

# Spontaneous Portoazygos Shunt in a Patient with Portal Hypertension

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## ABSTRACT

We present a case of spontaneous portoazygos shunt in a patient with liver cirrhosis and portal hypertension. The shunt was incidentally detected by abdominal magnetic resonance imaging for routine evaluation of liver cirrhosis. Multiplanar reconstruction images demonstrated the portal vein communicating with the azygos vein that was dilated and tortuous along its course to the mediastinum. Although there has been a case of congenital portoazygos shunt reported in a neonate with multiple congenital anomalies, to the best of our knowledge, this is the first case of spontaneous portoazygos shunt developed in an adult with portal hypertension.

## CASE REPORT

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A 46-year-old male with known liver cirrhosis secondary to alcohol and hepatitis C underwent an MRI of the abdomen in 2010 for routine evaluation of the liver for hepatocellular carcinoma. The patient's clinical history dates back to 2006 when he was incidentally found to have liver cirrhosis during a laparoscopic cholecystectomy. Between 2007 and 2009, he developed multiple episodes of hematemesis secondary to esophageal varices necessitating multiple endoscopic banding procedures. The patient also had multiple episodes of encephalopathy since 2007 for which he was maintained on lactulose. The patient's current MELD score was 10 and Child-Pugh class was B. He denied any symptoms in the chest and abdomen. His vital signs were stable.

Abdominal MRI was performed with a 1.5-T MR imaging system (Avanto, Siemens, Malvern, NJ, USA) and a phased-array torso coil. The pulse sequences included two-dimensional T1-weighted fast spin-echo, T2-weighted fast spin-echo fat-suppressed, T2-weighted gradient-echo fat-suppressed, and three-dimensional volumetric interpolated breath-hold examination (VIBE) fat-suppressed sequences after intravenous administration of paramagnetic contrast material (Gadobenate dimeglumine, Multihance, Bracco Diagnostics, Princeton, NJ, USA, 0.1mmol/kg). MRI revealed liver cirrhosis with no evidence of intrahepatic mass. T1-weighted VIBE images demonstrated that the posterolateral

aspect of the main portal vein was connected to the dilated and tortuous azygos vein via enlarged venous channel (Figures 1-3). The azygos vein was markedly dilated and tortuous with a maximal diameter of 4 cm. Multiple extensive paraesophageal and retroperitoneal varices were also noted along with small caliber parasplenic and gastric varices.

The patient did not undergo any interventional procedure or surgery. A follow-up abdominal MRI (Figure 4) performed 12 months later demonstrated no significant interval changes in portoazygos shunt and liver cirrhosis. He was managed medically and followed by the hepatology service without evidence of recurrent variceal bleeding or hepatic encephalopathy.

### DISCUSSION

The portal venous system drains blood flow of the gastrointestinal tract from the lower esophagus to the upper anal canal. The main tributaries of the portal venous system include splenic, superior and inferior mesenteric, left and right gastric, paraumbilical and cystic veins [1-3]. In patients with portal hypertension, numerous portosystemic collateral veins can develop, including esophageal, paraesophageal, coronary gastric, inferior phrenic, paraumbilical, abdominal wall, splenorenal, gastrosplenic, retrocaval, and mesocaval collateral

pathways [4]. To date, there has been only one case reported of congenital portoazygos shunt in a neonate with thoracoabdominal duplication cysts and multiple other congenital anomalies [5]. We report a case of spontaneous portoazygos shunt that developed in an adult patient with portal hypertension.

Portosystemic collaterals commonly occur as a consequence of portal hypertension. Prehepatic, intrahepatic, and posthepatic increased resistance to portal venous flow lead to portal hypertension. In portal hypertension, the high-pressure hepatopetal flow is redirected through the alternative pathways into the low-pressure systemic veins. Some of the clinical manifestations of liver cirrhosis, including gastrointestinal bleeding, ascites and hepatic encephalopathy, are the result of portal hypertension resulting in the development of portosystemic collaterals. To date, more than 20 different types of portosystemic collaterals have been described in the literature [3, 6, 7].

The portal vein is formed by the confluence of the superior mesenteric vein (SMV) and splenic vein. The inferior mesenteric vein (IMV) joins either the SMV or splenic vein. Right and left, frequently multiple, coronary (gastric) veins drain into the splenic vein or directly into the portal vein. The azygos and hemiazygos veins are continuations of the ascending lumbar veins on the right and left, respectively. Before draining into the superior vena cava (SVC), the azygos vein receives multiple paravertebral and intercostal tributaries as well as esophageal, mediastinal and pericardial branches [8].

