Inferior Mesocaval shunt as a rare spontaneous portosystemic shunt

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Learning objectives

- Describe the embryology of extrahepatic portosystemic shunts
- Recognize the characteristic imaging findings of inferior mesenteric vein shunts (morphology, pathway, drainage level, and size), classify them in different types and search potential association with other portosystemic shunts
- Analyze their connection with portal hypertension, hepatic encephalopathy or hepatic diseases

Background

PORTAL SYSTEMIC COLLATERAL VESSELS:

Portosystemic collateral vessels develop in patients with portal hypertension as a way to decompress the splanchic flow, and they can be divided into two groups depending on whether they drain toward the superior vena cava (SVC) or the inferior vena cava (IVC).

- Collateral vessels draining into the SVC include the left gastric vein (coronary), short gastric vein, posterior gastric vein, gastric varices, esophageal and paraesophageal varices.

- Vessels draining into the IVC include gastrorenal and splenorenal shunts, paraumbilical vein and abdominal wall veins, retroperitoneal and mesenteric shunts, mesenteric varices, gallbladder varices and omental collateral vessels.

Portosystemic shunts can be classified in intrahepatic or extrahepatic. Although the intrahepatic shunts have been the most common documented, the extrahepatic ones also have an important role, and can be also subdivided depending on their relation with superior mesenteric vein (SMV) or, less frequent, with inferior mesenteric vein (IMV).

Inferior mesenteric vein shunts (IMVS) connect IMV to inferior vena cava (IVC), and are an exceedingly rare type of shunt with few cases reported in the literature. They have a tortuous appearance, and have been described mainly in portal hypertension clinical situations or secondary to postsurgical fascia adhesions, although some of them are considered to be congenital.

Until recently it was thought that formation of collateral circulation was due to the passive opening of vascular channels in response to increased portal pressure. However, some recent studies have shown that in splanchic neovascularization may also be involved proangiogenic molecules like vascular endothelial growth factor (VEGF) or platelet-derived growth factor (PDGF).
EMBRIOLOGY:

In the fifth week of intrauterine life, there are three major paired veins in the abdomen: the vitelline (or also called onphalomesenteric), the umbilical and the cardinal veins.

The *vitelline veins* form the hepatic sinusoids. The left vitelline vein subsequently involutes and the blood is diverted into the right vitelline vein, which enlarges and forms the hepatocardiac portion of the inferior vena cava (IVC). Derivatives of the vitelline veins also form the hepatic and portal vein (Table 1).

![Diagram of hepatic venous system development](image)

**Table 1**: Development of the hepatic venous system. Vitelline veins form the hepatic sinusoids. Left umbilical vein connects to the right vitelline vein via the ductus venosus. Anastomotic network around the duodenum forms the portal vein. LCV: left cardinal vein; RCV: right cardinal vein; RUV: right umbilical vein; LUV: left umbilical vein.

**References**: Maria Pardo

The *cardinal* venous system consists of the anterior, posterior, and common cardinal veins. Afterwards, between fifth and seventh weeks, the cardinal venous system is formed additionally with supracardinal, subcardinal and sacrocardinal veins.
The IVC is formed between the sixth and eighth gestational weeks sequentially by the formation, anastomoses and posterior regression of *three paired embryonic veins*: the posterior cardinal, the subcardinal, and the supracardinal veins (Table 2). It is composed of four segments: infrarenal, renal, suprarenal, and hepatic segments. The subcardinal system gives rise to the prerenal segment of the IVC. The anastomosis between subcardinal and supracardinal systems forms the renal vein segment. The supracardinal system gives rise to the postrenal segment of the IVC.

**Table 2**: Development of the cardinal venous system from weeks 5-7 to birth. a) At 5-7 weeks. b) At term.

**References**: Maria Pardo

Aberrations in the complex embryogenesis of the IVC recently mentioned above result in several potential anatomic variants, present in approximately 4% of the population.

**IMV ANATOMY**:

On cross sectional imaging, the IMV is visualized running in the left paraduodenal space and forming an arc cephalad to the duodenojejunal junction before its termination. Its
major tributaries are the superior hemorrhoidal vein, the sigmoid vein and the left colic vein.

Usually the IMV terminates into the splenic vein, but anatomical variations can include the IMV draining into the splenoportal angle and the IMV draining into the SMV.

