

Radiological Diagnosis of Portal/Mesenteric Vein Occlusion

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Keywords

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Summary

Background: In contrast to an acute occlusion of the visceral arteries, which is the most important differential diagnosis for an occlusion of the portal venous system and which poses a highly dangerous situation ending in gangrene of the bowel wall, the symptoms of an acute occlusion of the portal venous system are quite unspecific. To rule out an acute arterial occlusion, diagnostic evaluation has to be carried out quickly in order to decide on the necessity of therapeutic steps concerning a recanalization of the occluded vessels. Only few therapeutic options are available to recanalize and remodel the portal venous system, depending on the underlying disease, the age of the occlusion, its extension, and the effect on the bowel wall, stomach, spleen, and abdominal wall. Moreover, the efficacy of recanalization procedures mainly depends on the formation and number of collateral venous blood supply, its degree, and the anatomic structure. Possible complications of portal hypertension like varices, gastrointestinal vasculopathy, ascites, and splenomegaly also influence the success of recanalization procedures. Only in cases of acute thrombotic occlusion systemic lytic therapy promises to be successful. Therefore, other options such as transjugular intrahepatic recanalization, e.g. by means of the TIPS (transjugular intrahepatic portosystemic shunt) procedure, have to be evaluated. **Methods:** Review of the literature. **Results:** Noninvasive methods such as ultrasound (US), computed tomography, and especially magnetic resonance imaging (MRI) allow the evaluation of therapeutic options as well as their success, the feasibility of technical procedures, the detection of possible risks, and a calculation of risks and benefits. **Conclusion:** In order to arrive at the correct therapeutic decision, a combination of MRI and US methods combined with color Doppler guarantee the most efficient diagnostic results in cases with acute or chronic occlusions of the portal venous system.

Schlüsselwörter

Pfortaderverschluss · Diagnostik

Zusammenfassung

Hintergrund: Während ein akuter Verschluss der Viszeralarterien die wichtigste Differenzialdiagnose zum portalvenösen Verschluss darstellt und innerhalb von Stunden in einer Darmgangrän enden kann, sind die Symptome beim akuten Verschluss des Portalvenensystems eher unspezifisch. Beim Ausschluss einer arteriellen Ursache spielt die Diagnostik in erster Linie eine Rolle in der Beurteilung der technischen Machbarkeit und therapeutischen Notwendigkeit der Rekanalisierung des Portalsystems. Die Wahl einer der wenigen verfügbaren Therapieoptionen ist abhängig von der Ursache, dem Alter, dem Ausmaß des Verschlusses und den durch die venöse Abflussbehinderung bedingten Auswirkungen auf die vorgeschalteten Organe wie Darm, Magen, Milz und Bauchwand. Ebenso therapieentscheidend sind die Zahl, das Ausmaß und die anatomische Zuordnung der Kollateralvenen sowie die Komplikationen der portalen Hypertension wie Varizen, gastrointestinale Vasculopathie, Aszites und Splenomegalie. Da nur bei sehr frischen thrombotischen Verschlüssen eine systemische Lysetherapie erfolgreich sein kann, muss die mögliche Indikationsstellung anderer Therapieoptionen, insbesondere zur transjugulär-intrahepatischen Rekanalisation über den TIPS (transjugulärer intrahepatischer portosystemischer Shunt)-Zugang, abgewogen werden. **Methoden:** Literaturübersicht. **Ergebnisse:** Nichtinvasive Verfahren wie Sonographie, Computertomographie und insbesondere Magnetresonanztomographie (MRT) erlauben eine Abwägung der therapeutischen Möglichkeiten, der technischen Durchführbarkeit, der möglichen Risiken, des Erfolgs sowie des Risikos bzw. des Nutzens. **Schlussfolgerung:** Für die korrekte Therapieentscheidung erscheint eine Kombination von MRT und Sonographie einschließlich Farbdoppler als die aussagekräftigste Diagnostik bei akuten oder chronischen Verschlüssen des Portalvenensystems.

