Review of Applied Anatomy, Hemodynamics, and Endovascular Management of Ectopic Varices

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Abstract

Portal hypertension leads to the opening up of collateral pathways to bypass the occlusion or resistance in the portal system. Ectopic varices are formed by such collaterals at many various sites along the gastrointestinal tract other than the usual location, that is, gastroesophageal region. Early diagnosis of ectopic varices needs strong clinical suspicion and contrast-enhanced computed tomography scan as endoscopy may often fail to pinpoint a source. In contrast to gastric varices where the understanding of the disease, as well as endovascular management, is widely studied and documented, the same is not true for ectopic varices due to low incidence. Understanding the applied anatomy and hemodynamic classification is important to decide the most suitable therapy. Interventional radiological procedures are aimed at either decompressing the varices or obliterating them and depend on the patency of the portal system, underlying etiology, and local expertise.

Keywords
- ectopic varices
- BRTO
- BATO

Introduction

Cirrhosis and extrahepatic portal venous obstruction (EHPVO) lead to the opening up of collateral pathways to bypass the portal occlusion or resistance.1,2 These collaterals could be either portosystemic or portoportal. These collaterals enlarge and form varices. The usual location of varices is the gastroesophageal region.

Ectopic varices are formed by such collaterals at many different sites along the gastrointestinal (GI) tract other than the usual location.3 Various reported sites of ectopic varices are described in - Table 1.4

The understanding of gastric varices and their endovascular management is widely studied and documented; the same is not true for ectopic varices due to low incidence.

In this review article, we will try to discuss the applied anatomy, hemodynamics, and endovascular management of ectopic varices.

Incidence and Prevalence

As a small fraction of ectopic varices become symptomatic and are often underreported, the true incidence is undetermined.5 In a nationwide Japanese survey conducted by Watanabe et al, the incidence of ectopic varices at different location was duodenal (57%), small intestinal (6.4%), colonic (3.5%), rectal (44.5%), biliary (4.6%), anastomotic (5.8%), stomal (1.7%), and diaphragm (0.6%). The primary modality to diagnose the ectopic varices was endoscopy and a computed tomography (CT) scan was done whenever
Table 1 Sites of ectopic varices

<table>
<thead>
<tr>
<th>Luminal</th>
<th>Extraluminal</th>
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<tbody>
<tr>
<td>Isolated gastric varices</td>
<td>Intrapitoneal</td>
</tr>
<tr>
<td>Duodenum</td>
<td>Retroperitoneal</td>
</tr>
<tr>
<td>Jejunum</td>
<td>Umbilicus</td>
</tr>
<tr>
<td>Ileum</td>
<td>Around the falciform ligament</td>
</tr>
<tr>
<td>Colon</td>
<td>Gallbladder and biliary tree</td>
</tr>
<tr>
<td>Rectum and anal canal</td>
<td>Perisplenic</td>
</tr>
<tr>
<td>Peristomal</td>
<td>Right diaphragm</td>
</tr>
<tr>
<td></td>
<td>Ovary</td>
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<td></td>
<td>Vagina</td>
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</table>

required. About 5% of the variceal bleed is caused by ectopic varices.6

### Etiology

Portal hypertension secondary to liver cirrhosis is the most common underlying cause followed by the EHPVO, splenic vein thrombosis (secondary to pancreatitis/malignancy), or superior mesenteric vein (SMV) thrombosis. Some other underlying causes include pancreatic or gastric malignancy, liver metastasis, congenital hepatic fibrosis, and biliary stricture. In a patient with old abdominal surgery, ectopic varices can develop at the site of adhesions.3,4,6

Some congenital or familial conditions can cause colonic varices without portal hypertension.7–9

### Pathophysiology

Portosystemic collaterals are at higher resistance than the portal circulation. Development of portal hypertension leads to shunting of blood through these collaterals. These collaterals enlarge and form varices.

The location and development of ectopic varices depend on a few factors. Generalized portal hypertension does not cause all ectopic varices. Additional locoregional hemodynamic factors also contribute. These locoregional hemodynamic factors include (1) visceral vein stenosis or occlusion (splenic vein, mesenteric vein, or portal vein), (2) postinflammatory and postoperative adhesions and scarring, (3) postoperative altered anatomy, and (4) occlusion of portosystemic shunts, for example, post-balloon-occluded retrograde transvenous obliteration (BRTO). Based on this, the ectopic varices can be divided into the nonocclusive type and occlusive type.

