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REVIEW ARTICLE

Updates in the pathogenesis, diagnosis and management of ectopic varices

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Abstract Ectopic varices (EcV) comprise large portosystemic venous collaterals located anywhere other than the gastro-oesophageal region. No large series or randomized-controlled trials address this subject, and therefore its management is based on available expertise and facilities, and may require a multidisciplinary team approach. EcV are common findings during endoscopy in portal hypertensive patients and their bleeding accounts for only 1–5% of all variceal bleeding. EcV develop secondary to portal hypertension (PHT), surgical procedures, anomalies in venous outflow, or abdominal vascular thrombosis and may be familial in origin. Bleeding EcV may present with anaemia, shock, haematemesis, melaena or haematochezia and should be considered in patients with PHT and gastrointestinal bleeding or anaemia of obscure origin. EcV may be discovered during panendoscopy, enteroscopy, endoscopic ultrasound, wireless capsule endoscopy, diagnostic angiography, multislice helical computed tomography, magnetic resonance angiography, colour Doppler-flow imaging, laparotomy, laparoscopy and occasionally during autopsy. Patients with suspected EcV bleeding need immediate assessment, resuscitation, haemodynamic stabilization and referral to specialist centres. Management of EcV involves medical, endoscopic, interventional radiological and surgical modalities depending on patients' condition, site of varices, available expertise and patients' subsequent management plan.

Abbreviations

APC	Argon plasma coagulation
B-RTO	Balloon-occluded retrograde transvenous obliteration
CT	Computed tomography
EcV	Ectopic varices
EUS	Endoscopic ultrasound
EPVO	Extrahepatic portal venous obstruction
G-O	Gastro-oesophageal
GV	Gastric varices
IGV	Isolated gastric varices
OV	Oesophageal varices
OG	Oesophago-gastric
PHT	Portal hypertension
RCT	Randomized-controlled trials
TIPS	Transjugular intrahepatic portosystemic shunt

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Introduction

The term "ectopic varices" (EcV) describes dilated portosystemic collateral veins located in unusual sites other than the gastro-oesophageal (G-O) region including the ectopic isolated gastric varices type 2 (IGV2). This definition has previously been given to any gastrointestinal mucosa-associated abnormally dilated tortuous veins that may lead to gastrointestinal bleeding. In addition, the term

has also been given to any portosystemic collateral veins located in the abdominal wall or in the retroperitoneal space.

As no randomized-controlled trials (RCTs) have previously addressed the therapeutic modalities of EcV, most of the available knowledge about this entity is obtained from small case series, case reports and mini-reviews. Awareness of the existence and the therapeutic options of EcV is essential for any physician dealing with patients with gastrointestinal bleeding. This is crucial not only because EcV represent up to 5% of all variceal bleeding episodes [1] but also because of the difficulty in their management and the high mortality secondary to their initial bleeding (up to 40%) [2].

Nowadays, EcV are diagnosed more frequently because of the recent advances in radiological and endoscopic techniques such as double-balloon enteroscopy (DBE) and video capsule endoscopy (PillCam). Indeed, about 8.1% of patients with portal hypertension (PHT) who underwent capsule endoscopy have small-bowel varices [3]. In this review, we present the currently available knowledge relating to the sites, types, pathogenesis, diagnosis and management of EcV.

Sites

Although EcV occur at several sites (Table 1), the bleeding EcV are most commonly found in the duodenum (Fig. 1), jejunum, ileum, colon, ano-rectal region (Fig. 2), biliary tract, umbilicus, peritoneum or at the sites of previous bowel surgery including stomas (peristomal varices) and trans-anastomotic porto-portal varices. Other infrequent sites include the peritoneum, ovary and vagina.

Norton et al. [4] reviewed 169 cases of bleeding from EcV. Twenty-six of them were peristomal in origin, 17% in

Table 1 Recognized sites of EcV

Duodenum
Jejunum
Ileum
Colon
Rectum
Peristomal area
Biliary tree
Peritoneum
Around the falciform ligament
Umbilicus
Urinary bladder
Along the splenic ligament
Ovary
Vagina
Right diaphragm

the duodenum, 17% in the jejunum or ileum, 14% in the colon, 8% in the rectum, 9% in the peritoneum, and few were located in other rare sites such as the vagina and the ovary [4]. Another rare site of EcV include the right diaphragm. On rupture, these varices present with acute dyspnoea and right-sided bloody pleural effusion [5]. Trans-anastomotic porto-portal varices are rare. They usually develop in the presence of extrahepatic PHT and presumably arise within the peri-anastomotic inflammatory tissue. Such varices may be difficult to manage and their prognosis is poor if they are bleeding [6].