Introduction

A large variety of underlying causes and effects of stenoses or occlusions of the splanchnic veins does exist. The clinical presentation of portal and/or mesenteric vein thromboses depends on how acutely the obstruction and its underlying cause develop. A wide spectrum of clinical presentations may occur, ranging from nonspecific abdominal pain [1] to complications of portal hypertension such as splenomegaly, varices, and massive ascites. If the thrombosis is due to septic pylephlebitis caused by an acute gastrointestinal inflammation, the disease may present with severe intestinal infarction or massive variceal hemorrhage. The severity of mesenteric occlusion depends on whether the occlusion results in transient mucosal changes or in a more severe transmural wall necrosis. In such cases, a clinical differentiation between an acute arterial mesenteric occlusion, in most cases caused by an embolism, and an acute mesenteric/portal venous thrombosis is not possible.

Caused by an increase of intravascular volume and an elevation of hydrostatic pressure, a concentric bowel wall thickening and a mesenteric edema may result if the collateral draining veins are inadequate. Intravascular flow could also be forced into the mesenteric space and other peritoneal spaces as ascites [2]. Hemorrhagic necrosis of the affected bowel may subsequently occur [3] but not in the same way as in cases with acute arterial mesenteric occlusion [4].

A special type of portal/mesenteric vein occlusion may occur in infants following umbilical catheter placement in the newborn period. They may present with ascites and major variceal hemorrhage or may suffer from a vascular emergency as the first manifestation of vein occlusion at the age of 8–10 years.

Due to the anatomy of the portal venous system, a direct arterial or venous approach via a catheter to the splanchnic veins is not possible. The diagnostic procedures are predominantly limited to ultrasound (US), computed tomography (CT), and magnetic resonance imaging (MRI). As a second-line possibility, invasive angiographic methods by means of an arterial catheter approach or direct needle puncture via transjugular transhepatic access are very promising procedures enabled by the transjugular intrahepatic portosystemic shunt (TIPS) (table 1).

Table 1. Diagnostic methods in cases of mesenteric/portal occlusion

<i>Non-invasive diagnostic imaging procedures</i>	
Sonographic methods (gray-scale/color Doppler)	
Computed tomography (multi-slice helical CT)	
Magnetic resonance imaging (MRI)	
<i>Invasive diagnostic imaging methods by angiography</i>	
Indirect access	
Indirect splenoportography	
Indirect mesentericportography	
Direct access	
Direct splenoportography	
Transjugular transhepatic portography	

In order to choose the appropriate therapeutic approach, it is necessary to know the exact underlying cause and the degree of the expansion of the occlusion. In cases of thrombosis, the age of the thrombus and the effects on the stomach, bowel, spleen, and abdominal wall caused by the vein occlusion must be evaluated. Because of the limited number of therapeutic procedures, the possibility of a minimally invasive interventional procedure, e.g. via a transjugular transhepatic approach, has to be discussed.

Noninvasive Diagnostic Imaging Modalities

Ultrasound

Sonography is the primary imaging modality for the diagnosis and follow-up of portal and mesenteric vein thrombosis. The examination is easy to perform. The sensitivity and specificity of detecting a thrombus of the main portal vein varies from 66 to 100% [5, 6]; this can even be improved by combining it with color Doppler studies (sensitivity 100%, specificity 93%) [1]. The use of an intravenous echo signal enhancer is not required. The vein thrombosis manifests as an intraluminal echogenic lesion that can partially or completely obstruct the vein, thus obscuring normal portal vein landmarks. The lumen of the vein may be expanded in acute thrombosis or when a tumor thrombus (hepatocellular carcinoma (HCC)) is present. However, depending on the age of the thrombus, clots exhibit variable echogenicity and may be hypoechoic (recent thrombus) or iso- and hyperechoic (elderly thrombus) in comparison to the vessel wall. Due to the similar background echogenicity of the liver, the diagnosis of an intrahepatic portal vein thrombus is more difficult to establish confidently. In such cases, the use of Doppler sonography, especially color flow Doppler sonography, allows the diagnosis of an occlusion of the veins by an elimination of the venous flow signal (fig. 1). It permits a quick diagnosis and reveals collateral flow such as cavernous transformation and varices and abnormal hepatofugal flow in the case of spontaneous splenorenal shunt that may be invisible on gray-scale images. Simultaneously, a tumor thrombus can be differentiated from a bland thrombus by demon-

