Portosystemic collateral pathways and interventions in portal hypertension

Murad Feroz Bandali,1 Anirudh Mirakhur2,*

ABSTRACT

Pathologic increase in portal pressure can be caused by increased resistance to blood flow at the level of the portal vein (pre-hepatic), hepatic sinusoids (hepatic) or hepatovenous outflow (post-hepatic). This results in recruitment and dilatation of tiny portosystemic collateral pathways, diverting portal venous blood flow to low pressure systemic veins. Based on the location of the causative factor of portal venous resistance, different collateral pathways and shunts may develop, resulting in unique syndromes of portal hypertension and in-turn requiring unique treatment options. Knowledge of the common and less-common portosystemic collateral pathways have important implication for clinicians and interventionalists. The objective of this pictorial review is to illustrate the various collateral pathways using diagrammatic and conventional non-invasive and invasive radiologic examples. Additionally, we will briefly address minimally invasive interventional techniques used to treat the sequelae of portal hypertension.

Keywords: Hypertension, portal; Portal hypertension; Radiology, interventional; Varices

Introduction

Portal venous system is a distinctive network of vessels that connect two separate vascular beds, the gastrointestinal/splanchnic veins and the hepatic sinusoids. The mesenteric and splenic veins join to form the portal vein, which in turn branches at the liver and terminates in the hepatic sinusoids. While the main feeding branches of the portal vein include the superior mesenteric vein (SMV) and splenic vein, and sometimes the inferior mesenteric vein (IMV), other tributaries also feed into the portal venous vasculature—creating a complex network of vessel that is functionally isolated from systemic circulation (Fig. 1). The portal vein functions to carry blood and nutrients from the splanchnic vasculature to the liver for metabolism, nutrient storage and detoxification.

A pathologic increase in portal pressure (above 5–10 mmHg) can be caused by increased resistance to blood flow at the level of the portal vein (pre-hepatic), hepatic sinusoids (hepatic) or hepatovenous outflow (post-hepatic). Portal pressure is measured by catheter-based hepatic venography, which indirectly measures the portal venous pressure and compares it to the systemic venous pressure in the inferior vena cava (IVC), termed the hepatovenous pressure gradient (HVPG). The HVPG is normally between 1–5 mmHg and portal hypertension is defined by HVPG greater than 10 mmHg.14 Caused by increased resistance to hepatopetal flow, portal hypertension is further aggravated by reactive and progressive splanchnic vasodilatation, as well as, hepatic sinusoidal endothelial dysfunction. Eventually, this leads to recruitment of collateral venous channels that bypass the liver and connect directly to systemic venous circulation, forming a network of shunts and end-organ varices. This is a complex process involving the recanalization, dilatation and hypertrophy of pre-existing portosystemic vascular channels, as well as, a possible component of angiogenesis.1,5–7

Based on the location of the causative factor of portal venous resistance, different collateral pathways and shunts may develop, resulting in unique syndromes of portal hypertension and in-turn requiring unique treatment options. Understanding of common, clinically relevant, portosystemic collaterals pathways is integral for any physician who aims to treat this complex condition.

In this review, we aim to discuss the spectrum of portosystemic collateral pathways in the abdomen and thorax, using imaging...
and pictorial examples. Additionally, a brief overview of minimally invasive interventional techniques will also be presented.

Basic Anatomy of Portosystemic Collaterals

Shunts are defined as collateral veins that simply bridge the portal venous and systemic venous systems. Varices, in contrast, are dilated end-organ capillary beds that have a propensity to bleed. The most common pathway of hepatofugal flow is via the coronary vein, forming esophageal, paraesophageal and cardiophrenic varices. Other pathways include gastric, paraumbilical and mesenteric venous channels. Pleuro-pericardial, peritoneal, pancreaticoduodenal, splenoazygos and mesocaval collaterals are far less common but may also be recruited to decompress the portal vein (Fig. 2). On cross-sectional imaging such as computed tomography (CT) or magnetic resonance imaging (MRI), shunts appear as well defined smooth, round, serpiginous and tubular structures which are separate from the viscera and enhance to a same degree as the adjacent normal portal or mesenteric veins. Varices have a similar appearance but are in a mural or submucosal location and are commonly seen in the walls of the hollow viscera such as the esophagus, stomach, and rectum.

