Diagn Interv Radiol 2009; 15:182–187

ORIGINAL ARTICLE

Intrahepatic portosystemic venous shunts: radiological evaluation

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PURPOSE
The purpose of our study was to evaluate the imaging findings of intrahepatic portosystemic venous shunts (IPSVS) in asymptomatic patients.

MATERIALS AND METHODS
Between 2002 and 2008, we examined 8 patients with IPSVS which were found incidentally. Diagnosis was based on ultrasonography, computed tomography and magnetic resonance imaging findings. Three patients had a history of liver cirrhosis without symptoms of encephalopathy.

RESULTS
Most IPSVS were located in the right liver lobe (7 cases) and in one case in the left liver lobe. Identification of type of shunt between portal and systemic veins was based on Park's classification. Type III shunts were found in five patients and type I in three patients.

CONCLUSION
IPSVS is a rare vascular abnormality that is usually asymptomatic. Radiologists must be aware of these communications because IPSVS may be an incidental finding in imaging control for unrelated reasons.

Key words: • *portosystemic shunts* • *ultrasonography* • *computed tomography, X-ray* • *magnetic resonance imaging*

Intrahepatic portosystemic venous shunts (IPSVS) are relatively rare anomalies that are usually asymptomatic. They are defined as communications between an intrahepatic portal vein and a systemic vein via an anomalous intrahepatic venous channel. Shunts may be discovered incidentally during imaging for an unrelated disease. A few patients may present with hepatic encephalopathy because of a high degree of shunting.

Materials and methods

In our patients, 4 men and 4 women, IPSVS were found incidentally during a six-year period (from May 2002 to June 2008). The average age of the patients was 59.25 years (range, 18–80 years). The reasons for imaging evaluation were liver cirrhosis (n = 3), anemia (n = 1), choledocholithiasis (n = 1), right upper quadrant pain (n = 1), diffuse abdominal pain (n = 1), and a calcified liver cyst (n = 1) (Table).

Three patients were imaged with ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI). Two patients had imaging procedures with both US and MRI, and three underwent imaging with helical CT only (Table). Sonographic examinations were performed with a Siemens Sonoline G60S scanner (Siemens Medical Systems, Erlangen, Germany) (probe, convex C6-2) in both gray-scale and color Doppler. CT scans were performed with a Picker PQ 5000 scanner. Images were obtained before and after a bolus injection of 150 mL (3-4 mL/s) of non-ionic contrast medium (iopromide, Ultravist 300, Schering, Germany) in arterial and portal phase with slice thickness, 5 mm; pitch, 2; reconstruction interval, 5 mm; and scan time, 1 second. MRI examinations of the abdomen were performed with a Siemens Expert Plus 1T device (Siemens Medical Systems, Erlangen, Germany). T2-HASTE images (TR, 6 ms; TE, 60 ms) and T1-FLASH images (TR, 11 ms; TE, 4.2 ms) after intravenous administration of 10 mL of contrast agent (gadopentetate dimeglumine, Magnevist, Schering, Germany) were obtained in axial and coronal planes, with a slice thickness of 8 mm. In selected cases (2 patients), T2-TRUFI images (TR, 10.2 ms; TE, 4.7 ms) in the axial plane and T1-weighted enhanced 3D gradient-echo sequence (TR, 6.4 ms; TE, 2.4 ms) in the coronal plane were also performed.

Results

In five patients, communication between portal and systemic veins was through a vascular lesion (aneurysm). The aneurysm was located in the right lobe of the liver in four cases and in the left lobe in one case. The size ranged from 20 mm to 40 mm in diameter (average, 32 mm). Connection was between the right branch of portal vein and the inferior vena cava in two cases and between the right branch of portal vein and the right hepatic vein in other two cases. The aneurysm, lo-

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Received 16 June 2008; revision requested 28 December 2008; revision received 19 January 2009; accepted 22 March 2009.

cated in the left liver lobe, connecting the left branch of portal vein with the inferior vena cava (Table). US showed a rounded hypoechoic lesion. On color Doppler images, flow was demonstrated within the lesions. On CT, the lesions (aneurysms) were hypodense on precontrast images, demonstrated intense enhancement on postcontrast images, and showed direct communication between branches of the portal vein and the systemic veins. On MRI, the lesions had low signal intensity on T2-weighted (T2W) images and were strongly enhanced on T1-weighted (T1W) images after contrast administration (Figs. 1–4).