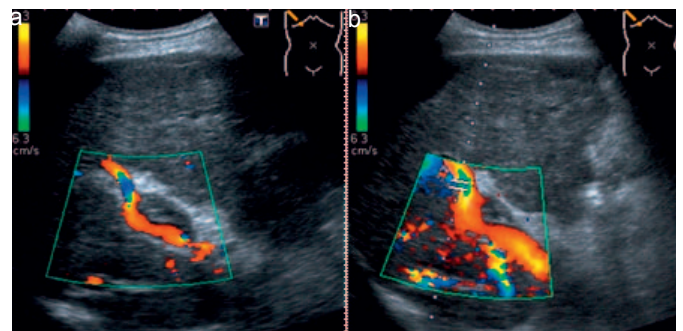


Fig. 1. a,b. Portal vein occlusion caused by tumor thrombus. Multiple small arterial vessels in the thrombus to verify the tumor infiltration by color Doppler sonography.

Fig. 2. Chronic portal vein occlusion in a 9-year-old girl caused by an umbilical vein catheter in a newborn period. Acute life-threatening upper gastrointestinal bleeding by rupture of esophageal varices. Multiple collaterals and cavernous transformation. **a** In gray-scale, hypoechoic, and variable echogenic, mainly hyperechoic old thrombosis. **b** The color Doppler sonography shows the irregular flow in the transformed veins. **c** Transjugular transhepatic angiography (DSA) before recanalization and TIPS.

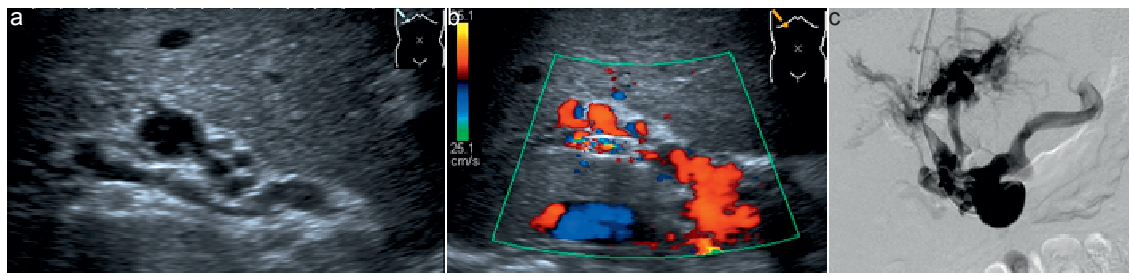
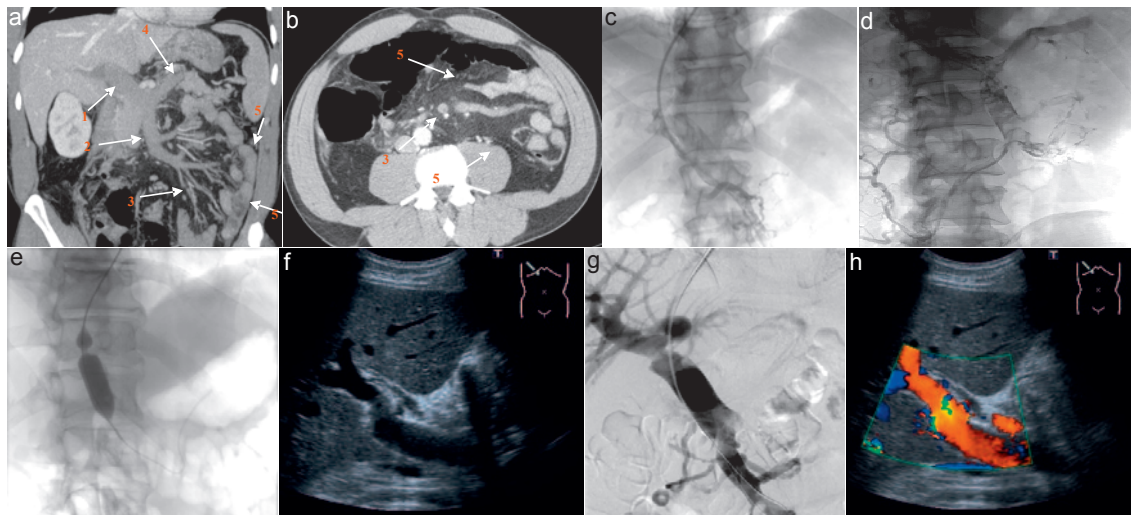