Coronary Vein Pathways

The coronary vein, or the left gastric vein, is situated within the lesser omentum and is the most common collateral pathway recruited in portal hypertension secondary to liver cirrhosis, occurring in an estimated 80% of cross sectional imaging studies. Typically, they appear as dilated collateral veins along the lesser curvature of the stomach and gastroesophageal junction. A coronary vein larger than 5 to 6 mm is a strong indicator of portal hypertension. These are also commonly accompanied by esophageal or paraesophageal varices. The anterior branch of the coronary vein typically supplies esophageal varices, while the posterior branch supplies paraesophageal varices (Fig. 3).

After forming a subepithelial and submucosal venous network

---

Fig. 1. Normal portal venous anatomy. Main portal vein (MPV) most commonly forms from the joining of the splenic vein (SV) and superior mesenteric vein (SMV). The inferior mesenteric vein (IMV) has a variable draining position but most common location is into the SV. Coronary vein (or left gastric vein [LGV]) courses along the lesser curvature of the stomach and drains directly into the MPV. As the MPV reaches the liver, it branches into the right portal vein and left portal vein, respectively.

Fig. 2. Hepatofugal portosystemic pathways in portal hypertension. Progressive resistance to portal venous blood flow results in decompression through pre-existing collateral pathways. Paraumbilical (PuVa) and abdominal wall (AwVa) varices develop after recanalization of the paraumbilical vein. Esophageal (EsoVa), Paraesophageal (PEsoVa), and Cardiophrenic (CPVa) develop when blood flow decompresses via the left gastric vein (LGV). Mesenteric (MVa) and rectal (RVA) varices may also develop to allow passage of portal venous blood into systemic circulation. MPV, main portal vein; GrSh, gastrorenal shunt; SrSh, splenorenal shunt; SV, splenic vein; SMV, superior mesenteric vein; IMV, inferior mesenteric vein.

Fig. 3. Axial contrast-enhanced computed tomography image (A) and thick-slab three-dimensional reformat image (B) of the abdomen and pelvis in a 54-year-old female with hepatitis C induced cirrhosis. Massive paraesophageal varices are seen surrounding the distal esophagus (asterisk) with a markedly dilated feeding coronary vein which arises from the proximal main portal vein (arrow).
within the esophageal wall, esophageal varices usually drain into the ayzygous or hemiazygous system. Their typical CT/MRI appearance is nodular and intraluminal protrusions with scalloped borders (Fig. 4). Formation of these varices is clinically significant in portal hypertension as they have a high propensity to hemorrhage into the esophageal and gastric lumen, at a rate of 10% to 30% per year, with an overall mortality of 20% to 35%. While cross-sectional imaging, is highly sensitive at the detection of large, clinically relevant, esophageal varices; endoscopy remains the mainstay of identification and diagnosis.

Parasoesophageal varices, in contrast, surround and are directly adjacent to the esophagus and descending thoracic aorta and form a network of dilated veins which are continuous with the coronary veins and can be followed superiorly to the ayzygous/ hemiazygous veins and paravertebral venous plexus. Unlike esophageal varices, they cannot be seen on endoscopy and require cross-sectional imaging for diagnosis (Fig. 3A). While they do not result in variceal hemorrhage, the presence of paraeosophageal varices on chest CT portends a poor prognosis for patients who have existing esophageal varices irrespective of whether they have undergone sclerotherapy.

Cardiophrenic varices also result from recruitment of the coronary vein collateral pathway and consist of dilated pericardial and cardiophrenic veins. On imaging, they manifest of as undulating lesions with venous enhancement along the inferior cardiac borders and cardiophrenic angles and may simulate a cardiophrenic mass at radiography. They care commonly seen in patients with post-hepatic causes portal hypertension, appearing in 18% of cases of cirrhosis secondary to membranous obstruction of the IVC.

