

# Asymptomatic Congenital Intrahepatic Portosystemic Shunt

Karen M. Brown, M.D., and Hassan Hal, M.D.

Citation: Brown KM, Hal H. Asymptomatic Congenital Intrahepatic Portosystemic Shunt . Radiology Case Reports. [Online] 2009;4:255.

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Abbreviations: CT, computed tomography

Karen M. Brown, M.D. (Email: [kbrown1@psu.edu](mailto:kbrown1@psu.edu)), and Hassan Hal, M.D., are in the Department of Radiology, Milton S. Hershey Medical Center, Hershey, PA, United States of America.

Published: January 31, 2009

DOI: 10.2484/rcr.v4i1.255

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## Abstract

Congenital (spontaneous) intrahepatic portosystemic shunt is rare in the English literature. Most cases of portosystemic shunt occur after trauma, surgery, liver biopsy or as a result of chronic portal hypertension. Chronic shunting may result in encephalopathy, bleeding or hyperinsulinism. We report a case of an asymptomatic adult female with a presumed congenital intrahepatic portosystemic shunt and discuss the pertinent imaging findings and important key concepts related to this condition.

## Introduction

Macroscopic congenital portosystemic shunts within the hepatic parenchyma are not uncommon as an incidental finding but have not been well characterized or described. Most adult patients with portosystemic shunts present with bleeding or hepatic encephalopathy as a first manifestation [1]. It has been hypothesized that patients become more symptomatic with advancing age as the tolerance for high ammonia levels diminishes [2]. We present a case of an asymptomatic congenital intrahepatic portosystemic shunt discovered as an incidental finding in a 48-year old female with pyelonephritis.