Extrahepatic portosystemic venous shunts are caused by persistent communication between the vitelline vein and subcardinal veins, resulting in abnormal confluence of the portal vein and the inferior vena cava tributaries. Since Retzius reported on, several anastomoses between the portal venous system and the IVC in the retroperitoneum, anastomoses between branches of the SMV or IMV and the IVC have generally been called the veins of Retzius.

These retroperitoneal pathways between the tributaries of the mesenteric vein and vena cava are considered a remnant of fetal communication, and it is often difficult to completely trace the drainage course due to the complex and often extensive nature of these collateral vessels that appear as small, rounded or tubular areas of increased attenuation that enhance to the same degree as the mesenteric veins.

A similar prevalence of veins of Retzius has been described in patients with and without portal hypertension.

Portal systemic encephalopathy in patients without liver cirrhosis or portal hypertension has also been described, although it is extremely rare.

**Imaging findings OR Procedure details**

We retrospectively reviewed 12 cases of inferior mesocaval shunts (IMS) identified in our tertiary referral hospital between 2004 and 2010, including 6 men and 6 women aged between 41-82 years (mean age 64.7 years), who underwent abdominal MDCT for different purposes, the most frequent one for evaluating liver disease. This is, to the best of our knowledge, the largest serie reported (Table 3).

10 of 12 patients had chronic hepatopathy, including Hepatitis B Virus, alcoholic diseases and unknown cause, and none of them had a history of abdominal trauma or surgery.

Studies were performed with a 4-row (n=3) or 16-row (n=9) MDCT scanner.

We revised imaging findings paying special attention to venous phase in axial plane and also in coronal multiplanar reconstruction (MPR). Maximum intensity projection (MIP)
and volume rendering (VR) were systematically performed on a workstation to analyze portosystemic collateral vessels.

Studies were analyzed by three different radiologists, assessing the pathway, the drainage level and the size of the shunt, as well as the presence of other portosystemic shunts that can lead to encephalopathy.

ANALYSIS OF RESULTS

IMV-systemic shunts are rarely encountered, although they have been reported with a frequency of about 13% in portal hypertension (including via the rectal vein, a type of shunt not depicted in our serie).

Mesenteric collateral vessels may arise from the inferior mesenteric veins, and may ultimately drain into the systemic venous system via the retroperitoneal or pelvic veins. It is usually difficult to trace their drainage due to the tortuous, cirrloid or dilated course.

We identified 17 shunts in 12 patients, being most of them (9/12) formed by a single drainage vein (Figures 1, 2, 3, 5, 6, 7, 8, 9, 12, 13, and 14), mainly to the IVC. However, several drainage veins have been reported, being observed in our series one patient with two (Figure 4) and two patients with three portosystemic connections (Figures 10 and 11) in most cases draining directly into the IVC. Recognition of these drainage pathways is important to guide the treatment.

The mean size of the shunts was 7'2 mm (ranging from 4 to 13 mm).

Noteworthy that the largest sizes (12 and 13 mm) were observed in those who had until 3 shunts. The lowest sizes (4 and 5 mm) were visualized in those patients without liver cirrhosis or portal hypertension, and are considered to be congenital, particularly when the shunt vessel is single, and occurred in one young patient.

Several different pathways from IMV toward IVC have been described.

Despite of the small percentage of cases described in literature of direct flow from the IMV toward the IVC and directly to the IVC (mesenteric-caval), we have observed it in 12 of 17 (70 %) of the portosystemic shunts, passing the aorta indistinctly anteriorly (n=6) (Figure 1, 2, 8, 12 and 13) or posteriorly (n=6) (Figures 4, 10, 11). We have noticed that the 3 patients who presented with encephalopathy had developed this last type of drainage, with mean size of the shunt of 9'3 mm (8, 8 and 12 mm), slightly higher than de mean size of all the shunts (7'2 mm).
Two cases presented with drainage through the left renal vein, (mesenteric-renal) (Figures 5, and 10), having both of them a large diameter (12 and 13 mm).