Ectopic varices are “true veins” histologically, have thicker walls, bigger diameter, and higher wall tension. Because of this, ectopic varices have four times more risk of bleeding compared with esophageal varices.16 Bleeding is often arduous to control and can be fatal in ~40%.11 Ectopic varices bleed at low portosystemic gradients in more than half the cases.12 Ectopic portosystemic varices/collaterals not only lead to variceal bleed but may also manifest as hepatic encephalopathy.

### Applied Venous Anatomy

Knowledge of afferent and efferent of a portosystemic shunt is of paramount importance to plan endovascular management. – Fig. 1 shows important portosystemic collaterals. Before we get on with venous anatomy related to specific sites of ectopic varices, we must understand here that the afferent is formed by a tributary of the portal vein and efferent in many cases by the vein of Retzius.

Veins of Retzius are a group of veins located in the abdomen which joins the inferior vena cava (IVC) with the SMV or inferior mesenteric vein (IMV). The veins of Retzius pathways get their name depending on anastomosing vein such as “mesenteric-gonadal, mesenteric-caval, mesenteric-renal, or mesenteric-iliac.” The veins of Retzius are frequently undilated and may not be well appreciated.13

### Duodenal Varices

Duodenal varices are 1 to 3% of all varices.14 Compared with esophageal varices, duodenal varices are smaller, shorter, and deeper in location, hence their bleeding incidence is less. The paraduodenal varices connect with submucosal duodenal varices through perforators. Submucosal duodenal varices if visible on endoscopy have higher chances of bleeding as compared with if not visible.15,16

Afferents—There are four small pancreaticoduodenal veins, namely, posterior superior pancreaticoduodenal vein, anterior superior pancreaticoduodenal vein, posterior inferior pancreaticoduodenal vein, and anterior inferior pancreaticoduodenal vein. The superior and inferior veins can anastomose and form an arcade which is usually anterior but rarely can be posterior.

The posterior superior pancreaticoduodenal vein joins the portal vein. The anterior superior pancreaticoduodenal and the anterior inferior and posterior inferior pancreaticoduodenal veins join either the SMV directly or one of its tributaries, namely, gastrocolic trunk or the first jejunal trunk. This multiplicity of communications can make any vessel contributes to the collateral pathway13,17 (– Fig. 2).

Efferents—The efferents can be either via portosystemic collaterals or portoportal collaterals.

### Portosystemic Collaterals

In cirrhosis, efferents start at the second or third part of the duodenum and have hepatopetal flow through retroperitoneal veins of Retzius, namely, right renal vein, right gonadal vein, right adrenal vein, or right inferior phrenic vein into the IVC. Another pathway is the subcostal vein and ascending lumbar vein via the azygos pathway into the superior vena cava.18

### Portoportal Collaterals

In extrahepatic portal vein obstruction, efferent starts at the first part of the duodenum and flow hepatopetally through portoportal collaterals.19 Portoportal collaterals can be formed by existing venous plexuses around the common bile duct (CBD) or by portal vein remnant above the
obstructed portal system. Collaterals in EHPVO are again described in detail in biliary varices.

**Small Bowel Varices**

These are often seen in patients with portal hypertension, chronic SMV thrombosis, or with a history of abdominal surgery. Postoperative adhesions between the surface of the bowel and the abdominal wall, coupled with portal hypertension allow varices development. “A triad of portal hypertension, hematochezia without hematemesis, and prior abdominal surgery characterizes hemorrhage from small bowel varices.” Small bowel varices are not seen on routine endoscopy and may require capsule endoscopy.

**Afferents**—Jejunal and ileal branches of SMV.

**Efferents**—The efferents of small bowel varices usually drain into veins of Retzius. The most common pathway is an ileocolic vein draining through the right gonadal vein.
either into the right renal vein or IVC.22 Occasionally, the left gonadal vein may act as an efferent for IMV varices.23

Large Bowel Varices

Colonic Varices
Common locations are cecum and rectosigmoid and mostly seen in cirrhosis or portal vein obstruction patients. Less commonly, it can be seen in congestive cardiac failure, mesenteric or splenic vein obstruction, and mesenteric vein compression.7,24–26

Afferents—SMV branches, namely, the ileocolic, right colic, and middle colic veins for the right colon and IMV branches for the remaining part of the colon.