Fig. 3. 50-year-old man with acute abdominal pain. **a, b** Multi-slice, contrast-enhanced helical CT, axially and in coronal reconstruction revealing a large recent thrombosis occluding the intra- and extrahepatic portal vein (1), the main vessel (2), and the left branches (3) of the superior mesenteric vein and the splenic vein (4), and the thickening bowel wall by intramural venous engorgement (5). **c, d** Angiography via a transjugular transhepatic approach before recanalization and TIPS. **e** Fluoroscopic balloon dilatation of a strong stenosis of the portal vein. **f-h** Gray-scale US and color Doppler US in comparison to the angiography after recanalization.



strating color Doppler flow and arterial waveforms (often hepatofugal in direction) within the tumor thrombus [7, 8]. Importantly, lack of flow does not exclude tumor thrombus. Pulsatile flow is fairly specific for a malignant thrombus, and likewise is the verification of arterial vessels (fig. 1) and echo enhancement by using US contrast medium [9]. However, US cannot only distinguish between tumor extension into the portal vein and extrinsic vein compression in the liver but can also evaluate the inflammatory status or malignancies in other organs such as the pancreas or the bowel.

In chronic thrombosis, the portal vein system can be normal in size and hyperechoic, small, or vanished (fig. 2). Bridging collaterals may therefore develop in the hepatic hilus. These vessels are often quite prominent in the case of cavernous transformation of the portal vein, which occurs in 30% of the patients with portal vein occlusion [10]. A typical continuous, low-frequency portal venous flow pattern is found in these patients by Doppler examination of these vessels.

Furthermore, sonographic signs of thrombosis of the portal vein system are often splenomegaly, ascites, venous collaterals, and intestinal congestion or infarction. Serial examinations are useful in these patients to determine the efficacy of e.g. lytic or anticoagulative therapeutic procedures or to mon-

itor the development of cavernous transformation after acute portal vein thrombosis (fig. 3).

An interventional, radiological, minimally invasive therapy via the transjugular transhepatic approach essentially intends to recanalize the portal/mesenteric venous system and allows the preinterventional evaluation of target vessels (visibility of the (hypoechoic) intrahepatic portal branches) to control the correct guiding of the needle during the puncture into the occluded portal vein as well as the correct guidance of the guide wire into the portal branch by means of US.

Computed Tomography

Contrast-enhanced CT is considered to be the 'gold standard' in the diagnostics of the portal venous system because of its lower costs compared to MRI, its wide availability, and its excellent sensitivity in diagnosing and differentiating between arterial infarction by embolism or nonocclusive mesenteric ischemia and venous thrombosis. Technical advances in volumetric multislice CT scanning performed with helical multislice CT have facilitated the development of CT angiography. This technique permits the evaluation of vascular structures, the bowel wall, and the adjacent mesentery and organs. Sensitivity and specificity rates amount to at least 90% [11]. CT

findings of portomesenteric venous thrombosis include persistent, well-defined intraluminal filling defects with central low density, which may be surrounded by well-defined, rim-enhancing venous walls. Accompanying collateral circulation, engorgement of mesenteric veins, and mesenteric edema may be present [12]. Multivein occlusion is common in splanchnic venous thrombosis.

