Noncirrhotic Portal Hypertension: Imaging, Hemodynamics, and Endovascular Therapy

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Patients with noncirrhotic portal hypertension (NCPH) present with clinical features of portal hypertension, but without evidence of significant hepatic parenchymal dysfunction. Standard as well as emerging imaging modalities can assist in diagnosis and management of this condition and will be described in the following sections.

Ultrasound

Ultrasonography is the first-line imaging modality in patients with elevated portal pressures, because it is noninvasive, relatively inexpensive, and can accurately evaluate the portal venous system with the added advantage of determining the velocity and direction of flow.¹ In noncirrhotic portal fibrosis/idiopathic portal hypertension (NCPF/IPH), findings may include normal liver size and echotexture, splenomegaly, and echogenic thick-walled portal veins.² The intrahepatic portal veins (PV) will often demonstrate a “withered tree” appearance, or a sudden narrowing or cut-off of intrahepatic second- and third-degree PV branches.²

In the setting of extrahepatic portal venous obstruction (EHPVO), Doppler imaging can easily identify a portal venous thrombosis or portal cavernous transformation. Flow is generally hepatopetal and continuous, with little if any respiratory or cardiac variation.³ Sonography has been shown to have an overall diagnostic accuracy of 80% in establishing the presence and etiology of portal hypertension due to cirrhosis, NCPF/IPH, and EHPVO.⁴ Features of portal hypertension included splenomegaly, hepatic and splenic hilar collaterals, and portal and splenic vein diameters greater than 10 mm. Splenic infarcts and absence of ascites were features of NCPH

(NCPF and EHPVO) and that nonvisualization of the portal vein and presence of a cavernoma had a diagnostic accuracy of 98% in EHPVO.⁴

Although sonography provides a good initial assessment of the portal venous system, it is extremely operator-dependent, and evaluation of the entire portal venous system is often limited by uncontrollable patient characteristics such as body habitus, overlying bowel gas, and so forth. Therefore, in patients who are potential candidates for surgery or an interventional procedure, further evaluation with cross-sectional examinations such as computed tomography (CT) and magnetic resonance (MR) angiography are required.¹

Contrast-enhanced ultrasonography has also shown some promise in differentiating NCPH from cirrhosis. In a small prospective study designed to differentiate NCPF/IPH from cirrhosis using contrast-enhanced ultrasound using a perflubutane microbubble agent, delayed periportal enhancement was demonstrated more frequently in NCPF/IPH than in controls/cirrhosis, suggesting that it may be a characteristic of NCPF/IPH.⁵

Transient elastography is a noninvasive imaging technology for estimating hepatic fibrosis with a high degree of histopathologic correlation. Correlation between liver stiffness and clinical outcomes has mainly been shown in patients with cirrhosis; its utility in evaluation of NCPH has not been extensively explored and is yet to be established.⁶

CT/MRI

CT and MR angiography and portography are useful in the diagnosis of EHPVO as well as for preprocedural planning

Abbreviations: CT, computed tomography; DIPS, direct intrahepatic portocaval shunt; EHPVO, extrahepatic portal venous obstruction; MELD, Model for End-Stage Liver Disease; MR, magnetic resonance; MRE, magnetic resonance elastography; MRI, magnetic resonance imaging; NCPF/IPH, noncirrhotic portal fibrosis/idiopathic portal hypertension; NCPH, noncirrhotic portal hypertension; TIPS, transjugular intrahepatic portosystemic shunt.

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TABLE 1 Portal and Hepatic Venous Pressure Measurements

Cause	FHVP	WHVP	HVPG
Prehepatic	Normal	Normal	Normal
Hepatic presinusoidal	Normal	Normal to increased	Normal to increased
Hepatic sinusoidal	Normal	Increased	Increased
Hepatic postsinusoidal	Normal	Increased	Increased
Posthepatic	Increased	Increased	Normal to increased

Adapted from Berzigotti et al¹⁰ and Kirby et al.¹⁴

Abbreviations: FHVP, free hepatic vein pressure; HVPG, hepatic vein pressure gradient; WHVP, wedged hepatic vein pressure.