Three patients had tubular lesions connecting the right portal vein with the inferior vena cava (one case) and peripheral branches of the portal vein with the inferior vena cava (two cases). These tubular connections were isoattenuating to the hepatic veins on CT and isointense to the hepatic veins on MRI (Fig. 5).

Discussion

Portal to systemic venous communications are classified as extrahepatic and intrahepatic. Extrahepatic communications are usually seen in patients with portal hypertension from cirrhosis and are commonly through the coronary vein, esophageal varices, or retroperitoneal collaterals. Intrahepatic communications are found between intrahepatic portal veins and systemic veins and are less common than extrahepatic communications.

The presence of IPSVS was first described by Raskin et al. in 1964 (1). Although intrahepatic shunts are rare vascular abnormalities, advances in diagnostic imaging techniques have resulted in an increased number of cases reported. According to the location of the communicating systemic vein and pathogenic mechanism, IPSVS are subdivided into two main types (2, 3). The first (internal type) is a shunt that consists of an intrahepatic portal venous-hepatic venous pathway, whereas the other (external type) is a shunt that consists of an intrahepatic portal venous-perihepatic venous pathway. In the internal type, the shunt is depicted as tubular or aneurysmal communication (single or multiple) between intrahepatic portal veins and hepatic veins. The external type communicates between the intrahepatic portal vein and perihepatic veins and drains into the inferior vena cava. Direct communication between the right portal vein and the inferior vena cava around the right lobe is also included in this category.

Park et al. (4) classified published cases of IPSVS into the following types: (1) a single large tube of constant diameter connecting the right portal vein to the inferior vena cava, (the most common), (2) a localized peripheral shunt with single or multiple communications between the peripheral branches of the portal and hepatic veins in one hepatic segment, (3) a connection between peripheral portal and hepatic veins through an aneurysm, and (4) diffuse and multiple communications between peripheral portal and hepatic veins in both lobes.

According to Chevallier et al., IPSVS measuring more than 1 mm in diameter are called macroscopic intrahepatic shunts (5). Based on anatomic, clinical, and pathophysiological criteria, Chevallier et al. divided these shunts into four different types. Type I consists of patent paraumbilical veins located in the liver and are found in patients with portal hypertension. They communicate between the portal vein and systemic venous system. Type II includes isolated or multiple communications between a portal venous branch and a hepatic venous branch that involve two contiguous liver segments. Type III shunts provide multiple communications between portal and hepatic venous branches found in liver segments that are not contiguous. Type IV corresponds to a tubular communication between the right portal vein and the inferior vena cava (5). Small type II and III shunts may be difficult to differentiate from small hypervascular liver lesions. In these difficult cases, only portography should be considered as a pretherapeutic procedure.

According to Park et al., in 14 cases reported in the literature, a single large tube connecting the portal vein to the inferior vena cava was the most common type and was found commonly in cirrhotic patients (4). Tanoue et al. and Remer et al. found that more than 50% of the shunts in their series had aneurysmal communications (3, 6).