The mesenteric-gonadal shunt are often due to ileocolic veins from the superior mesenteric vein, anastomosed with right gonadal vein, while the anastomosis between the left gonadal vein with a venous network developed from the IMV is exceedingly rare, although we have identified this type of pathway in 3 cases (Figures 4, 6, and 9). In such cases, a variceal plexus around the uterus may develop (Figure 9), affect the opposite gonadal vein and even produce ileocecal varices.

Another type of drainage pathway described is toward the internal iliac veins (mesenteric-iliac), not identified in our series.

Two patients presented 3 drainage pathways, one of them with all connections toward the IVC, meanwhile the other patient with two shunts to the IVC and the other one to the left renal vein.

In 7 cases the IMV was larger (12’5 mm of mean size) than the trunk of the portal vein (10’8 mm of mean size).

Although our studies are not dynamic studies for evaluating vascular flows, we observed in 4 patients a jet within the IVC at the shunt level (representing inflow from an opacified drainage pathway toward an unenhanced IVC) (Figure 3) and a "flow artifact" in left renal vein at the shunt level during the arterial phase (representing inflow from unenhanced collateral vessel toward a high opacified left renal vein) (Figure 9).

SYMPTOMS RESULTING FROM THE SHUNT
The clinical manifestations of IMV-IVS shunts are the result of the retrograde direction of flow from the IMV and the size of the shunt. In cirrhotic patients, the development of hepatic encephalopathy depends largely on the function of residual hepatocytes and the portal blood diverted to the systemic circulation.

In our serie 3 patients presented encephalopathy (disorientation and/or disturbance of consciousness), corresponding all of them to shunts toward the IVC and with a mean size of 9’3 mm, slightly higher than the mean size of all the shunts (7’3 mm).

The IMV-IVC shunt is not associated with gastrointestinal bleeding, although exceptional ruptures into the retroperitoneal cavity have a severe prognosis.

TREATMENT
In some series it's been observed that with treatment there's usually improve of symptoms secondary to portosystemic encephalopathy, coinciding with decrease of the serum ammonia levels.

The treatment of symptomatic shunts includes conservative therapy in most cases, related to the symptomatolgy (encephalopathy), but surgical ligation, and transcatheter embolization (percutaneous transhepatic obliteration or retrograde transcaval obliteration) have also been described.

One case of our serie was treated with embolization, placing an Amplatzer Vascular Plug (AVP) in the IMV. A follow-up MDCT demonstrated an important decrease of IMV size, although persistence of the portosystemic shunt was detected through the left colic vein, as well as dilated superior haemorrhoidal veins, not present before Amplatzer placement (Figure 14).

However, the therapeutic blockade in some cases can lead to overload of the portal venous system, increasing the risk for ascites formation and variceal bleeding. This is the reason why it's widely believed that the management of these patients should be judged according to their characteristic risk/benefit ratio.

Some recent studies have shown that proangiogenic molecules like VEGF or PDGF may have an important role in splanchnic neovascularization. According to this, antiangiogenic therapies like sorafenib® could be useful to reduce portosystemic collateral circulation, improve splanchnic hyperdynamics and decrease portal pressure.

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**Table 3:** Table depicts results observed in our 12 patients with portosystemic shunts.
Fig. 1: Case 1. Axial CT images and volume rendering reconstruction show a dilated inferior mesenteric vein (yellow arrows), and mesenteric varices (arrow heads), with a single draining vein (red arrow) into the dilated inferior vena cava.
**Fig. 2:** MDCT volume rendering reconstruction corresponding to case 1 (same as figure 1), clearly shows the inferior mesenteric vein shunt through a tortuous vein passing the abdominal aorta anteriorly toward the inferior vena cava.