Prevalence

EcV account for only 1–5% of all variceal bleeding episodes [1, 2, 7]. EcV represent a relatively common finding during endoscopy in patients with PHT. The prevalence of EcV varies according to the modality used for their detection, the patient population studied and the aetiology of PHT. For example, duodenal varices were detected in 40% of patients with intrahepatic PHT undergoing angiography [7], whereas ano-rectal varices have been reported in 10–40% of patients with liver cirrhosis undergoing colonoscopy [8, 9]. Colonic varices were reported in 3.4% of patients with intrahepatic PHT [10]. In addition, a recent study of 37 patients with liver cirrhosis who underwent capsule endoscopy, 3 (8.1%) were found to have small-bowel varices [3].

On upper endoscopy, most patients with duodenal varices have extrahepatic PHT. In fact, most patients with portal or splenic vein thrombosis on angiography are likely to show duodenal varices [4, 11–14]. Intestinal varices have also been reported in a patient with prehepatic PHT secondary to cavernous malformation of the portal vein (portal cavernoma) and were associated with oesophageal varices (OV) [15]. Varices in other parts of the small bowel (jejunum and ileum) and in the colon are commonly seen in patients with intrahepatic PHT who have previously undergone abdominal surgery [11].

Stomal varices, particularly common in patients with intrahepatic PHT, are caused by primary sclerosing cholangitis [11]. Because of the low prevalence of extrahepatic PHT in the western countries, most cases of bleeding from EcV reported in the West are usually associated with intrahepatic PHT [11, 16]. These varices usually bleed at hepatic venous pressure gradients of 12 mmHg or less [17]. Similar to gastric varices (GV), a possible explanation for the lower portal pressure in patients with EcV could be the development of spontaneous spleno-renal shunts or a reduced portal collateral resistance [18]. The bleeding can be massive enough to cause mortality, and the source in such cases can only be identified at autopsy [19].

Fig. 1 (**a**) Upper endoscopy picture showing serpiginous varices in the postbulbar duodenum with a focus on intermittent bleeding. (**b**) Close-up view
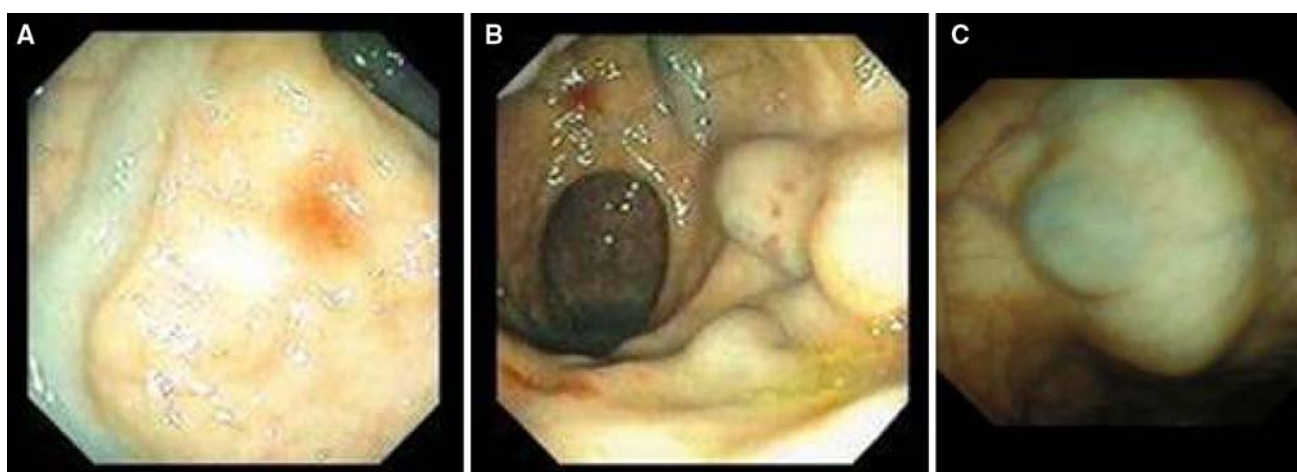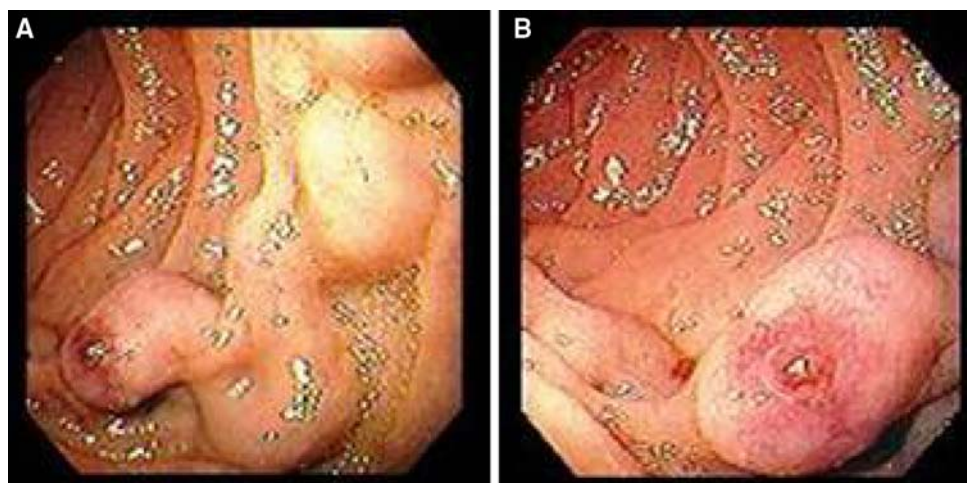