Several strategies exist for treating esophageal varices. For patients with medium to large sized esophageal varices, non-selective beta-blockers, endoscopic ligation and/or balloon tamponade should be considered as initial therapies. However, patient who do not respond to these therapies, image-guided interventions may be considered. Transjugular intrahepatic portosystemic shunt (TIPS) is an image-guided procedure where a transhepatic shunt is created using a large needle-trocar set, angioplasty balloon and polytetrafluoroethylene (PTFE)-covered stents; creating a parenchymal tract between the portal and hepatic veins and reducing HVPG below 12 mmHg (Fig. 5). TIPS has shown up to a threefold decrease in recurrent variceal bleeding when compared to endoscopic therapy. Strong evidence also exists for TIPS placement improving transplant-free survival in cirrhotic patients who suffer from refractory ascites, when compared to intermittent large-volume paracentesis. Limited evidence exists for additional indications which include: acute gastropathy, hepatopulmonary syndrome, and Budd-Chiari syndrome (Fig. 6).

Unfortunately, TIPS is not without disadvantages. Deterioration of hepatic function can result from diversion of portal venous blood flow. Hepatic encephalopathy may develop due to the flow of non-detoxified blood to systemic circulation; now being allowed to bypass the liver parenchyma unimpeded. Additionally, frequent surveillance and potential maintenance procedure may also be needed to ensure long-term stent patency.

In some instances, optimal tract creation via the right or middle hepatic vein is not possible. In such cases, direct intrahepatic portocaval shunt (DIPS) may be created. Using intravascular ultrasound, a trans-caudate tract may be created directly from the IVC to the portal vein. DIPS has demonstrated shorter procedure times, as well as, may be ideal in cases of hepatic vein thrombosis, challenging anatomy or in where ideal parenchymal tract is not possible.

**Gastric Venous Collateral Pathways**

Gastric varices are less prevalent than esophageal varices but can be seen in up to 33% of patients with portal hypertension. Esophageal and gastric varices frequently co-exist with esophageal varices, as described in the commonly used Sarin classification for gastric varices (Table 1). Gastric varices are more likely to be supplied by the short gastric and posterior gastric veins, while esophageal varices are more likely to be supplied by the coronary veins. Rarely they are supplied by the gastroepiploic vein, typically in the context of endovascular or surgical exclusion of other feeding veins. Short gastric varices usually course along the lesser curvature of the stomach and drain...
into the splenic vein. Isolated short gastric varices are seen in the context of splenic vein stenosis or thrombosis. On cross sectional imaging, they typically appear as a tangle of vessels in the region of the splenic hilum and gastric fundus. It can often be difficult to distinguish between the gastric walls and individual vessels.

Gastric veins may drain into esophageal/paraesophageal varices in approximately 84% of cases.27 Occasionally gastric varices may drain via a gastrorenal shunt, which appears as a large left-sided inferior phrenic vein which connects the gastric varices to a dilated renal vein. This shunt may be recruited by existing tiny portosystemic collaterals from the adrenal venous system.27 Large gastric varices are commonly seen in the absence of esophageal varices when a gastrorenal shunt is seen. Alternative pathways include direct drainage into the IVC by way of the left inferior phrenic vein or pericardiophrenic veins. Other smaller drainage pathways include paravertebral venous plexus, intercostal veins ascending lumbar veins and azygous veins.10,27,28

Gastric varices tend to bleed at a lower rate than esophageal varices but they are associated with a higher overall mortality secondary to their larger size and higher flow rate.29 Additionally, life-threatening gastric variceal hemorrhage can occur at HVPG below 12 mmHg, making TIPS placement a less favorable option.30,31 Balloon-occluded retrograde transvenous obliteration of gastric varices (BRTO) is a technique which involves advancing paired catheters from femoral vein access through the outlet of a gastrorenal shunt (Fig. 7). Following balloon-occlusion of the shunt, the distal catheter is placed in the gastric variceal bed and is used to inject a sclerosing agent to obliterate the varices. The sclerosing agent and balloon-catheter are left in-place for 4 to 48 hours, prior to removal.30–32

BRTO has shown excellent efficacy at controlling variceal bleeding with low re-bleed rates and some have advocated for its employment as a prophylactic measure.25,33 BRTO is advantageous in that it diverts the blood flow away from collateral pathways; improving hepatopetal flow and overall hepatic reserve. As such, BRTO is also an excellent treatment option for refractory encephalopathy. It is also advantageous in that it does not require general anesthesia (unlike TIPS).32 However, occlusion of certain portosystemic shunts may aggravate other symptoms of portal hypertension: either worsening abdominopelvic ascites or aggra-