The pathogenesis of IPSVS is controversial. Some authors believe that IPSVS is congenital, caused by a persistent embryonic venous anastomosis. When portosystemic communication is confirmed in a patient without history of liver disease or trauma, a congenital origin is presumed (6–9). Others support the acquired nature of

Table. Demographic characteristics of patients and location, size, and types of intrahepatic portosystemic venous shunts

Patient	Age	Sex	Type (Park)	Location	Size	History	Examination
1	25 y	M	III	RLL	20 mm	Biliary cirrhosis	US, CT, MRI
2	80 y	F	III	LLL	36 mm	Choledocholithiasis	US, CT, MRI
3	67 y	M	III	RLL	30 mm	Diffuse abdominal pain	US, MRI
4	64 y	F	III	RLL	40 mm	Right upper quadrant pain	US, MRI
5	73 y	F	III	RLL	34 mm	Cirrhosis	CT
6	80 y	M	I	RLL	–	Anemia	CT
7	18 y	F	I	RLL	–	Calcified liver cyst	CT
8	67 y	M	I	RLL	–	Cirrhosis	US, CT, MRI

y, years; M, male; F, female; LLL, left liver lobe; RLL, right liver lobe; CT, computed tomography; MRI, magnetic resonance imaging; US, ultrasound.

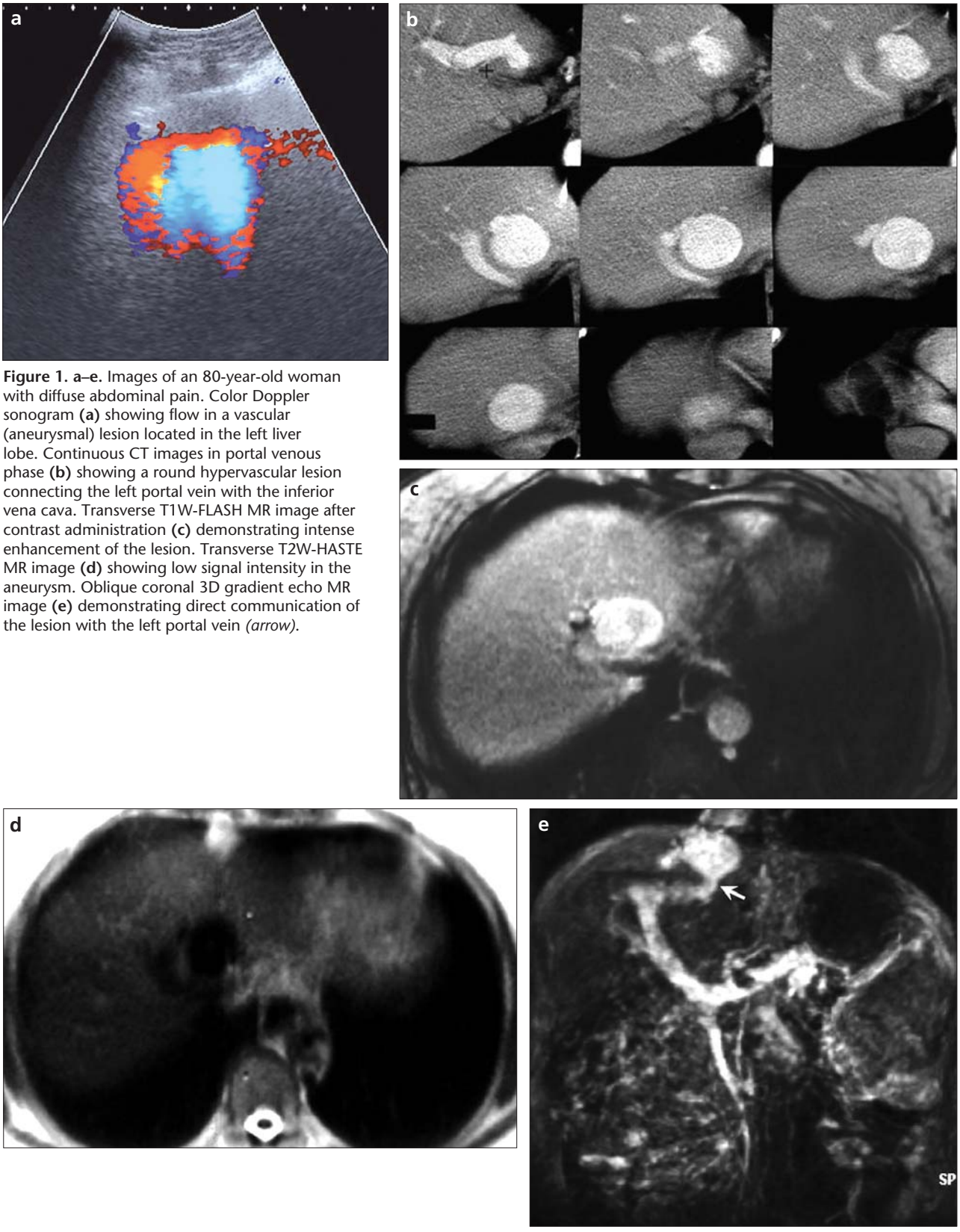


Figure 1. a–e. Images of an 80-year-old woman with diffuse abdominal pain. Color Doppler sonogram **(a)** showing flow in a vascular (aneurysmal) lesion located in the left liver lobe. Continuous CT images in portal venous phase **(b)** showing a round hypervascular lesion connecting the left portal vein with the inferior vena cava. Transverse T1W-FLASH MR image after contrast administration **(c)** demonstrating intense enhancement of the lesion. Transverse T2W-HASTE MR image **(d)** showing low signal intensity in the aneurysm. Oblique coronal 3D gradient echo MR image **(e)** demonstrating direct communication of the lesion with the left portal vein *(arrow)*.

IPSVS, resulting from portal hypertension due to cirrhosis or chronic hepatitis, iatrogenic or traumatic episodes, or rupture of a portal venous aneurysm into a hepatic vein (10–12). The internal type of IPSVS is thought to be congenital in origin because of its low prevalence of coexisting liver cirrhosis. This is supported by the finding of associated anomalies of hepatic vessels, such as portal vein aneurysm, hepatic venous anastomosis, and portal vein

