic interventions are of vital importance in this condition. Despite the availability of many imaging techniques for the diagnosis and assessment of patients with BCS, a team approach between the treating physician, radiologist and surgeon is essential for selection of an optimal combination and sequence of imaging modalities in an individual patient.

References