Portal vein thrombosis appears as a low-density central zone surrounded by an intensely enhanced periphery on contrast-enhanced scans. It is not entirely clear whether this peripheral enhancement is due to flow around the clotted material or enhancement of the vasa vasorum of the portal vein wall. There is also transient inhomogeneous enhancement of the periportal hepatic parenchyma. Enlargement of the occluded vein increases the likelihood of the presence of tumor thrombus. There may also be streaky enhancement of the clots, which correlates with the angiographic finding of 'threads' with septic thrombophlebitis and 'streaks' seen in tumor thrombus. On precontrast scans, the contents of the portal vein may be high in attenuation because of the high protein content of concentrated red blood cells [1].

Bowel wall thickening is the most common manifestation of accompanying bowel ischemia caused by intramural venous engorgement [13]. This nonspecific finding may manifest as a target sign, with alternating intramural areas of high and low density resulting from submucosal edema or hemorrhage [14]. The triad of low density in the mesenteric vein, thickening of the bowel wall, and especially the presence of intraperitoneal fluids highly suggests a bowel infarction.

Arteriportal shunting is another indirect sign of chronic portal vein obstruction. In these cases, enhancement of the involved portal vein branch occurs early in the arterial phase. Calcification of the portal vein thrombus is also readily detected by CT.

Other important CT findings including bowel dilatation which reflects aperistaltic activity, absent or poor enhancement of the bowel wall secondary to both venous and arterial perfusion abnormalities, and, less common, intestinal pneumatosis as a so-called 'signum mali ominis' are more specific signs of ischemic bowel disease caused by an arterial occlusion. The appearance of mesenteric or portal venous gas suggests a dissection of intramural gas into the venous system by bowel infarction. Free intraperitoneal air from the perforation of an affected bowel segment may also be seen in a later state of infarction.

Dynamic evaluation of the splanchnic veins may be performed prospectively with dedicated CT angiography during the venous phase of the bolus injection. CT angiography provides exquisite anatomic detail and reveals intra- and extraluminal abnormalities, intimal calcifications, mural thrombosis, and mesenteric edema. Hepatopetal extension of thrombosis into the intrahepatic portal venous system may also be detected, permitting an assessment of the severity of disease and an initiation of the appropriate management. Furthermore, the remaining extravascular organ anatomy is exquisitely depicted.

Magnetic Resonance Imaging

MRI has been proven to be an excellent diagnostic tool for the visualization of vessel occlusions of the portal venous system. In comparison to US and CT, MRI offers a simultaneous demonstration of the vessel occlusion and thus a better diagnostic visualization of its possible underlying cause, e.g. in patients with HCC, liver cirrhosis, and pancreatitis. A large variability of sequences to visualize thrombi and occlusions of the portal venous system does exist. Generally, there are conventional MRI sequences as well as sequences especially for magnetic resonance angiography (MRA). Besides the common sequences like T2 spin echo or T1 gradient echo, susceptibility-weighted sequences (T2*) for the direct visualization of the thrombus have been published, although the same sequence for the demonstration of thrombi has been used long since in the neurovascular system [15].

The analysis of variations of signals in T1- and T2-weighted sequences allows defining the age of thrombus formations. Moreover, the increase of signal intensity in both sequences in comparison to the liver parenchyma or the surrounding muscle tissue is mostly due to fresh thrombus material. Fresh thrombi fewer than 5 weeks old seem to show higher signals both in the T1- and T2-weighted sequences. The decrease of signal intensity in T1-weighted images is mainly seen in chronic occlusions [1]. Diffusion-weighted imaging (DWI) sequences are an adequate tool for imaging abdominal anatomic structures, and recent studies on the differentiation of HCC-induced thrombi and normal thrombi reveal the diagnostic possibilities, although controversial results have been reported in the recent literature [16, 17].