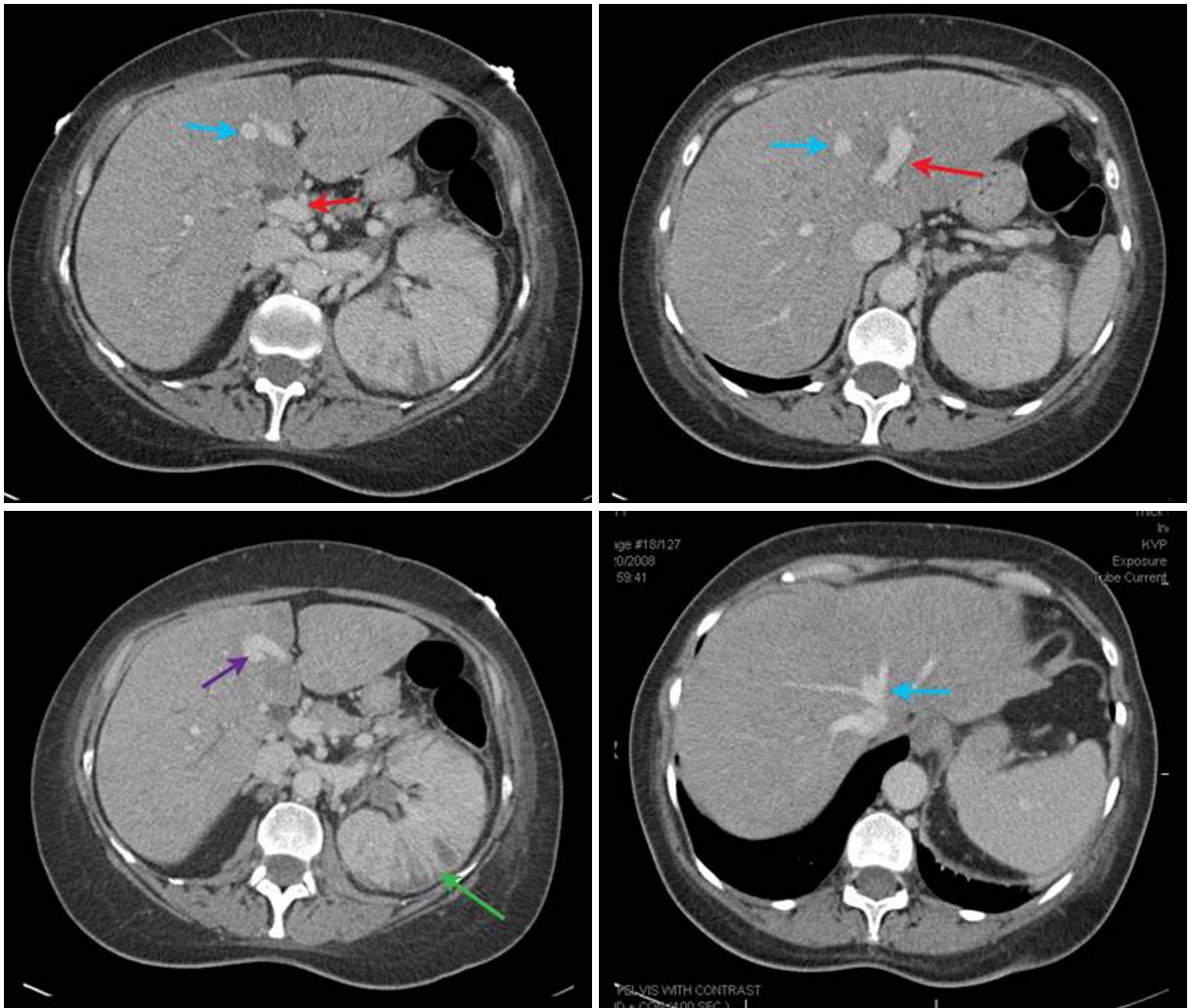
## Case Report

A 48-year-old woman was evaluated for left flank pain and possible perinephric abscess. Her past medical history was significant for recurrent renal calculi and pyelonephritis. She also had a remote history of stroke at age 22, attributed to oral contraceptive use. She had no history of liver disease, liver biopsy, abdominal trauma, cirrhosis, mental status changes or diabetes. Medications included iron supplement, acetaminophen, cephalixin, and pyridium. She denied alcohol use. She did have a positive smoking history. Physical exam was negative for hepatomegaly or jaundice, positive for left upper quadrant tenderness with no guarding, rebound, or rigidity. Heart rate was slightly irregular with occasional premature beats.