### Table 1 Sarin Endoscopic Classification for Grading Gastric Varices

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gastroesophageal varix type 1 (GOV-1)</td>
<td>2–5 cm below the gastroesophageal junction and continuous with esophageal varices which extend along the lesser curvature of the stomach.</td>
</tr>
<tr>
<td>Gastroesophageal varix type 2 (GOV-2)</td>
<td>Continuation from esophageal varices which extend along the lesser curvature of the stomach but are more tortuous than GOV-1.</td>
</tr>
<tr>
<td>Isolated gastric varix type 1 (IGV-1)</td>
<td>Absence of esophageal varices and are located at the gastric fundus. Varices are tortuous and complex.</td>
</tr>
<tr>
<td>Isolated gastric varix type 2 (IGV-2)</td>
<td>Absence of esophageal varices and are located at the gastric body, antrum or pylorus.</td>
</tr>
</tbody>
</table>

Fig. 6. A 27-year-old male with a history of schistosomiasis associated portal hypertension presents with recurrent upper gastrointestinal bleeding secondary to esophageal varices. (A) Digital subtraction angiography (DSA) performed through a pigtail catheter within the main portal vein after a Rosch-Uchida transjugular intrahepatic portosystemic shunt trochar-needle set (Cook Medical) are advanced from the right hepatic vein through the hepatic parenchyma into the right portal vein. Note the dilated coronary vein arising from the main portal vein and Linton balloon in the stomach. (B) Delayed DSA images demonstrate prominent and extensive esophageal varices and gastric varices along the lesser curvature of the stomach (asterisk). (C) Subsequently, polytetrafluoroethylene-covered stent was placed across the parenchymal tract (arrow). (D) Transvenous injection of liquid embolic agent in the coronary vein and esophageal varices.
vating esophageal varices. Additionally, severe adverse reactions may result from extended exposure to a sclerosing agent such as: anaphylaxis, portal vein thrombosis, diffuse intravascular coagu-
lopathy, renal dysfunction and pulmonary edema.31,34

A potential solution to this is employment of other occlusion
methods at the level of the gastrorenal shunt. Coil-assisted retro-
grade transvenous obliteration of gastric varices is a modified ver-
sion of BRTO, where dual microcatheters are employed but rather
than balloon occlusion, the gastrorenal shunt is coil-embolized.34 A
microcatheter is placed beyond the coil-pack and gel-foam
slurry is injected into the variceal bed. Both catheters are then
immediately withdrawn with coil-pack left in place; alleviating
the need for a sclerosing agent.34 A similar modification using a
vascular plug instead of embolization coils (termed PARTO: vas-
cular plug-assisted transvenous obliteration of gastric varices) has
demonstrated similar technical success and clinical efficacy for
the treatment of gastric varices and hepatic encephalopathy.15

Splenlic Vein Collateral Pathways

Splenic/perisplenic varices are seen in the anteroinferior re-
gion of the splenic hilum where they traverse the splenocolic liga-
ment. This collateral pathway may also communicate and serve
as a drainage pathway for gastric varices. Dilated and tortuous
splenic vein within the hilum of an enlarged spleen should not be
confused with perisplenic varices (Fig. 8).11

Splenic venous flow also commonly circumvents the liver
by way of large splenorenal or splenocaval shunts. Splenorenal
shunts appear as large tortuous veins connecting the splenic and
left renal vein; although the exact origin of the connection
along the splenic vein is usually difficult to discern.11 As with
gastrorenal shunts, the left renal vein is usually dilated. Rarely a
splenocaval shunt can develop, which extends inferiorly into the
pelvis and drains into the IVC via the internal iliac or gonadal
veins. Splenocavalous shunts are even less common, where a di-
lated venous connection can be seen from the splenic vein to the
hemiazygous or posterior abdominal wall/chest wall veins.10

Paraumbilical and Abdominal Wall Collateral Pathways

The paraumbilical vein is a fetal remnant which, when re-
canalized in the context of portal hypertension, arises from the
left portal vein and courses along the anterior edge of the falci-
form ligament to the abdominal wall (Fig. 9A). This is a common
portosystemic pathway in cirrhosis, seen in up to 30% to 35% of
cases.16 Typically this can be seen as 2 to 3 mm tubular and ser-
piginous vessels within the abdominal wall (Fig. 9B) which drain
via the superior epigastric vein and/or internal thoracic veins,
superior vena cava or to the IVC by way of the inferior epigastric
and external iliac veins.16 For patients with medically refrac-
tory encephalopathy, the paraumbilical vein is a common shunt
that can serve as a target for transvenous obliteration to achieve
symptomatic improvement (Fig. 9C).17,18