For imaging vascular structures by means of MRA, different techniques have been reported. Non-contrast-enhanced MRA and conventional sequences such as time-of-flight and phase-contrast techniques, the 3D half-Fourier fast spin echo, and true steady-state free-precession (SSFP) in combination with two time-spatial labeling inversion pulses (T-SLIPs) have been proven as an effective method for the direct visualization of intra- and extrahepatic thrombi of the portal venous system [18].

In several studies, contrast-enhanced MRA, especially the quick 3D technique, has been shown to be a precise imaging modality with comparable diagnostic accuracy when compared with invasive conventional digital subtraction angiography (DSA) and other methods for the visualization of the portal venous system [19–21].

In cases in which an accumulation of the contrast medium in the wall of the thrombosed venous system can be demonstrated, this can be interpreted as a sign of older thrombi with partial recanalization or as the existence of septic thrombotic material. The enhancement of the contrast medium in thrombotic material is an important diagnostic sign in cases of a HCC-induced thrombus.

Last but not least, the excellent soft tissue contrast and the focal change of the liver tissue with wedge-shaped hyperintensive change in T2-weighted sequences induced by the obstruc-

Table 2. Necessary MRI sequences for the diagnostics of PVT

Thrombi/Occlusion	MRI sequence		
	T1w	T2w	T1w with CM (e.g. 3D VIBE)
Age	+++	+++	+++
Expansion	++	++	+++
Collateral	+	+	+++
Surrounding effect	+	+++	+++
Underlying cause, e.g. compression tumor, inflammation	+	+++	+++

tion of the venous drainage in cases of intrahepatic thrombus can be interpreted as a typical sign of a focal liver tissue infarction or edema. Accordingly, a transient increased arterial perfusion in regions with reduced portovenous flow due to the existence of thrombi and venous occlusions is visualized in the arterial vessel phase of contrast-enhanced sequences.

Moreover, MRI allows for a better visualization of intramural edema of the bowel wall and a reduced uptake of the contrast medium in cases of intestinal infarction following vein occlusion. The visualization of motility of the bowel loops in the area of infarction is also possible with the so-called real-time MRI.

The lack of X-ray exposure and the marked decrease of nephrotoxicity of gadolinium are well-known advantages compared to CT. The general disadvantages of MRI are its limited availability and the high cost of the investigations. Diagnostic disadvantages of MRI are motion-induced artefacts in cases of noncompliant patients and also in patients with a high quantity of ascites combined with the movements of the small bowel. An additional diagnostic failure is seen in those patients with implanted stent material.

A precise selection of the patient group and the intensive instruction of the patients prior to the investigation are essential for an optimized MR diagnostics (table 2, fig. 4).

Invasive Diagnostic Imaging: Conventional Angiography

Diagnostic angiographic procedures include conventional methods such as direct and indirect portography. Although these techniques are invasive, they may be limited by flow dynamics and require the use of potentially nephrotoxic iodinated contrast material and ionizing radiation. Diagnostic conventional angiography is usually reserved for cases in which clinically suspected portomesenteric venous thromboses cannot be visualized with noninvasive modalities. Especially direct invasive procedures, such as transcatheter delivery of thrombolytic agents, mechanical thrombectomy, balloon dilatation, or stent implantation including the TIPS procedure in selected patients, can be combined with endovascular therapeutic maneuvers to unblock occluded veins.