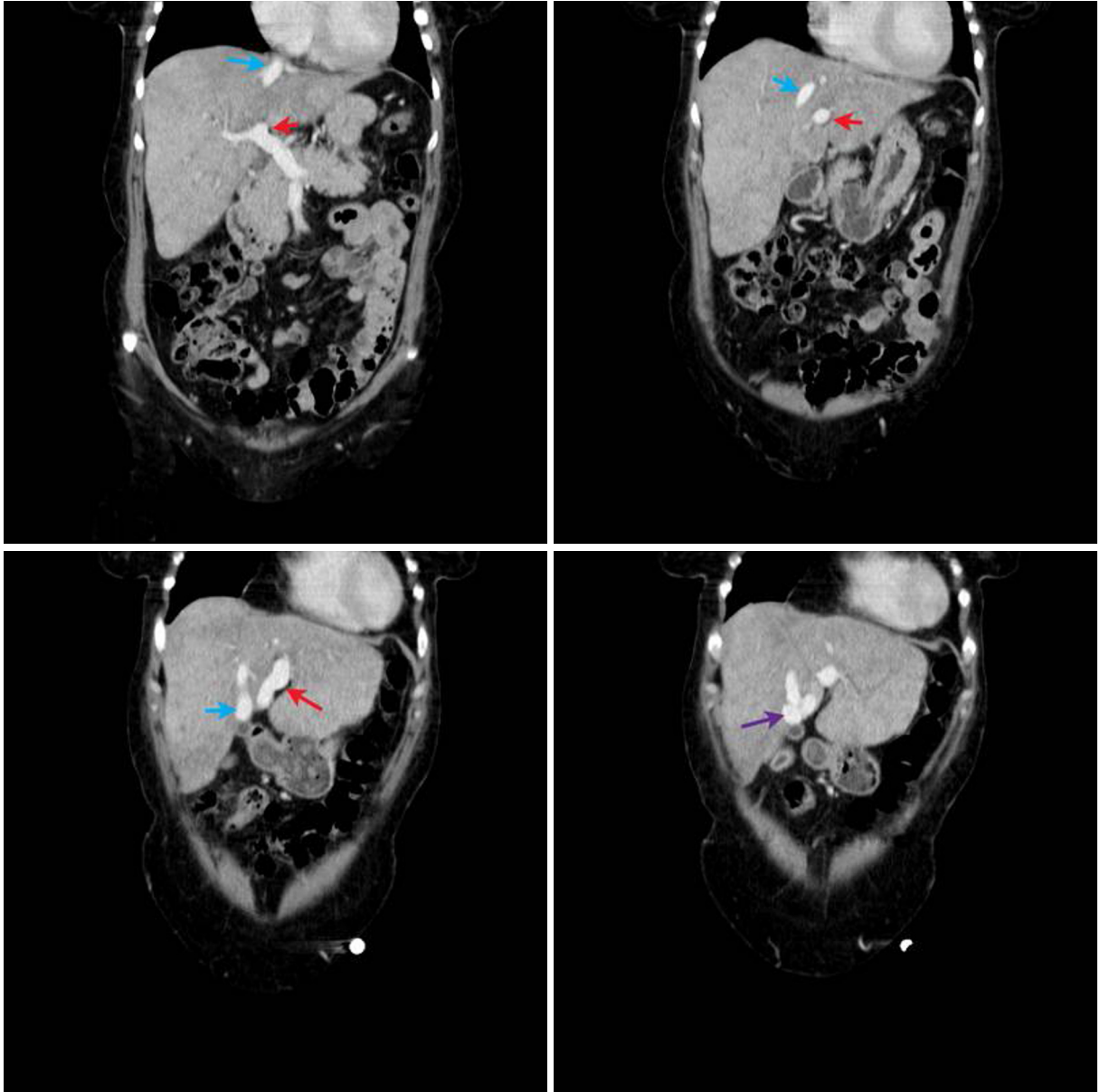
Pertinent laboratory values were: platelets 566K/microL (elevated), ALT 13 units/L, total bilirubin 0.3 mg/dL, and alkaline phosphatase 117 units/L, all normal. Sodium, potassium, chloride, bicarbonate, blood urea nitrogen, creatinine and glucose levels were all within normal limits.

A CT scan, performed to evaluate the kidneys, demonstrated CT findings in keeping with left pyelonephritis. Incidentally noted was an enlarged, tortuous vessel within the left hepatic lobe connecting the left portal vein with the middle hepatic vein at the fissure for the ligamentum venosum. The vessel measured 1.4 cm at its widest diameter (Figs. 1-2). The liver parenchyma was homogeneous and normal with no focal lesion identified. The main portal vein was patent. Ultrasound evaluation of the right upper quadrant performed three years prior demonstrated a direct communication of color flow signals between the middle hepatic and portal veins within the left hepatic lobe (Fig. 3) Additionally, an enhanced abdominal CT three years prior demonstrated the same intrahepatic portosystemic shunt, unchanged in appearance or size. No additional hepatic abnormalities were identified.