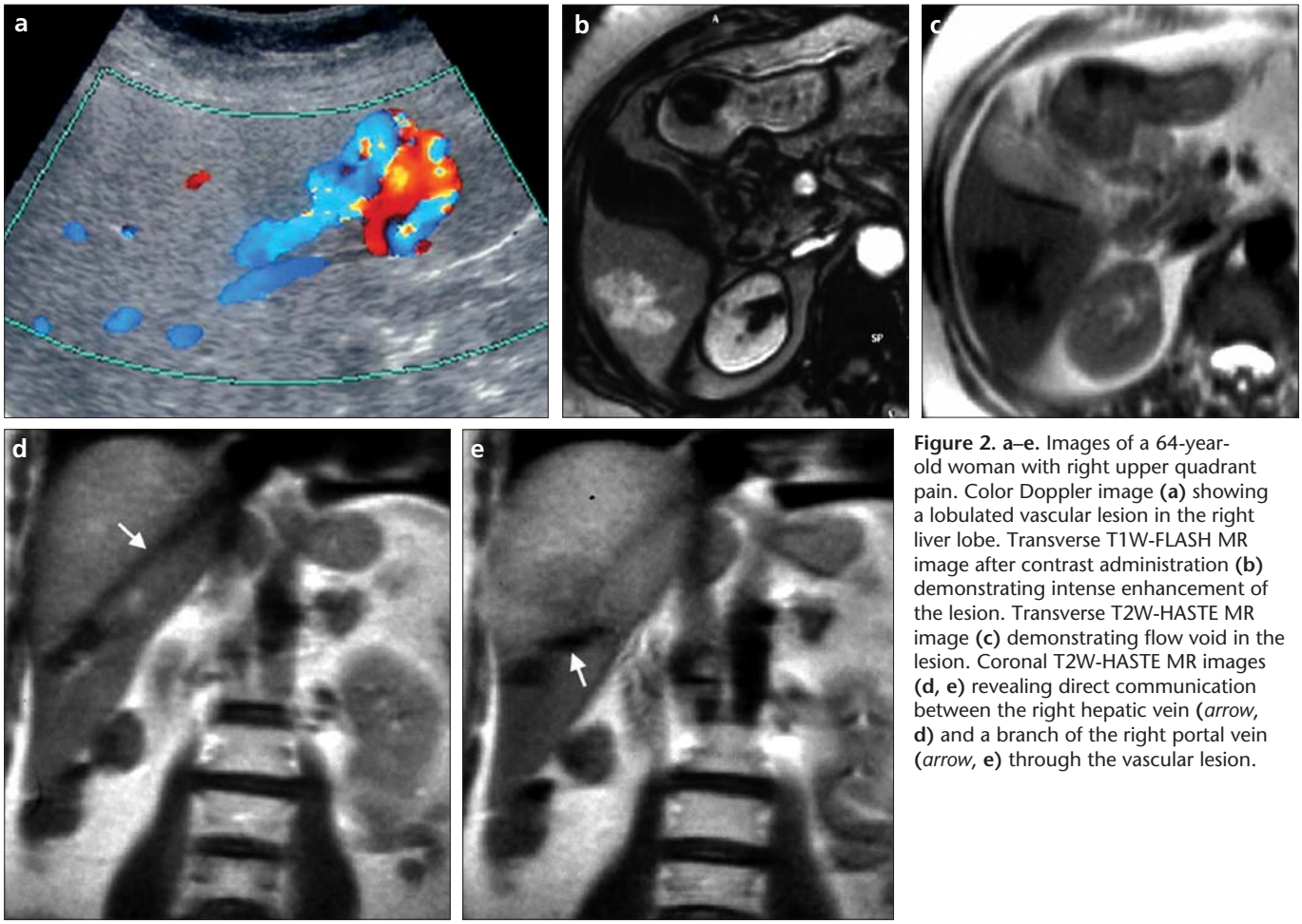


Figure 2. a–e. Images of a 64-year-old woman with right upper quadrant pain. Color Doppler image **(a)** showing a lobulated vascular lesion in the right liver lobe. Transverse T1W-FLASH MR image after contrast administration **(b)** demonstrating intense enhancement of the lesion. Transverse T2W-HASTE MR image **(c)** demonstrating flow void in the lesion. Coronal T2W-HASTE MR images **(d, e)** revealing direct communication between the right hepatic vein **(***arrow*, **d)** and a branch of the right portal vein **(***arrow*, **e)** through the vascular lesion.