**Fig. 3:** Case 2. Axial CT image demonstrates an 8 mm shunt to the inferior vena cava (red arrow). Coronal reconstruction depicts the draining vein (red arrow) and a "jet" flow into the dilated IVC (arrowhead). Volume rendering reconstruction shows the direct IMV-IVC shunt (red arrow: draining vein, yellow arrows: IMV). Note the dilated IMV, larger than the portal vein (white arrow).
Fig. 4: Case 3: IMV-systemic shunt through two drainage veins. The coronal MIP reconstruction shows a shunt to a dilated left gonadal vein (white arrow) that drains into the left renal vein (asterisk). Note the normal size of the gonadal vein (arrowheads) caudal to the shunt. The axial CT image demonstrates a retroaortic shunt (red arrow) to the IVC. The volume rendering reconstruction depicts the dilated IMV (yellow arrows), and tortuous retroperitoneal varices.
Fig. 5: Case 4: Portosystemic shut from IMV to Left renal vein. Volume rendering image shows connection between a dilated IMV (yellow arrows) and left renal vein (asterisk) through one single drainage vein (white arrow). Coronal MIP reconstruction depicts left renal vein shunt (LRV), next to the left gonadal vein drainage (white arrows).
**Fig. 6**: Case 5: Shunt between IMV and left renal vein. Volume rendering image shows the portosystemic shunt with a dilated IMV (yellow arrows). MIP reconstruction depicts retroperitoneal varices originating from IMV and draining to a dilated left gonadal vein (red arrow), meanwhile the distal segment maintains a smaller calibre (white arrows).
**Fig. 7:** Volume rendering reconstruction of case 5 (same as Figure 6), which shows shunt between IMV and left renal vein.

**Fig. 8:** Case 6: Direct connection between IMV and IV. Volume rendering reconstruction (left) and coronal MIP reconstruction (right) depict dilatation of IMV (yellow arrows) and connection with medial aspect of the IVC (asterisk) through a single drainage vein (red arrow) with a preaortic course.
**Fig. 9:** Case 7: Shunt between IMV and right gonadal vein. Coronal volume rendering reconstruction (left) shows dilatation of the IMV (yellow arrows), multiple gonadal varices and dilatation of the left gonadal vein (red arrows). Variceal plexus around the uterus connect with the opposite gonadal vein, which is also slightly dilated (white arrows). Blood from the IMV is drained to the left renal vein through the left gonadal vein and directly to the IVC through the left gonadal vein. Curved reconstruction in the late arterial phase (on the right) depicts a flow artifact (blue arrow) in the left renal vein (asterisk) caused by the unenhanced blood from the left gonadal vein (red arrows) entering into the enhanced left renal vein.
Fig. 10: Case 8: IMV-left renal vein and IVC shunts. Volume rendering reconstruction (left) demonstrates systemic shunt between IMV and LRV (arrow) and the dilatation of the IVC (asterisk). In axial and coronal images (right) we observe a shunt with LRV (white arrow) but also two other shunts with IVC, which is shown dilated and heterogeneous due to inflow thereof.
Fig. 11: Case 9: IMV-IVC shunt. Axial CT images (left) and coronal reconstruction (middle) depict the presence of three retroperitoneal drainage veins toward IVC (yellow arrows), all of them passing the aorta posteriorly. Volume rendering reconstruction (right) shows dilatation of the IMV (white arrows) and presence of tortuous varices draining to it. Note also a dilated IVC (asterisks).
**Fig. 12:** Case 11: IMV-IVC shunt. Volume rendering and coronal MPR demonstrate a dilated IMV (yellow arrows) and a single draining vein (white arrows) anterior to the abdominal aorta and entering a dilated IVC (asterisks).
Fig. 13: Case 12: IMV-IVC shunt. Volume rendering image (left) shows dilated IVC and IMV (yellow arrows), with tortuous and large distal veins. Angiographic study of the celiac trunk in venous return phase (a) depicts an inverted flow in IMV, which is dilated (yellow arrows). Angiographic study of the IVC (b) confirms the existence of an IMV-IVC shunt through a preaortic branch (white arrow) that communicates with the IMV through a network of varicose vessels. The shunt was embolized using an Amplatzer Vascular Plug in the IMV (not shown).
**Fig. 14:** Case 12: Persistence of a systemic shunt. Coronal MIP image (left) and volume rendering reconstruction (right) show an Amplatzer Vascular Plug in IMV (white arrows), with an important decrease of its calibre (yellow arrows), though we observe a persistence of a portosystemic shunt through the left colic vein (red arrows), as well as dilated superior haemorrhoidal veins (blue arrows), not present before Amplatzer placement. Volume rendering reconstruction demonstrates persistence of the communication with the IVC (white arrowhead).
Conclusion

- It's important to recognize the MDCT findings of IMVS that allow its identification and classification, since there are important clinical manifestations associated.
- Knowledge of the imaging features and the drainage pathways are also critical to guide the treatment strategy for symptomatic shunts.

References


**Personal Information**