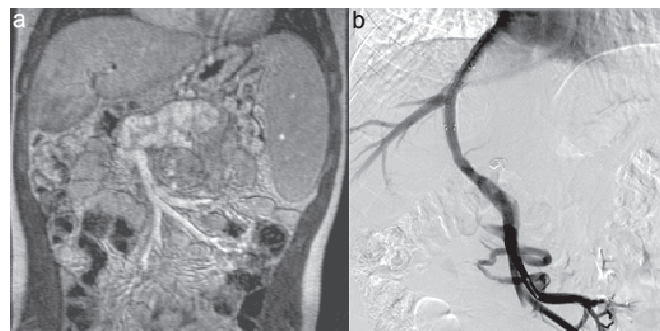


Fig. 4. a, b. 16-year-old boy with portal venous occlusion caused by an umbilical vein catheter in the newborn period. Multiple episodes of life-threatening variceal bleeding in spite of surgical meso-Rex and Warren shunt. T1 weighted with contrast medium shows an old coronary occlusion and large abdominal collaterals. Angiography after recanalization of the portal vein and TIPS.

Two types of angiographic techniques for the visualization of the spleno-mesenterico-portal system are possible:

- The indirect spleno-mesenterico-portography involves selective arterial injections of iodinated contrast medium into the superior mesenteric artery or splenic artery followed by delayed imaging, which was previously used as the preferred nonsurgical approach for confirming splanchnic venous thrombosis. In contrast to direct portography, this method is less invasive; however, the mesenteric or splenic arteriography with delayed venous phase imaging may not always demonstrate the exact cause and the expansion of the venous occlusion or an intraluminal thrombus. Nevertheless, the collateral veins, e.g. varices and spontaneous (splenoportal) shunts, can be clearly visualized in most cases.
- Direct portography includes percutaneous transhepatic portal venography, transjugular portography, and direct splenoportography. Direct splenoportography with an injection of contrast medium directly into the parenchyma of the spleen via a percutaneous needle puncture was established in the seventies of the last century to explore the portal system but has been completely deserted meanwhile. More and more, the transjugular transhepatic approach has taken over as the therapeutic procedure of choice. Complications such as intra-abdominal bleeding, which is a complication of percutaneous transhepatic direct portography or therapy, are not observed.

Discussion and Evaluation

Many diagnostic methods are available or have been adapted to the therapeutic needs of the patients with thrombosis of the portal system. However, the efficiency and the diagnostic value of these methods differ immensely. It is mandatory to know the value of these methods, which are highly complicated and combined with a major risk of complications,

Table 3. Diagnostic methods concerning significance in portal/mesenteric occlusion

	Ultrasound	Color Doppler	CT	MRI	Angiography
Cause of occlusion	++	++	++	+++	(+)
Expansion	++	++	+++	+++	++
Age	+++	(+)	++	+++	+
Collaterals	++	+++	+++	+++	+++
Surrounding effects	++	+	+++	+++	(+)

CT = Computed tomography; MRI = magnetic resonance imaging.

when planning a minimally invasive radiological procedure or an operation. Therefore, CT, MRI, and US are the most efficient tools to answer the following questions: i) is a therapy necessary or not, ii) which therapeutic step is the most efficient one, iii) conservative ‘watch and see’, iv) operation or minimally invasive interventional radiological therapeutic procedure? The domain of US is the evaluation of the onset of occlusions resulting in the formation of collateral venous blood supply, its extension, and the velocity of flow conditions in the occluded vessels.

Other diagnostic modalities such as CT and MRI have the potential to visualize the underlying cause and the effects of vessel thromboses on organs and their surrounding areas and tissues.

Although contrast-enhanced CT as a fast diagnostic tool is available nearly everywhere nowadays and accepted as the gold standard, in comparison to MRI, not all questions can be answered by means of CT (table 3).

Conclusion

In order to answer all questions regarding suspected venous occlusions in clinical and diagnostic practice, a combination of MRI and US methods combined with color Doppler guarantees the most reliable diagnostic results in cases with acute or chronic occlusions of the portal venous system. Not only portal and splanchnic veins into the peripheral branches but also the splenic veins, and therefore the effect on neighboring structures in cases of acute and chronic occlusions, can thus be optimally visualized and evaluated.

Disclosure Statement

No conflicts of interest for any of the authors.

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