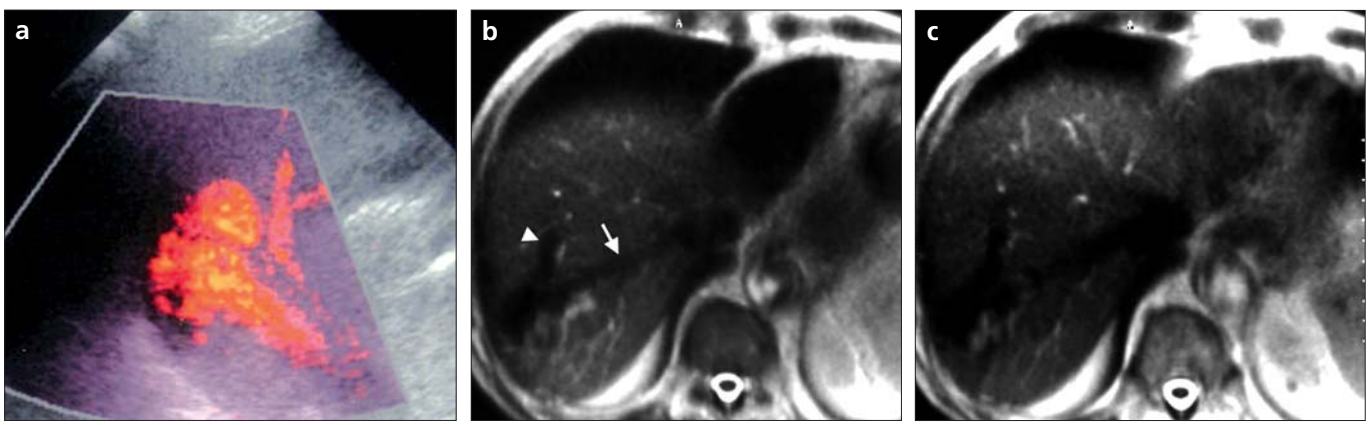


Figure 3. a–c. Images from a 67-year-old man with choledocholithiasis. Color Doppler image **(a)** showing a vascular lesion in the right liver lobe. Transverse T2W-HASTE MR images **(b, c)** demonstrating communication between a branch of the right portal vein *(arrowhead)* with the right hepatic vein *(arrow)* through a vascular lesion (aneurysm).

anastomosis, by Tanoue et al. (3). The acquired theory could explain the external type of IPSVS. In these patients there is high incidence of liver cirrhosis, and perihepatic veins are developed as a result of portal hypertension, as intra- and extrahepatic collateral pathways (3).

The theory of congenital development of IPSVS suggests that anastomosis exists between subcardinal venous system and vitelline venous system (the precursor of portal and hepatic veins) in an early stage of embryologic development. At 5 weeks gestation, hepatic cords surround vitelline venous plexus; this plexus forms hepatic sinusoids. Bilateral umbilical veins also form sinusoids. At 8 weeks, sinusoids start to develop, and the portal and hepatic venous system is formed. At the same time, the right umbilical vein and the cranial portion of left umbilical vein regress. The ductus venosus, a