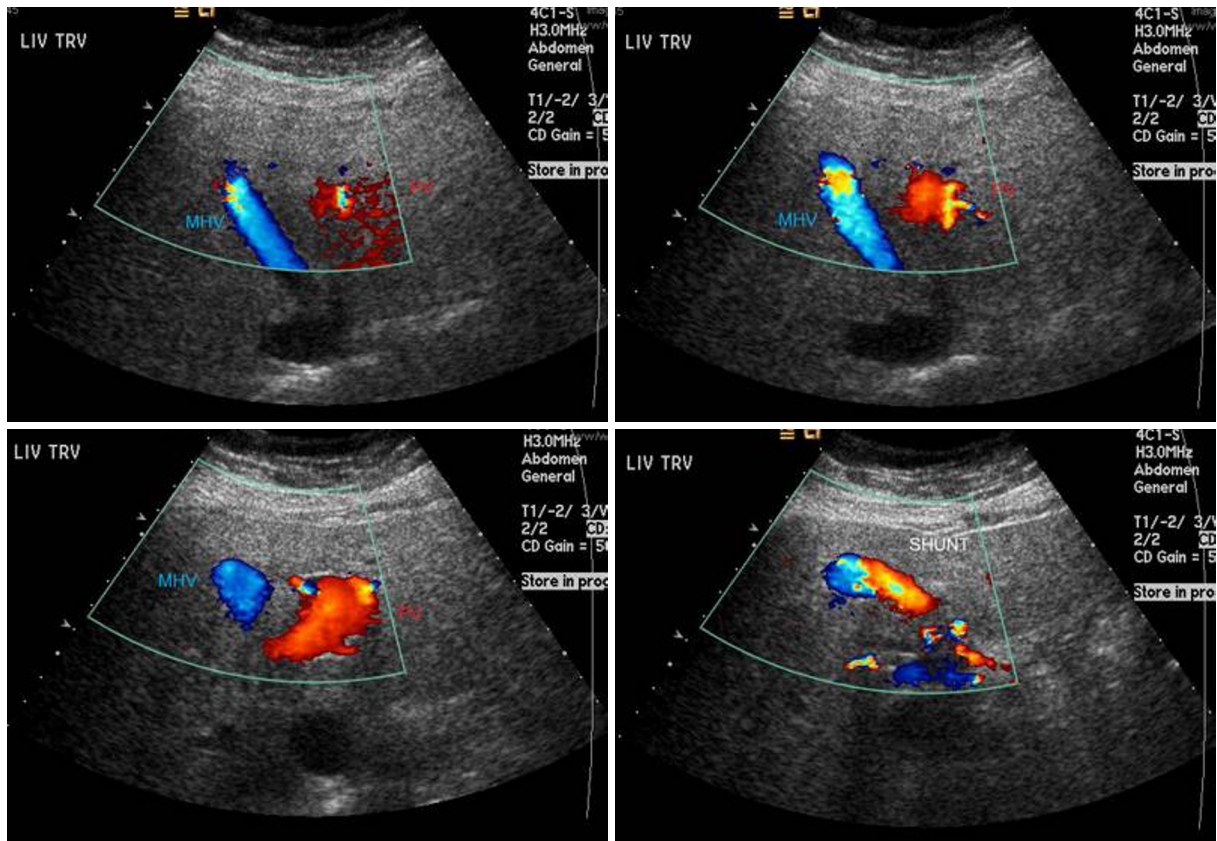
No intervention was performed as she was asymptomatic from this finding. The etiology of this shunt is assumed to be congenital, as she had no history of trauma, cirrhosis, portal hypertension or surgical intervention.



**Figure 1.** 48-year-old woman with congenital intrahepatic portosystemic shunt. (A-D) Axial enhanced CT images demonstrate the portal vein (red arrow) and middle hepatic vein (blue arrow) with a portosystemic shunt within the medial segment of the left hepatic lobe (purple arrow). Note also a striated left nephrogram (curved green arrow).



**Figure 2.** 48-year-old woman with congenital intrahepatic portosystemic shunt. (A-D) Coronal enhanced CT images demonstrate branches of the portal vein (red arrow) and middle hepatic vein (blue arrow) with a portosystemic shunt within the medial segment of the left hepatic lobe (purple arrow).



**Figure 3.** 48-year-old woman with congenital intrahepatic portosystemic shunt. (A-D) Transverse sonographic images through the liver demonstrate hepatopedal flow through the portal vein (PV) and hepatofugal flow through the middle hepatic vein (MHV) with turbulence within a portosystemic shunt within the left hepatic lobe.