Efferents—Drains into the veins of Retzius, which include right gonadal, right renal, or lumbar veins.13

Rectal Varices
Rectal varices and internal hemorrhoids are different entities. Rectal varices characteristically are seen above the dentate line and yield to compression in contrast to hemorrhoids. The prevalence of hemorrhoids is not increased by the presence of portal hypertension.27 Rectal varices can manifest as mild or uncontrolled bleeding per rectum.

Afferents—The superior rectal vein is a direct continuation of the IMV. Superior rectal vein further divides into extrinsic rectal venous plexus and intrinsic rectal venous plexus and both are interconnected via perforators across the muscular propria. The superior part of intrinsic venous plexus is present in the rectal submucosa and the inferior part is present in the anal subcutaneous tissue.

Efferents—Rectal venous plexuses communicate via the rectogenital and interrectal communications and eventually drain via the middle rectal vein and inferior rectal veins into the internal iliac vein.28

Umbilical Varices
A bunch of vessels in the subcutaneous tissue of the anterior abdominal wall near the umbilicus can lead to the formation of varices. Sometimes, these can connect to varicose veins in the anterior right thigh.29

Afferents—The left branch of the portal vein via the recanalized umbilical and paraumbilical veins.

Efferents—Umbilical varices drain via superior and inferior epigastric veins into external iliac veins with the latter being the commonest path. Sometimes, paraumbilical veins can also drain via internal thoracic veins into the superior vena cava.13

Biliary Varices
In extrahepatic portal vein obstruction, multiple collateral veins can develop at the porta hepatitis from two existing venous plexuses located around the extrahepatic part of the CBD. These venous plexuses are known as the paracholedochoval venous plexus of Petren and the epicocholedochoval venous plexus of Saint. The paracholedochoval venous plexus of Petren courses side by side and the epicocholedochoval venous plexus of Saint courses on the surface of the CBD as a plexiform network.30,31

Enlargement of these collaterals may cause portal biliopathy. Enlarged paracocholechoval collaterals cause extrinsic compression of the CBD and enlarged epicocholechoval collaterals cause irregularity of the mural surface of the CBD.32,33 Sometimes, biliary varices can cause hemobilia or life-threatening hemorrhage.34

Stomal Varices
Sometimes, varices can develop in the stomal mucosa especially at ileostomies following protocolectomy for inflammatory bowel disease.35 The apposition of bowel and abdominal wall brings portal and systemic circulation together allowing for portosystemic collateralization. Bleeding is due to trauma or variceal erosion.

Often stomal varices are not seen on endoscopy; therefore, high clinical suspicion along with CT imaging is necessary in obscure cases. The morbidity is often high, but mortality is usually low around 3 to 4%. Simple maneuvers such as compression bandaging with or without adrenaline, packing with gelatine foam, and suture ligation often effectively stop bleeding.36

Miscellaneous Infrequent Sites
Vesical varices can develop following abdominal surgery and may present as hematuria or rarely life-threatening intraperitoneal bleed. Intraperitoneal and cutaneous varices can cause an intraperitoneal or percutaneous bleed.5

Classification
Ectopic varices are divided based on the patency of the portal system: Type a “nonocclusive or oncotic” (normal portal system or global portal hypertension) and Type b “occlusive” (occluded portal system). Again subdivided into Type 1 having only portoportal collaterals, Type 2 having primarily portoportal collaterals with few portosystemic collaterals, and Type 3 having primarily portosystemic collaterals with few portoportal collaterals as per Saad—Caldwell’s classification37,38 (Fig. 3).

Diagnosis and Site Localization
Endoscopy and Clinical Examination
Inspection and palpation can help diagnose stomal or para-stomal varices; otherwise, endoscopy is the primary modality to diagnose and localize the ectopic varices. On clinical examination or endoscopy, high-risk varices typically appear large, distended, or have features of the previous bleed such as “red and white spot.”39 Some distal duodenal, jejunoileal, and colonic varices may not be seen on endoscopy. A high index of suspicion is required to diagnose ectopic varices in the case of brisk upper or lower GI bleed, especially if upper and lower GI endoscopy fails to identify the source of bleed in a patient with portal hypertension.
Sonography
Doppler ultrasound effectively diagnoses patency of the entire portal venous system and direction of mesoporal blood flow which can be toward the liver, away from the liver, or fluctuating. Ultrasound has the advantage of repeatability and can be performed bedside in unstable patients.