Figure 1 Digital subtraction angiography (DSA) image demonstrating patent TIPS (white thin arrow) connecting the right portal vein (black thin arrow) to the right hepatic vein (black thick arrow) through the liver parenchyma. Embolization coils are present in the coronary vein (white thick arrow) due to persistent reflux into varices.

prior to shunt placement.² Multiphase CT and MR can confirm the presence of EHPVO, which will demonstrate numerous periportal venous collaterals that enhance in the portal venous phase and possibly linear calcifications within a thrombosed portal vein, suggesting chronic thrombosis.⁷

Contrast-enhanced CT and MRI are also helpful in differentiating NCPF/IPH from cirrhosis. In a retrospective study conducted by Glatard et al, the following CT findings were observed significantly more frequently in NCPF/IPH than in cirrhosis: extrahepatic portal vein thrombosis, intrahepatic portal vein abnormalities (such as reduced caliber, occlusive thrombosis, and non-visualization), focal nodular hyperplasia-like nodules, and perfusion defects.⁸ Conversely, the combination of hypertrophy of the caudate lobe, atrophy of segment IV, and nodular hepatic contour were seen significantly more often in cirrhosis.⁸

Figure 2 DSA image demonstrating patent DIPS (white thin arrow) connecting the portal vein (black thin arrow) to the inferior vena cava (IVC) (white thick arrow) through the caudate lobe of the liver. Also visualized is the proximal splenic vein (black thick arrow) and superior mesenteric vein (SMV) (white bent arrow).

Hepatic fibrosis has also been assessed using magnetic resonance elastography (MRE), which utilizes shear waves delivered to the liver from an external generator. The liver is then imaged with a gradient echo MRE pulse sequence, and reconstructed images display stiffness of the liver. MRE has advantages over ultrasound elastography, because the entire liver may be easily imaged without acoustical window limitations.⁹ Thus far, MRE has not been utilized for assessment of liver disease associated with NCPH.

Portal Hemodynamics

The onset of clinical symptoms related to portal hypertension provides the impetus for invasive studies to accurately document portal pressures and to provide a roadmap for intervention planning. Contrast portal venography obtained during venous phase of an arteriogram or portal phase CT can confirm portal vein occlusion and cavernous transformation or other collaterals. Alternatively, portal venography may be obtained by wedged hepatic vein injection with iodinated contrast material or carbon dioxide. Hepatic vein-to-vein communications (HVVC), identified on hepatic venography, should raise suspicion of NCPH etiologies.⁶

The hepatic sinusoidal pressure (wedged hepatic vein pressure [WHVP]), which reflects the portal pressure, can be estimated by transiently occluding a hepatic vein, using a balloon catheter with a transducer introduced via the internal jugular vein. Normal hepatic vein pressure gradient

Figure 3 (A) Coronal contrast-enhanced CT image demonstrating a chronically occluded portal vein (white thick arrow) with cavernous transformation (white thin arrow). The superior mesenteric vein (black thick arrow) is widely patent. (B) DSA image demonstrating a patent mesocaval shunt (white thin arrow) connecting the superior mesenteric vein (black thin arrow) to the IVC (white thick arrow). A pigtail flush catheter is in place in the superior mesenteric vein.

Figure 4 (A) Axial contrast-enhanced computed tomography (CT) image demonstrating marked gastric varices (white arrow). (B) DSA with balloon occlusion retrograde venography of the gastroduodenal shunt (GRS) (white thin arrow) demonstrates opacification of gastric varices (white thick arrow) and the inferior phrenic vein (black thin arrow). The GRS is catheterized via the left renal vein from the IVC. (C) Fluoroscopic image with balloon occlusion retrograde venography demonstrates opacification of the gastric varices (white thin arrows). (D) DSA from GRS (white thin arrow) after embolization of the gastric varices with sclerosant and coils (white thick arrow) demonstrates persistent opacification of the inferior phrenic vein (black thin arrow).

