

# Congenital intrahepatic portosystemic shunt with variant inferior right hepatic vein

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## ABSTRACT

We present a case of congenital intrahepatic aneurysmal portosystemic shunt in which the right portal vein communicates with both a variant inferior right hepatic vein and the right hepatic vein. We also describe the importance and efficacy of three dimensional computed tomography reconstruction images in determining subtle, small shunt vessels.

**Key words:** • tomography, spiral computed  
• ultrasonography, Doppler, color • liver • portal vein  
• hepatic veins

**P**ortosystemic shunts are classified into four morphological types (1). The most common, type 1, is a single large vessel of constant diameter that connects the right portal vein to the inferior vena cava. Type 2 is a localized peripheral shunt in which single or multiple communications are found between peripheral branches of portal and hepatic veins in one hepatic segment. Type 3 is an aneurysmal communication between the peripheral portal and hepatic veins. Type 4 has multiple communications between portal and hepatic veins distributed in both lobes (Fig. 1). In this report, we present a case with an aneurysmal connection between the right portal vein and the inferior right hepatic vein, which represents a type 3 portosystemic shunt. In addition, the right hepatic vein, which was noted to have a very small caliber and a long, curved course, was connected to the aneurysm. Contrast-enhanced computed tomography (CT) scans and especially, three-dimensional reconstructions clearly demonstrated the right portal vein and its connection to the aneurysm that could not be identified on color Doppler ultrasonography (US) examination.

## Case report

A 56-year-old man was referred for CT examination with a presumptive diagnosis of left renal colic. On unenhanced CT images (Presto Multi-slice CT Scanner, Hitachi Medical Systems, Singapore) the urinary system was normal; however, a hypodense, well circumscribed lesion was detected in the right lobe of the liver. On contrast-enhanced CT, an aneurysm connecting the inferior right hepatic vein to the right portal vein was detected (Fig. 2). In addition, the variant inferior right hepatic vein was seen to have a very small caliber (2.5 mm) and a long, curved course, and to communicate with the same aneurysm. The long course of the right hepatic vein and its connection to the aneurysm were clearly demonstrated by using three-dimensional image processing techniques (Fig. 3). Color Doppler US (Sonoline G50; Siemens, Erlangen, Germany) demonstrated the flow entering the aneurysm from right portal vein and draining into inferior right hepatic vein (Fig. 4); however, the connection between the aneurysm and right hepatic vein was not demonstrated definitively. The liver and the spleen were of normal size with smooth borders and normal internal architecture on both US and CT examinations. Portal venous system was normal. Laboratory data including liver function tests and blood ammonia levels were within normal limits. Viral serology test results were negative. Physical examination was unremarkable. There was no previous history of liver disease, abdominal trauma, surgery, or liver biopsy. Because the results of laboratory tests and radiological examinations were not suggestive of chronic liver disease, liver biopsy was not performed. Intrahepatic portosystemic shunts in this case were thought to be congenital.

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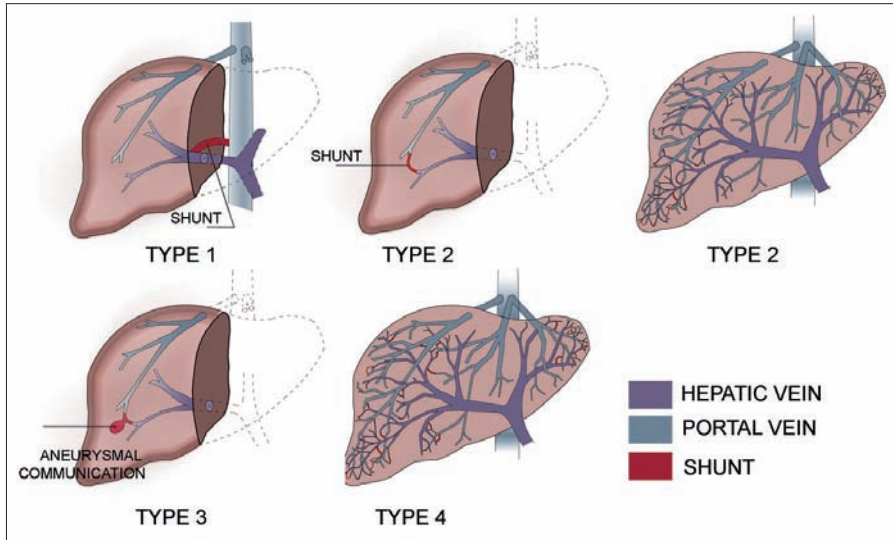


Figure 1. Portosystemic venous shunts according to the classification of Park et al. (1).