Both diagnosis and image-guided treatment of stomal varices can be done with the help of Doppler ultrasound.40

Contrast-Enhanced CT Scan
A contrast-enhanced CT scan is the modality of choice. It helps in the diagnosis, site localization, and treatment planning of variceal hemorrhage. The 64-slice and higher slice CT scanner can quickly image even actively bleeding or hemodynamically unstable patients. Multiphasic CT allows the detection of blood loss as little as 0.5 mL/s. The raw data postprocessing and multiplaner reconstruction provide accurate anatomical detail for interventional, endoscopic, or surgical planning.

CT is effective in the diagnosis of underlying etiology such as liver cirrhosis, secondary signs of portal hypertension, or locoregional surgical causes. CT can detect extraluminal varices, particularly those located at the unusual site such as postoperative adhesions, parastomal, surgical anastomoses, or mesentery.

Magnetic Resonance Imaging
T2-weighted or postcontrast T1-weighted magnetic resonance imaging (MRI) can be used. MRI can be helpful in patients with hepatorenal syndromes where a contrast-enhanced CT cannot be done.

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Fig. 3  Hemodynamic classification of ectopic varices. PC, portal venous circulation; P-O, portal venous outflow; P-P, portoportal collaterals; P-S: portosystemic collaterals; PvB, portal vein branch; SC, systemic venous circulation; S-O: systemic venous outflow. (Reproduced with permission from Philips et al.37)
Management

There is a lack of randomized trials or guidelines for ectopic varices management. The management depends on the location of hemorrhage, the severity of hemorrhage, institutional expertise, and the etiology of portal hypertension. A collaborative team approach of intensivists, endoscopists, vascular interventional radiologists, and GI surgeons is desirable.

Medical, endoscopic, and surgical management are beyond the scope of this article. Here, we will elaborate on endovascular interventional radiological management.

Endovascular Interventional Radiological Procedures

Multiple image-guided approaches have been extensively reported. A very practical algorithm is mentioned by Saad et al based on etiology and patency of mesoportal circulation to guide the treatment\(^ \text{38} \) (Fig. 4).

Decompression by Transjugular Intrahepatic Portosystemic Shunt

In cirrhosis with patent portal system, transjugular intrahepatic portosystemic shunt (TIPS) has a role in the management of bleeding ectopic varices.\(^ \text{41} \) Here, it is important to note that adequate ectopic varix decompression is not achieved at the usual 12 mm Hg gradient reduction because ectopic varices decompress through many different pathways. TIPS creation should be combined with embolization or sclerotherapy to prevent rebleed\(^ \text{42} \) (Fig. 5).

Whether to use the TIPS approach or not depends on the experience of the interventional radiologist and the severity of the underlying liver disease, in terms of Model for End-Stage Liver Disease score or encephalopathy. Contraindications for creating TIPS are detailed in Table 2.\(^ \text{43} \)

### Table 2 Absolute and relative contraindications of creating TIPS

<table>
<thead>
<tr>
<th>Absolute</th>
<th>Relative</th>
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<tbody>
<tr>
<td>Primary prevention of variceal bleeding</td>
<td>Hepatoma, particularly if central</td>
</tr>
<tr>
<td>Severe congestive heart failure</td>
<td>Obstruction of all hepatic veins</td>
</tr>
<tr>
<td>Tricuspid regurgitation</td>
<td>Hepatic encephalopathy</td>
</tr>
<tr>
<td>Multiple hepatic cysts</td>
<td>Significant portal vein thrombosis</td>
</tr>
<tr>
<td>Uncontrolled systemic infection or sepsis</td>
<td>Severe uncorrectable coagulopathy (INR &gt; 5)</td>
</tr>
<tr>
<td>Unrelieved biliary obstruction</td>
<td>Thrombocytopenia (≤ 20,000 platelets/mm(^3))</td>
</tr>
<tr>
<td>Severe pulmonary hypertension</td>
<td>Moderate pulmonary hypertension</td>
</tr>
</tbody>
</table>

Abbreviations: INR, international normalized ratio; TIPS, transjugular intrahepatic portosystemic shunt.
Even if decompression is contraindicated, a TIPS approach can still provide direct access to varices for sclerotherapy. One may avoid putting a stent graft, can reduce the shunt, or reverse if the need arises.\(^5\)

**Decompression by Portal or Mesenteric Vein Recanalization**

If there is occlusion of portomesenteric circulation, then recanalization by venoplasty or stenting is a very effective decompressive strategy. Such decompression provides anatomical continuity.