The radiologic appearances of the portal and azygos systems and portosystemic collaterals are well described using CT and MRI, and these cross-sectional imaging techniques have essentially replaced arterial and percutaneous transhepatic portography [6]. Thin-slice helical CT angiography with contrast enhancement is considered the best imaging modality for demonstrating portosystemic collateral vessels as enhancing lobulated or serpentine structures [3]. However, MRI is performed increasingly these days in surveillance for hepatocellular carcinoma in patients with chronic liver disease. In many cases, MR angiography (MRA) with contrast enhancement is comparable, and can provide excellent information about the portosystemic collateral pathways [9]. However, some of the rare described pathways can be missed on MRI and MRA given their lower spatial resolution [3]. Knowledge about the different collateral pathways and their appearance on MRI is essential in early diagnosis and intervention.

In a retrospective review of 92 CT scans with evidence of liver cirrhosis and varices, Cho et al [6] described several different types of varices. The described portosystemic collateral pathways in decreasing order of frequency are: coronary (gastric), esophageal, paraumbilical, abdominal wall, perisplenic, retrogastric, paraesophageal, omental, retroperitoneal-paravertebral, mesenteric, splenorenal and gastrosplenic veins. Other rare shunts have also been described and they included retrocaval, mesocaval, inferior phrenic [4], pleuropericardial-peritoneal, splenoazygos, and splenic- or

gastric-to-inferior pulmonary veins [1]. Esophageal varices are located inside the wall of the esophagus and thus endoscopy is the modality of choice for the diagnosis. Paraesophageal varices are commonly associated with gastric varices and are located outside the wall of the esophagus. Contrast-enhanced CT or MRI shows lobulated enhancing structures in the posterior and inferior aspect of the mediastinum. Paraesophageal varices may mimic mediastinal mass on nonenhanced CT. Gastric varices may simulate gastric neoplasm on nonenhanced CT. They are typically located at the superior and posterior aspect of the gastric fundus and most commonly drain into the esophageal or paraesophageal varices, although occasionally drain through the gastrosplenic shunt into the left renal vein [3]. Splenorenal shunt is the collateral vessel between the splenic hilum and the left renal vein [4]. Enlarged left renal vein is frequently observed on MRI or CT [3]. Paraumbilical collateral veins arise from the left portal vein and drain through the inferior epigastric vein into the external iliac vein. Occasionally paraumbilical veins communicate with subcutaneous varicose veins of the abdominal wall around the umbilicus creating the caput medusae [4, 6]. Mesenteric collateral veins may develop either between the superior hemorrhoidal vein and the middle/inferior hemorrhoidal vein or between the IMV and the IVC. If mesenteric collaterals are located in the rectum, they may simulate a rectal mass protruding into the lumen on nonenhanced CT and a biopsy may result in devastating consequences. Although they are less frequent than other collateral veins, rectal bleeding can be associated with this type of portosystemic shunt [3, 4, 6].

In the present case, MRI of the abdomen with intravenous administration of paramagnetic contrast material performed for the routine evaluation of the liver demonstrated a rare collateral pathway between the main portal and the azygos veins. The azygos vein was markedly dilated and tortuous along its course to the mediastinum. Additional paraesophageal, gastric, and perisplenic varices were present, but small compared to the portoazygos shunt. Spontaneous portoazygos shunt may be asymptomatic but can cause hepatic encephalopathy or variceal bleeding. Treatment options may include transvenous coil embolization by interventional radiology and surgical ligation. Although the clinical implication and prognosis of the portoazygos shunt are not known, understanding the anatomy and the course of the shunt may help avoid potential complications related to interventional procedure or surgery.

In conclusion, although there has been a case of congenital portoazygos shunt reported in a neonate with thoracoabdominal duplication and absent intrahepatic portal vein [5], to our knowledge, the present case is the first published report to demonstrate a portoazygos shunt in an adult patient with liver cirrhosis and portal hypertension. Coronal imaging that included the thorax was essential to evaluate the full extent of the varices. Contrast-enhanced cross-sectional imaging with CT or MRI provides a detailed evaluation of collateral pathways between the portal and systemic veins that may influence clinical management.