(HVPg) is less than 5 mm Hg and represents the difference between WHVP and free hepatic vein pressure (FHVP). Clinical complications of portal hypertension including varices, ascites, or hepatorenal syndrome are not observed until the HVPg equals or exceeds 10 mm Hg.¹⁰ In addition, good correlation exists between stiffness values, presence of esophageal varices, and abnormal HVPg.¹¹ A HVPg less than 10 mm Hg with clinical sequelae of portal hypertension should raise suspicion of NCPH such as from IPH or chronic portal vein thrombosis.⁶ Compared with sinusoidal portal hypertension/cirrhosis, which demonstrates an elevated HVPg, presinusoidal portal hypertension from IPH often has a normal HVPg¹⁰ (Table 1).

Minimally Invasive Interventions

The major indications for portal vein decompression in the setting of PH are refractory variceal hemorrhage and ascites. Historically, surgically placed portosystemic shunts have been effective but associated with operative morbidity. Percutaneous transjugular intrahepatic portosystemic shunts (TIPS) offer the additional advantage of adjustable flow rates after placement to address hemodynamic requirements of individual patients. In NCPH, TIPS is generally reserved for patients with variceal hemorrhage refractory to endoscopic management. Percutaneous portosystemic shunt creation is generally contraindicated in clinical settings such as hepatic encephalopathy, right-sided heart failure (pressure >20 mm Hg), uncontrolled sepsis, and marked hepatic insufficiency (bilirubin >3 mg/dL). Patients with NCPH are known to have better hepatic function and lower incidence of encephalopathy after surgical portosystemic shunts. TIPS-related mortality has been shown to relate to Model for End-Stage Liver Disease (MELD) category; low-, intermediate-, and high-risk MELD classifications have shown a 3-month overall mortality post-TIPS approaching 15%, 33%, and 80%, respectively.^{12,13}

Three major percutaneous shunt types have been described. The greatest clinical experience exists with TIPS from the hepatic vein to the portal vein. Disadvantages include potential resultant functional deterioration, hepatic encephalopathy, and need for surveillance imaging. Direct intrahepatic porto-

caval shunts (DIPS) connect the portal vein and IVC via the caudate lobe of the liver. DIPS is advantageous in the setting of hepatic vein or intrahepatic parenchymal tract unsuitability for TIPS such as in Budd-Chiari syndrome and polycystic disease of the liver. However, DIPS may interfere with liver transplantation. Percutaneous mesocaval shunts are the most recent methodology, although clinical experience is limited. These shunts connect the mesenteric vein and IVC and can be employed in the setting of portal vein occlusion but are the most technically challenging. They preserve hepatopetal blood flow and do not affect subsequent transplantation.¹³

Sinistral (left-sided) portal hypertension with isolated gastric varices and portal and/or splenic vein occlusion is considered a relative contraindication to TIPS. Of note, gastric varices may bleed at portosystemic pressure gradients less than the 12 mm Hg target residual gradient following TIPS. Potential causes include Crohn's disease post-proctocolectomy and pancreatitis. The gastric varices may be treated by balloon occlusion retrograde transvenous obliteration (BRTO) with sclerosant embolization via spontaneous portosystemic shunts such as a gastroduodenal shunt. Advantages over TIPS include preservation of liver function, improved hepatopetal flow, and prevention of encephalopathy. Surveillance is important, because BRTO may potentiate esophageal varices.^{14,15}

Hepatic or portal vein recanalization with angioplasty or stenting and partial splenic embolization are other potential therapies in the setting of TIPS contraindication and certain clinical scenarios.^{14,16,17}

Summary

Imaging modalities can greatly assist in the diagnosis, classification, and management of NCPH-related disorders. Further research is needed to establish the role of novel modalities in the evaluation of this condition. [Corrections to figures, references, author list and affiliations added to online publication 11 October 2016.]■

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