Surgically created ileostomies and colostomies within the
abdominal wall create portosystemic collaterals which develop
in portal hypertension, by way of unique mucocutaneous con-
nections.10,26,39 As a result, stomal varices are very common and
reported in up to 50% patient with surgical digestive stomas and
concomitant portal hypertension. Hemorrhage from these varices
is a common occurrence, present in up to 27% of patients with
stomal varices, and can be challenging to manage.39 Stomas that
are diffusely engorged with diffuse venous oozing tend to respond
favorably to TIPS therapy. Conversely, patients with focal stom-
al varical bleeding respond better to manual compression and
transvenous obliteration (which include BRTO, percutaneous tran-
shpatic obliteration, or trans-TIPS balloon-occluded antegrade
transvenous obliteration) (Fig. 10).40

Mesenteric and Omental Collateral Pathways

Omental varices can be seen in up to 30% of patients with
cirrhosis but are not considered common collateral pathways by
interventionalist, as they are not commonly seen at the time of
angiography.11 They tend to be much smaller than their counter-
parts elsewhere in the abdomen but are typically quite numerous
and should not be mistaken for peritoneal metastasis at cross sec-
tional imaging. On imaging, they appear as sparse tubular vessels

![Fig. 7. Schematic drawing demonstrating the BRTO procedure. Via femoral
vein access, a pair of catheters are advanced into the outlet of the gastrorenal
shunt. A balloon catheter is inflated and sclerosant is injected in the gastric
varices. The sclerosant and balloon are left inflated for 4 to 48 hours while
the patient is under close clinical surveillance. The balloon catheter is then
deflated and removed.](image1)

![Fig. 8. Axial computed tomography image depicting cirrhosis, splenomegaly
and associated perisplenic varices within the left upper quadrant. Note a large
dominant splenic varix posterior to stomach (arrow).](image2)
coursing through the greater omentum (Fig. 11). They arise from the SMV or IMV and drain into retroperitoneal or pelvic veins.\textsuperscript{10} There have been few but fatal reports of bleeding and rupture from omental varices.\textsuperscript{41,42}

Mesenteric varices appear as dilated and tortuous branches of the SMV and IMV within the mesenteric fat.\textsuperscript{11} They also drain into systemic circulation by way of the retroperitoneal or pelvic veins but rarely mesorenal shunts may develop between the mesenteric varices and the right renal vein or mesocaval shunts directly to the IVC.\textsuperscript{43} Of note, inferior mesenteric varices may drain into systemic circulation via the superior or middle rectal veins to the internal iliac veins. Rectal varices more commonly result in encephalopathy rather than life-threatening hemorrhage.\textsuperscript{44}

Other Collateral Pathways

Intrahepatic portal veins may also create direct venous connections with hepatic venous branches or direct communication with the coronary vein, typically via the left hepatic lobe.\textsuperscript{45} Pleuropericardial–peritoneal collaterals may develop as a loose venous plexus which pierces the diaphragm to join the pericardial, pleural and pulmonary veins.\textsuperscript{46} Pericholecystic varices in or outside the gallbladder wall are present in up to 12% of patients with portal hypertension but are most common in patients with extrahepatic portal vein obstruction.\textsuperscript{28}
Fig. 11. Axial computed tomography image depicting tiny omental collaterals along the left ventral abdomen (arrow).

Conclusion

A strong background knowledge of the basic anatomy of portosystemic collateral pathways is a necessity for any physician treating patients with portal hypertension. Not only is it essential for accurate diagnosis, appropriate characterization of portosystemic collateral pathways can lead to identification of causative factors, therapy selection and mitigate potential procedure-related complications. As non-invasive imaging techniques continue to advance, so too does the ability to visualize this complex disease process.

Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

Acknowledgments

We would like to acknowledge Salima Hirji for her contribution in creating the anatomical illustrations. M.F.B. and A.M. contributed to this review article with conception, literature review and analysis, image preparation and editing, manuscript drafting and critical revision and editing, and approval of the final version.

References