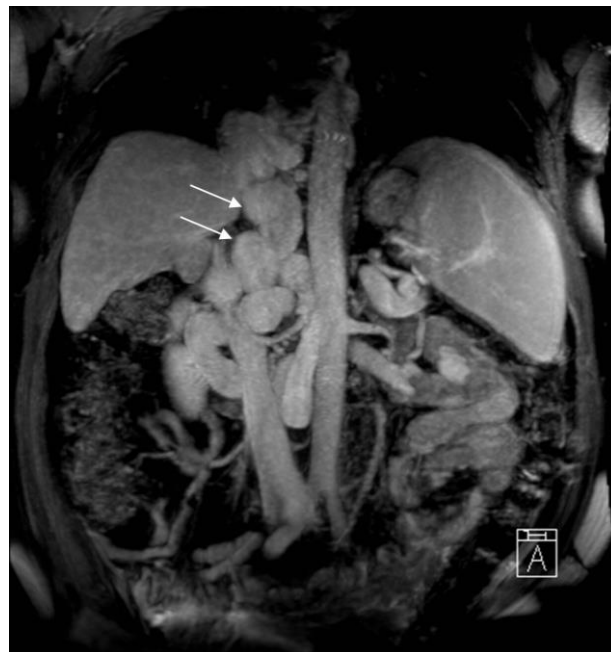
## TEACHING POINT

Spontaneous portoazygos shunt is a rare portosystemic shunt in an adult patient with liver cirrhosis and portal hypertension. Contrast-enhanced cross-sectional imaging with CT or MRI provides a detailed evaluation of collateral pathways between the portal and systemic veins that may influence clinical management.

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## FIGURES

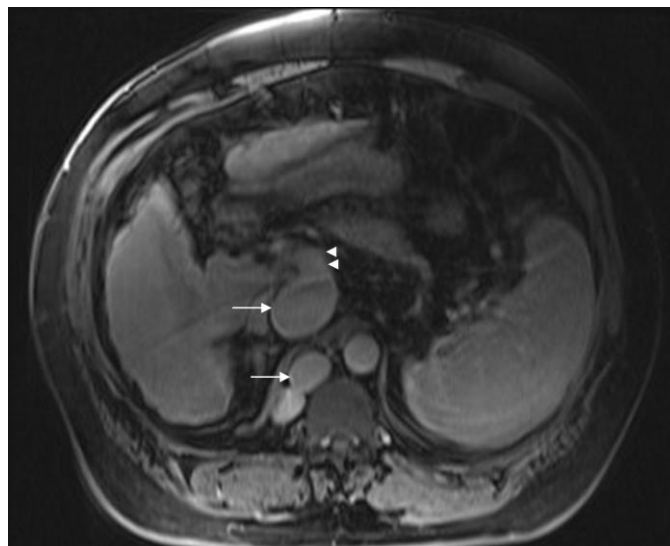
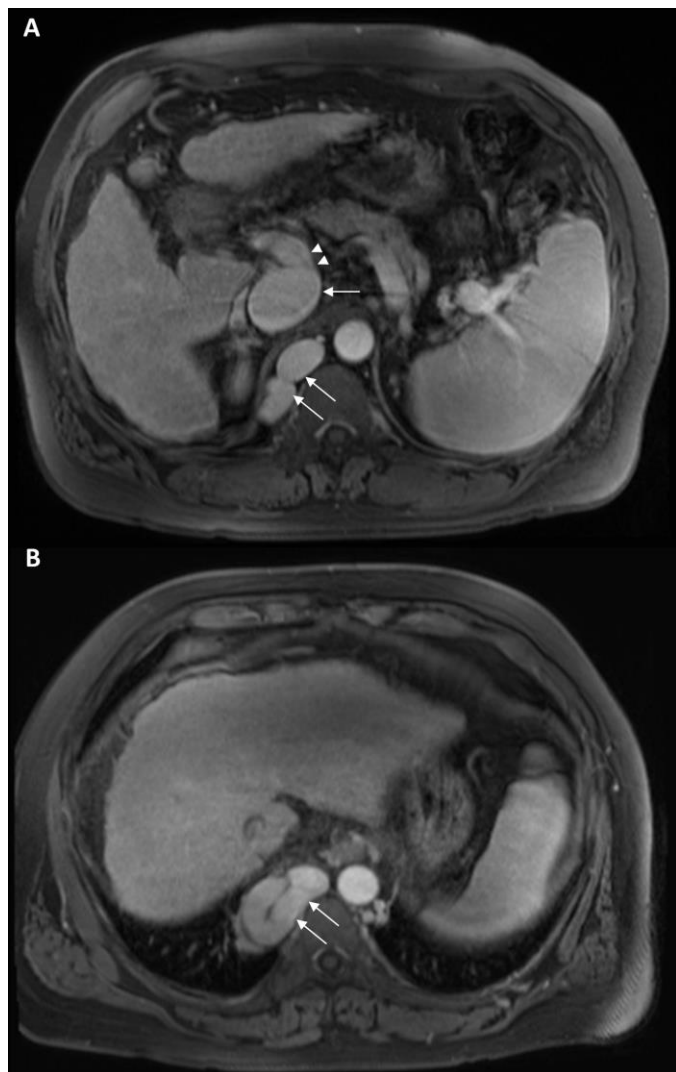