## Discussion

Intrahepatic portosystemic shunts are rare in the English literature and are usually associated with portal hypertension, trauma, surgical intervention or liver biopsy. Embryologically, the intra- and extrahepatic portal venous system develops by selective persistence of the vitelline and umbilical systems between the fourth week and third month of fetal life [1, 3]. In the congenital origin theory, intrahepatic portosystemic venous shunts are thought to represent persistent communication between the cranial and caudal hepatic sinusoids formed by vitelline veins and the umbilical vein [4]. Park et al have characterized four different morphologic types [5]. The most common type is a single large tube of constant diameter connecting the right portal vein to the inferior vena cava. This includes a patent periumbilical vein, considered to be a collateral pathway developing as a result of portal hypertension. The second type is a localized peripheral shunt with single or multiple communications found between branches of the portal vein and hepatic veins in one hepatic segment. The third type is an aneurysmal connection between peripheral portal and hepatic vein segments. The fourth type consists of multiple communications between peripheral portal and hepatic veins within both hepatic lobes. Our patient presented with a variant of a type 1 shunt, consisting of a single large vessel of constant diameter connecting the left portal vein to the middle hepatic vein. Of note, the connection is not through a patent ductus venosus, as the anastomosis occurs within otherwise normal hepatic parenchyma within the medial left hepatic lobe. Chevallier et al [6] developed a different classification with type 1 including patent periumbilical veins within the liver parenchyma, as seen in portal hypertension. Types 2 and 3 include shunts, single or multiple, between a portal branch and the

hepatic vein located either in two adjacent liver segments (type 2), or in non-adjacent liver segments (type 3). Type 4 includes any vascular connection between the right portal branch and the inferior vena cava. According to this classification, our patient presented with a variant of type 4, with a vessel connection between the left portal vein and the middle hepatic vein.

The diagnosis of intrahepatic portosystemic venous shunt can be made by ultrasound with color Doppler, CT, MRI or conventional angiography. Color Doppler imaging reveals a direct communication of color flow signals between the portal and hepatic veins, with changes within the spectral waveform from a continuous waveform signal, as seen in the portal vein, to a turbulent signal within the shunt region to a biphasic waveform as seen within the hepatic vein[7]. MRI demonstrates flow void within the region of the shunt. Additionally, MRI may be useful in visualizing multiple diffuse shunts [8]. Transarterial/transhepatic portography is diagnostic for shunt visualization demonstrating either the shunt itself or large pooling from the dilated portal branch with subsequent visualization of the hepatic vein [3]. In our case, contrast-enhanced CT demonstrated an abnormal vein between the left portal vein and middle hepatic vein. However, CT cannot demonstrate reversal of flow in either vessel, as can be seen with color Doppler imaging. We believe this vascular malformation to be a congenital abnormality as no signs of cirrhosis or trauma were found. Additionally, the patient had no history of surgical intervention. The etiology of spontaneous intrahepatic portosystemic shunt is unknown. Some authors postulate a persistent venous anastomosis such as patent ductus venosus and right vitelline vein. Alternatively, the shunt may be acquired from rupture of a portal venous aneurysm into the hepatic vein or from a dilated hepatic vein communicating with the inferior vena cava via inferior phrenic and suprarenal veins. [9,10]. Our patient likely demonstrates a variant of a patent ductus venosus with failure of regression.

Of note, intrahepatic communications have rarely resulted in encephalopathy and hyperinsulinism with secondary hypoglycemia [11, 12]. Most shunts can be completely cured by transcatheter embolization. Potential complications of coil embolization include portal hypertension caused by altered portal hemodynamics and dislodgment of coils into the systemic circulation [4,13]. Surgical intervention such as shunt ligation, hepatic resection, and creation of an alternative portosystemic shunt are additional therapeutic approaches.

Previous studies have demonstrated an association between intrahepatic portacaval shunts and neonatal jaundice, congenital heart disease, congenital biliary atresia, polycystic ovary syndrome, pelvic myoma, coronary artery fistulas, hemangiomas, and membranoproliferative glomerulonephritis, focal nodular hyperplasia, hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu syndrome), and pulmonary hypertension due to vasoconstrictive agents bypassing metabolism in the liver [10,14-19]. Only one other case in the literature describes a truly asymptomatic portacaval shunt with morphology such as seen in our patient [20]. Like the previous presentation, our patient presented with an unrelated complaint and previous studies demonstrate no change over three years. As she is asymptomatic from this incidental finding, no intervention was performed or recommended. It is hypothesized that hepatic encephalopathic symptoms may develop with increasing age [2,5]; we hope that further followup may elucidate the natural evolution of this portosystemic shunt.

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