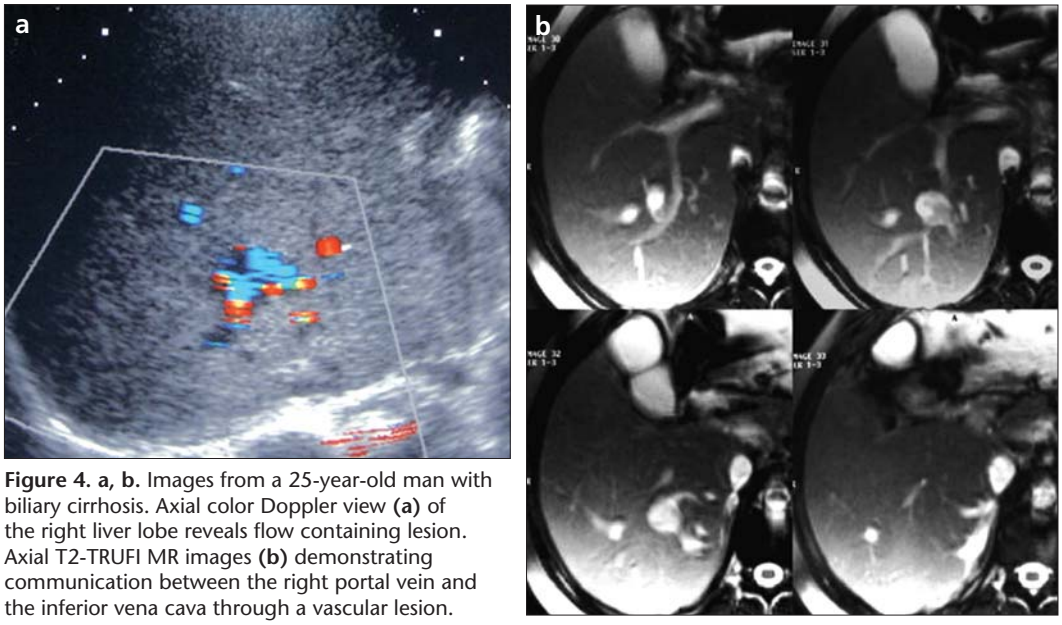
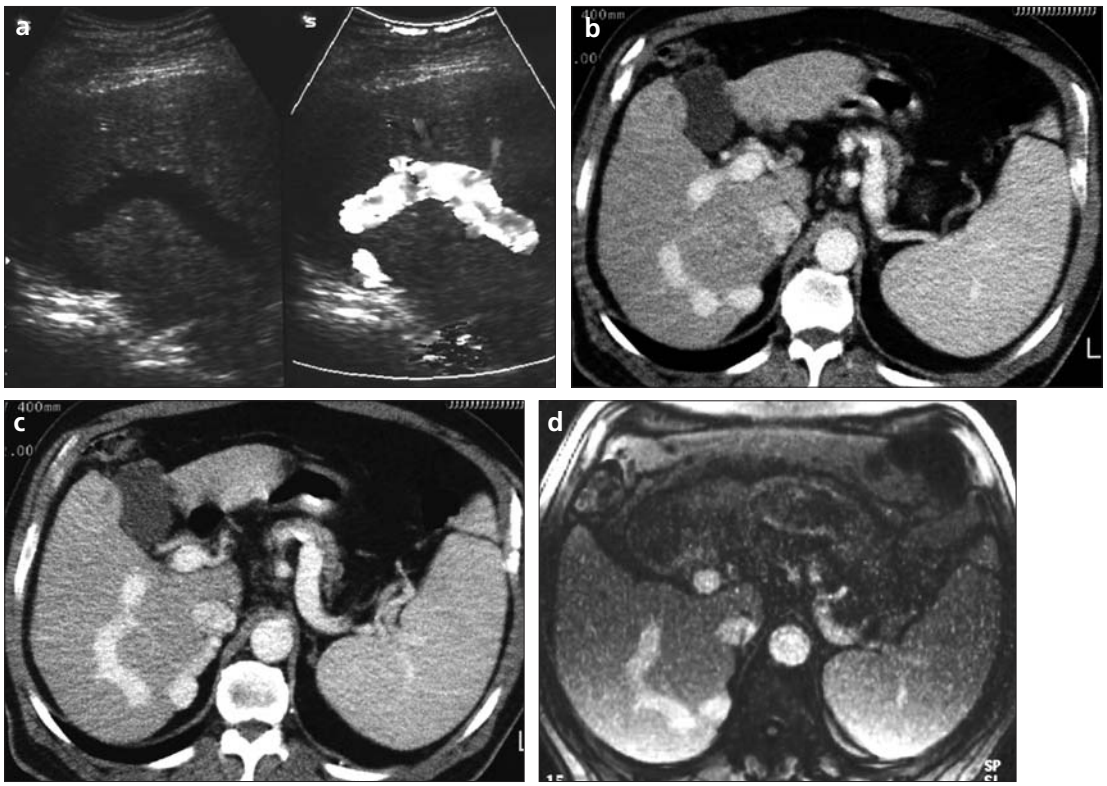


Figure 4. a, b. Images from a 25-year-old man with biliary cirrhosis. Axial color Doppler view **(a)** of the right liver lobe reveals flow containing lesion. Axial T2-TRUFI MR images **(b)** demonstrating communication between the right portal vein and the inferior vena cava through a vascular lesion.



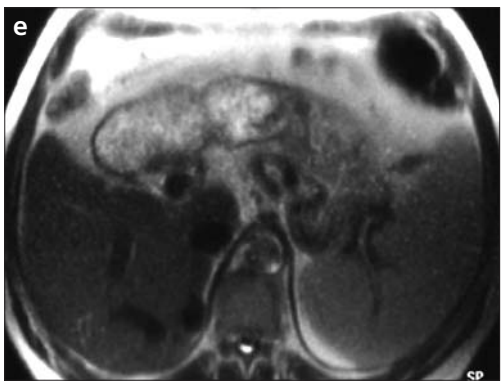


Figure 5. a–e. Images from a 67-year-old man with cirrhosis. Color Doppler sonogram **(a)** shows a large tubular lesion in the right liver lobe (images in gray scale). Axial CT images **(b, c)** in portal phase demonstrating a tubular lesion with constant diameter, connecting the right branch of the portal vein with the inferior vena cava. Transverse T1W-FLASH MR image after contrast administration **(d)** demonstrating intense enhancement of the lesion. Transverse T2W-HASTE MR image **(e)** demonstrating flow void in the lesion.