**Figure 1:** A 46-year-old male with spontaneous portoazygos shunt and liver cirrhosis secondary to alcohol and hepatitis C. Coronal reformatted image obtained from contrast enhanced 3D VIBE (Gadobenate dimeglumine, Multihance, Bracco Diagnostics, Princeton, NJ, USA, 0.1mmol/kg) shows a markedly dilated (3.9 cm in maximum diameter) azygos vein (arrows) located between the liver and the aorta (1.5T MR imaging system, Avanto, Siemens, Malvern, NJ, USA; TR=4.3, TE=1.88, matrix 320x65, FOV=41cm, slice thickness/spacing=3/0mm).



**Figure 2:** A 46-year-old male with spontaneous portoazygos shunt and liver cirrhosis secondary to alcohol and hepatitis C. Sagittal reformatted image obtained from contrast enhanced 3D VIBE (Gadobenate dimeglumine, Multihance, Bracco

Diagnostics, Princeton, NJ, USA, 0.1mmol/kg) shows a dilated azygos vein (long arrow) in communication with the portal vein (short arrow) (1.5T MR imaging system, Avanto, Siemens, Malvern, NJ, USA; TR=4.3, TE=1.88, matrix 320x65, FOV=41cm, slice thickness/spacing=3/0mm).



**Figure 4 (top):** A 46-year-old male with spontaneous portoazygos shunt and liver cirrhosis secondary to alcohol and hepatitis C. One-year follow-up axial contrast enhanced 3D VIBE (Gadobenate dimeglumine, Multihance, Bracco Diagnostics, Princeton, NJ, USA, 0.1mmol/kg) image shows no interval change in size and shape of dilated azygos vein (arrows) with connection to the posterior aspect of the portal vein (arrowheads) (1.5T MR imaging system, Avanto, Siemens, Malvern, NJ, USA; TR=4.7, TE=2.4, matrix 320x65, FOV=41 cm, slice thickness/spacing= 3.5/0mm).

**Figure 3 (left):** A 46-year-old male with spontaneous portoazygos shunt and liver cirrhosis secondary to alcohol and hepatitis C. A, B) Axial contrast enhanced 3D VIBE (Gadobenate dimeglumine, Multihance, Bracco Diagnostics, Princeton, NJ, USA, 0.1mmol/kg) images showing a dilated azygos vein (arrows) with connection to the posterior aspect of the portal vein (arrowheads) (1.5T MR imaging system, Avanto, Siemens, Malvern, NJ, USA; TR=4.3, TE=1.91, matrix 320x65, FOV=41cm, slice thickness/spacing= 3/0mm).

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<b>Etiology</b>	Recanalization of potential channel between the portal vein and systemic veins due to portal hypertension
<b>Incidence</b>	Rare, only one case of congenital portoazygos shunt in a neonate with thoracoabdominal duplication and absent intrahepatic portal vein has been reported [5].
<b>Gender ratio</b>	Unknown
<b>Age predilection</b>	Unknown
<b>Risk factors</b>	History of portal hypertension, liver cirrhosis, or congenital absence of intrahepatic portal vein
<b>Treatment</b>	Transvenous coil embolization by interventional radiology, surgical ligation.
<b>Prognosis</b>	Typically asymptomatic but can cause hepatic encephalopathy or variceal bleeding
<b>Findings on imaging</b>	Originates from the lateral posterior aspect of the main portal vein, extends to the right aspect of the lumbar vertebral body, ascends along the right aspect of the thoracic vertebral bodies to the mediastinum, and drains into the superior vena cava.