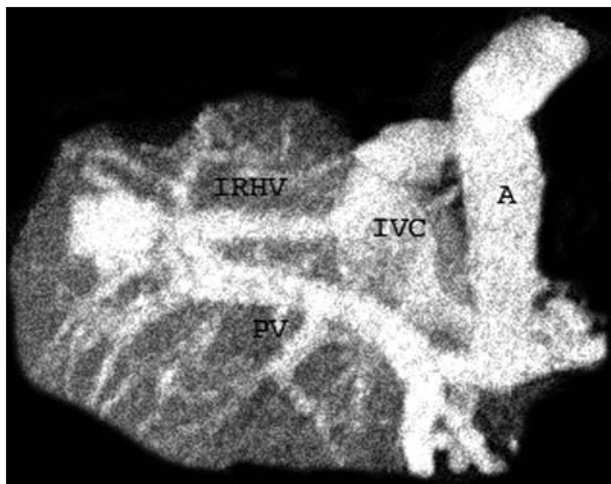


Figure 2. Volume-rendered reconstruction CT image shows aneurysmal communication between right portal vein and inferior right hepatic vein. A, aorta; IVC, inferior vena cava; PV, portal vein; IRHV, inferior right hepatic vein.

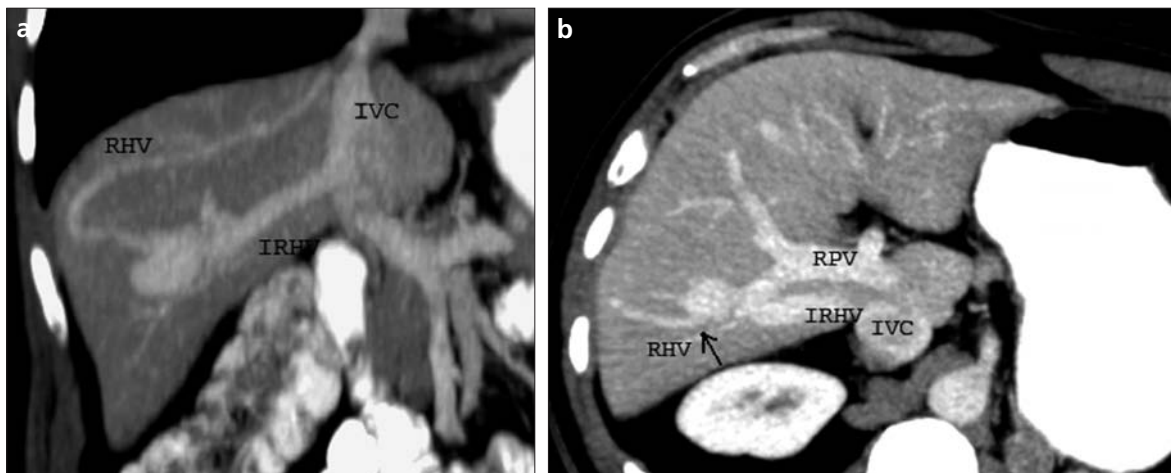


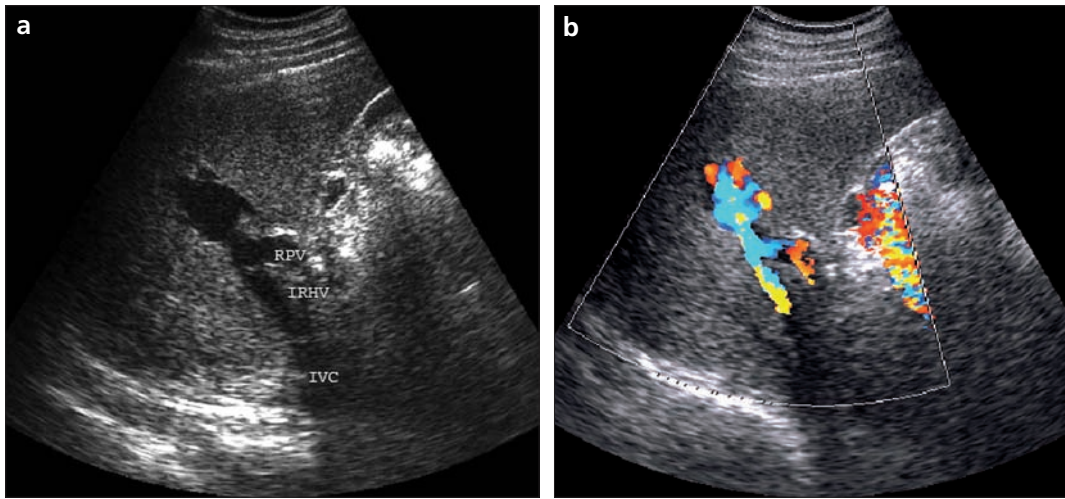
Figure 3. a, b. Coronal oblique maximum-intensity-projection (MIP) CT image (a) depicts the course of the right hepatic vein and its connection with the aneurysm. Communication between the aneurysm and the inferior right hepatic vein can also be seen. Axial oblique MIP CT image (b) demonstrates the connection between right hepatic vein and aneurysm (arrow). Aneurysmal communication between right portal vein and inferior right hepatic vein is apparent. IVC, inferior vena cava; RHV, right hepatic vein; RPV, right portal vein; IRHV, inferior right hepatic vein.

## Discussion

The right hepatic vein is usually a single vessel that drains the right lobe of the liver; however, there may be a variant inferior right hepatic vein in about 18% of the population. As a general rule, the inferior right hepatic vein drains segment VI and adjacent hepatic parenchyma, and flows directly into the right posterior aspect of the inferior vena cava, 3 to 5 cm distal to the right hepatic vein (2). The inferior right hepatic vein may be larger than the right hepatic vein. In the present case, we found incidentally an aneurysmal portosystemic communication between the right portal vein and the inferior right hepatic vein, which may represent a type 3 portosystemic shunt according to the classification of Park et al. (1). The right hepatic vein was also connected to this formation.

Intrahepatic portosystemic venous shunt is a congenital or acquired condition consisting of a communication between the portal and systemic venous circulation, situated at least partially within the liver (3). Acquired shunts may develop as intrahepatic collateral pathways secondary to portal hypertension and cirrhosis (4), or may result from trauma, liver biopsy, or surgery.

Congenital intrahepatic portosystemic venous shunt is an uncommon condition that is best understood in the context of the embryological development of the portal and hepatic venous systems. By the fourth week of fetal