The optimum decompression depends on the extent of occlusion. If limited to only the extrahepatic portal vein, then a portal vein recanalization will suffice. In case of occlusion of the confluence of the splenic and mesenteric veins, recanalization or stent placement should be done from the SMV to the portal vein and not from the splenic vein. Additional sclerotherapy of varices can be done if there is a filling of varices after decompression (►Fig. 6). Often decompression alone is enough for regression of varices (►Fig. 7). In the case of splenic vein occlusion, partial splenic artery embolization can be performed also in such cases to decompress left-sided portal hypertension.

If intrahepatic portal vein branches are also occluded, then recanlized portal vein stent should be extended by TIPS to provide outflow to the portal vein to achieve an effective reduction in portal pressure.\(^38\)

**Balloon-Occluded Retrograde Transvenous Obliteration**

When decompression is not feasible or contraindicated, shunt and variceal occlusion procedures such as BRTO can be performed accessing the shunt retrogradely through the systemic venous approach. BRTO is routinely used for the treatment of gastric variceal hemorrhage with reported success rates around 89 to 100%.\(^44,45\)

The only contraindications to BRTO are relative (soft) contraindications. These include (1) severe uncorrected coagulopathy (which probably in this clinical setting is associated with liver failure), (2) splenic vein thrombosis (segmental portal hypertension), (3) portal vein thrombosis, and (4) uncontrolled esophageal variceal bleeding.\(^46\)

There is often an improvement in hepatic function or hepatic encephalopathy because of increased portal flow to the liver. However, the resultant increase in portal pressure can cause bleeding from unsclerosed varices, the occurrence of new varices at remote sites, development of ascites, or worsening of esophageal varices.\(^47\) To minimize these problems, various additional procedures such as TIPS,\(^48\) percutaneous obliteration, and endoscopic interventions can be utilized.\(^49\)

BRTO can be helpful in the management of duodenal, jejunileal, colonic, stomal, and mesenteric ectopic varices.\(^50–54\)

A mixture of air, sodium tetradecyl sulfate, and lipiodol in a 3:2:1 ratio can be used. We can also add gel foam slurry to embolize competing collaterals, accelerate thrombosis, and
reduce sclerosant dose. We have used BRTO for ileal varices (► Fig. 8) and umbilical varices (► Fig. 9). In our experience, BRTO is a safe and effective lifesaving procedure for ectopic varices if a suitable shunt can be found for balloon navigation via a transfemoral or transjugular approach. Sometimes instead of the balloon, a modification of technique using coils called coil-assisted retrograde transvenous obliteration can be used if shunt tortuosity makes it difficult to take an occlusive balloon35 (► Fig. 10).

**Balloon-Occluded Antegrade Transvenous Obliteration and Direct Percutaneous Embolization**

Percutaneous embolization via transhepatic route is another procedure to occlude ectopic varices with reported success...
rates around 80% but up to 65% rebleeding rate within the next 5 months.\textsuperscript{56}

The transhepatic route provides easy access to the portal system especially if the operator is not experienced for TIPS, but it has risks of tract bleeding, recurrent bleeding because of the inability of portal decompression, and is contraindicated in the presence of ascites. Sometimes, antegrade obliteration may be required using a transsplenic/transhepatic route in cases of EHPVO depending on the location and anatomy of the varices to be sclerosed.

In the case of multiple collaterals, coiling of adjacent extraluminal collaterals can be done to help create blood flow stagnation which in turn helps in increasing sclerosant contact time and prevents accidental seepage of sclerosant.
It is important to avoid putting coils in the submucosal varices as it can ulcerate in the lumen. The afferent and efferent of stomal or parastomal varices can both be seen by ultrasound. Afferent can be accessed antegrade under ultrasound guidance using a micropuncture set. Sclerotherapy or coil embolization by direct ultrasound-guided puncture of cutaneous or stomal varices is described in the literature anecdotally. In the case of portal vein thrombosis where percutaneous transhepatic access to the portal side (►Fig. 11) is difficult, sclerotherapy or coil embolization can be performed via direct ultrasound-guided puncture of cutaneous or stomal varices.
portal vein is not possible, via small pararectal incision ileocolic vein can be used to access SMV or IMV. This is mainly used for sclerosis of rectal varices.

**Conclusion**

Early diagnosis of ectopic varices needs strong clinical suspicion and contrast-enhanced CT scan as endoscopy may often fail to pinpoint a source. Understanding the applied anatomy and hemodynamic classification is important to decide the most suitable therapy. Depending on the patency status of splanchnic circulation, interventional radiological procedures in the form of either decompressing the varices or obliterating them work effectively.

Confict of Interest
None declared.

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