tubular structure between the left umbilical vein and the inferior vena cava, appears at this stage. At 12 weeks gestation, further differential growth of the portal and hepatic venous system is noted; if complete segregation between these systems does not occur, residual communications between them correspond to intrahepatic portosystemic venous shunt after birth (13).

The clinical significance of IPSVS is the potential for development of hepatic encephalopathy (3, 9, 11, 14, 15). Uchino et al. found that in 51 cases of congenital IPSVS, there were 12 patients with hepatic encephalopathy at the time of diagnosis (9). Some authors report that the prognosis of IPSVS depends on the shunt ratio and patient age (16). The rate of hepatic encephalopathy increases with the age because of decreasing tolerance of the brain to toxic metabolites. Also, large intrahepatic shunts are more often responsible for encephalopathy than small shunts because of the higher degree of shunting. However, according to Remer et al., this theory does not fit the pathophysiology of hepatic encephalopathy as it is understood today (6).

US, CT, and MRI findings are sufficient for imaging evaluation of IPSVS. Imaging findings differ in proportion to the type of the shunt. In case of aneurysmal communication between portal and hepatic veins, color Doppler imaging demonstrates a vascular lesion supplied by a vascular branch with monophasic flow (portal flow shows subtle phasic variation caused by respiration-related changes in thoracic pressure) and drained by a vessel with biphasic flow (hepatic vein flow has a pulsatile pattern that results from transmission of right atrial pulsations into the veins). On post-contrast enhanced CT, a rounded mass with homogenous strong enhancement is present, accompanied by a portal vein branch entering the lesion and a hepatic vein branch exiting it. MRI provides findings similar to CT, but MRI offers the advantages of sagittal and coronal imaging and the potential of MR venography. In tubular communication between right portal vein and the inferior vena cava, US adequately demonstrates this tubular vessel, whereas CT and MRI provide more global visualization.

Treatment should be considered only for symptomatic patients. Dietary management with limitation of protein intake and supplementation with lactulose are the first approach. If dietary modifications fail to control the symptoms of encephalopathy, interventional methods must be implemented. Symptomatic intrahepatic portosystemic venous shunts are adequately treated by transcatheter embolization. Access routes that can be followed are transileocolic obliteration, percutaneous transhepatic obliteration, and retrograde transcaval obliteration. Symptoms related to portal-systemic encephalopathy improve or completely disappear after embolization (3).

In our cases, the findings of IPSVS were well depicted with US, CT, and MRI. Most shunts (7 of 8) were located in the right lobe of the liver; the shunt was found in the left lobe in only 1 case. Aneurysmal communication between the portal and systemic circulations (type III) was the most common type of IPSVS (62.5%) in our series. Type III was also the most frequent type of IPSVS in the series of Tanue et al. (70%) and of Remer et al. (54%) (3, 6). Remer et al. found that 76% of shunts were located in the left lobe of the liver, whereas only 1 (12.5%) case had this location in our study (6). We found type I shunts in three of eight patients (37.5%), whereas Remer et al. found only 1 case of type I shunt in 22 patients (4.5%) (6).

In conclusion, IPSVS is a rare vascular abnormality that can be adequately diagnosed with US, CT, and MRI with the exception of very small lesions. Identification of asymptomatic shunts has increased because of advances in imaging techniques. In our series, IPSVS was located in the right lobe of the liver in almost all cases, with a vascular lesion or an aneurysm bridging a branch of the portal vein with a branch of the hepatic vein. Radiologists should be aware of this vascular anomaly because it can be recognized in asymptomatic patients in whom treatment is not required.

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