**Table 1:** Summary table for spontaneous portoazygos shunt

Portosystemic collateral veins	Location	MRI	CT	Clinical significance
<b>Esophageal collateral veins</b>	<ul style="list-style-type: none"> <li>Located in the wall of the esophagus</li> </ul>	<ul style="list-style-type: none"> <li>Thick wall of the esophagus with heterogeneously low signal intensity on T2WI</li> <li>Bright on T1WI with contrast enhancement</li> </ul>	<ul style="list-style-type: none"> <li>Difficult to see on nonenhanced CT</li> <li>Thick wall of the esophagus with hyperdensity on contrast-enhanced CT</li> </ul>	<ul style="list-style-type: none"> <li>Endoscopy is more sensitive in diagnosis.</li> <li>Esophageal bleeding</li> </ul>
<b>Paraesophageal collateral veins</b>	<ul style="list-style-type: none"> <li>Located outside the wall of the esophagus</li> </ul>	<ul style="list-style-type: none"> <li>Signal void tubular/tortuous structures outside the wall of the esophagus on T2WI</li> <li>Bright on T1WI with contrast enhancement</li> </ul>	<ul style="list-style-type: none"> <li>Hyperdense tubular/tortuous structures along the outside the wall of the esophagus on contrast-enhanced CT</li> <li>May simulate posterior mediastinal mass on nonenhanced CT</li> </ul>	<ul style="list-style-type: none"> <li>Associated with gastric varices</li> </ul>
<b>Gastric collateral veins</b>	<ul style="list-style-type: none"> <li>Located at the posterior and superior aspect of the gastric fundus</li> <li>Occasionally drains into the left renal vein [3]</li> </ul>	<ul style="list-style-type: none"> <li>Tubular/tortuous structures with signal void or heterogeneous low signal intensity on T2WI</li> <li>Bright on T1WI with contrast enhancement</li> </ul>	<ul style="list-style-type: none"> <li>May simulate gastric mass on nonenhanced CT</li> <li>Hyperdense tubular/tortuous structures on contrast-enhanced CT</li> </ul>	<ul style="list-style-type: none"> <li>Commonly associated with esophageal or paraesophageal varices</li> <li>Gastric bleeding</li> <li>Hepatic encephalopathy</li> </ul>
<b>Paraumbilical collateral veins</b>	<ul style="list-style-type: none"> <li>Originates from the left portal vein</li> <li>Drains through the epigastric and external iliac veins</li> </ul>	<ul style="list-style-type: none"> <li>Signal void on T2WI</li> <li>Bright on T1WI with contrast enhancement</li> </ul>	<ul style="list-style-type: none"> <li>Hyperdense tubular/tortuous structures on contrast-enhanced CT</li> </ul>	<ul style="list-style-type: none"> <li>Not associated with gastrointestinal bleeding</li> <li>Associated with varicose veins around the umbilicus (caput medusa)</li> </ul>
<b>Splenorenal collateral veins</b>	<ul style="list-style-type: none"> <li>Between splenic hilum and the left renal vein</li> </ul>	<ul style="list-style-type: none"> <li>Signal void on T2WI</li> <li>Bright on T1WI with contrast enhancement</li> <li>Enlarged left renal vein</li> </ul>	<ul style="list-style-type: none"> <li>Hyperdense tubular/tortuous structures on contrast-enhanced CT</li> <li>Enlarged left renal vein</li> </ul>	<ul style="list-style-type: none"> <li>Hepatic encephalopathy</li> </ul>
<b>Mesenteric collateral veins</b>	<ul style="list-style-type: none"> <li>Between the superior hemorrhoidal vein and the middle/inferior hemorrhoidal veins via the hemorrhoidal plexus</li> <li>Mesocaval shunt between the IMV and IVC through lumbar and retroperitoneal veins</li> </ul>	<ul style="list-style-type: none"> <li>Tubular/tortuous structures with signal void or heterogeneous low signal intensity on T2WI</li> <li>Bright on T1WI with contrast enhancement</li> </ul>	<ul style="list-style-type: none"> <li>Can be mistaken for a rectal mass protruding into the rectal lumen on nonenhanced CT [3]</li> <li>Hyperdense lobulated mass protruding into the rectal lumen on contrast-enhanced CT [3]</li> </ul>	<ul style="list-style-type: none"> <li>Rectal bleeding</li> </ul>
<b>Portoazygos collateral vein</b>	<ul style="list-style-type: none"> <li>Between the posterior aspect of the main portal vein and the azygos vein along the right aspect of the thoracolumbar vertebrae</li> </ul>	<ul style="list-style-type: none"> <li>Tubular/tortuous structures with signal void on T2WI</li> <li>Bright on T1WI with contrast enhancement</li> </ul>	<ul style="list-style-type: none"> <li>Hyperdense tubular/tortuous structures on contrast-enhanced CT</li> </ul>	<ul style="list-style-type: none"> <li>Hepatic encephalopathy</li> </ul>

**Table 2:** Differential diagnosis table for portosystemic shunts

#### ABBREVIATIONS

CT: computed tomography  
 IMV: inferior mesenteric vein  
 IVC: inferior vena cava  
 MELD: model for end-stage liver disease  
 MRA: magnetic resonance angiography  
 MRI: magnetic resonance imaging  
 SMV: superior mesenteric vein  
 SVC: superior vena cava  
 VIBE: volumetric interpolated breath-hold examination

#### KEYWORDS

Portal Hypertension; Portosystemic shunt; CT; MRI; Portoazygos shunt

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