**Figure 4.** a, b. US scan (a) demonstrates an anechoic lesion on the right hepatic lobe that communicates with inferior right hepatic vein and portal vein. IVC, inferior vena cava; RPV, right portal vein; IRHV, inferior right hepatic vein. Color Doppler US scan (b) shows communicating color signals of the inferior right hepatic vein and right portal vein with the aneurysm.

life, the vitelline and umbilical systems begin to break into intrahepatic sinusoids that give rise to the intrahepatic portal and hepatic veins, respectively. Congenital portal-systemic shunts are thought to represent persistent venous communications (3, 5). Early diagnosis and periodic evaluation of the shunt are important because of the ongoing risk of hepatic encephalopathy with exposure to high ammonia levels, especially if the shunt may be progressive (6, 7). Additionally, reduction in blood flow to the liver may result in fatty degeneration, hepatic dysfunction, and atrophy of the liver (8).

Color Doppler US is recommended as the primary imaging modality for diagnosis of portosystemic venous shunts. In addition to demonstrating flow signals between the involved vessels, and evaluating flow direction, it may also determine the shunt ratio by estimating flow volume (6, 7). Contrast-enhanced CT with three-dimensional image processing techniques is usually performed to supplement US, especially in obese patients or in pa-

tients with marked liver atrophy (6). In the present case, the communication between the aneurysm and the right hepatic vein that eludes observation on color Doppler US is demonstrated clearly on contrast-enhanced CT, on coronal and axial oblique maximum intensity projection (MIP) images. We were not able to visualize this communication on color Doppler US, most likely because of the small caliber (2.5 mm) and long course of the inferior right hepatic vein. Although several suspicious small vessels were seen near the aneurysm, standard anatomic orientation with planar images was not adequate to follow the long course of such a small and curved vessel.

Multiplanar CT images created by three-dimensional image processing techniques are more effective than two-dimensional images in demonstrating variant small and curved vessels. With extensive use of these techniques shunt vessels such as these that may elude observation by other imaging modalities may be recognized more commonly.

## References

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