Fig. 2 Sigmoidoscopy (**a**, **b**) and colonoscopy (**c**) pictures showing dilated venous collaterals and spider angiomas in a patient with liver cirrhosis and PHT

Table 2 shows the estimated prevalence of EcV in different studies. To the best of our knowledge, data regarding the incidence and/or severity of bleeding in each site are not currently available. A proposed international Web-based EcV registry would be very valuable in this respect.

Aetiology and pathogenesis

As shown in Table 3, EcV may develop secondary to PHT, including the idiopathic PHT [20], surgical procedures involving abdominal organs and vessels, anomalies in the venous outflow vessels, abdominal vascular thrombosis, hepatocellular carcinoma, and may be familial in origin [21]. It has also been reported that duodenal and rectal varices develop as a result of band ligation of varices in the oesophago-gastric (OG) junction [22, 23].

Most of EcV represent natural portosystemic shunts that develop as a result of high portal pressure anywhere in the gut, around the ovaries, the biliary tree and the bare area of the liver. Under normal circumstances, such collaterals remain collapsed. However, in portal hypertensive patients, the portosystemic collaterals open in a trial to reduce the increased intrahepatic vascular resistance.

Any surgical operation involving apposition of abdominal structures (drained by the systemic veins) to the bowel (drained by the portal tributaries) may result in the formation of collateral vessels at an unusual site. An example is the development of ileostomy varices (stomal varices) in patients with primary sclerosing cholangitis who have undergone colectomy for ulcerative colitis [24]. In addition, bleeding jejunal varices were detected in a case of severe extrahepatic portal vein stenosis secondary to pancreaticoduodenectomy with portal vein resection and intraoperative radiotherapy [25].

Colonic varices may develop in the absence of PHT because of the anomalies in portal venous outflow as in patients with congenital anomalies of portosystemic

Table 2 Estimated prevalence of EcV in different studies

References	Author(s)	Diagnostic modality	Patient population	n	n (%) EcV	Sites
[1]	Kinkhabwala et al.	Transhepatic portography	Predominantly IHPHT	500	25 (5)	Ileal
[3]	De Palma et al.	Capsule endoscopy	IIHPHT	37	3 (8.1)	Small bowel
[7]	Stephan and Miething	Arteriography	IHPHT	73	1 (1.4)	Duodenum
[7]	Stephan and Miething	Arteriography	EHPHT	33	9 (27.3)	Duodenum
[10]	McCormack et al.	Colonoscopy	Predominantly IHPHT	102	4 (3.6)	Rectal
[12]	Salam et al.	Arteriography	GI bleeders	200	6 (3)	GIT and GB
[13]	Wilson et al.	Arteriography	GI bleeding	309	5 (1.6)	GIT
[14]	Itzchak and Glickman	Arteriography	EHPHT	20	8 (40)	Duodenum
[17]	Tripathi et al.	Portography	TIPS patients	472	12 (2.5)	GIT
[131]	Sarin et al.	Endoscopy	IHPHT and EHPHT	1,128	53 (4.6)	IGV2

EHPHT, extrahepatic portal hypertension; GI, gastrointestinal tract other than the O-G area; IHPHT, intrahepatic portal hypertension

Table 3 Recognized causes of EcV

PHT (intrahepatic and extrahepatic)

Surgical procedures involving abdominal organs and vessels

Anomalies in the venous outflow vessels

Abdominal vascular thromboses

Hepatocellular carcinoma

Secondary to band ligation of O-G junction varices

Familial

Table 4 Recognized presentations of bleeding from EcV

Overt gastrointestinal bleeding of obscure origin

Occult gastrointestinal bleeding

Accidental finding

Iron-deficiency anaemia

Haematemesis

Haematochezia

Internal haemorrhage (haemoperitoneum)

Hypovolaemic shock

Haemorrhagic pleural effusion

At autopsy

anastomoses [26], abnormal vessel structure [27], arterio-venous fistulae [28] or they may be familial in origin [21, 27]. In addition, rectal varices may occur secondary to intra-abdominal vascular thrombosis [29].

Because tension in the varix wall is proportional to the radius of the vessel and the transmural pressure across the vessel wall, the major determinants of rupture of EcV are mostly vessel size, portal pressure and varix wall tension [30]. In addition, the prevalence of haemorrhage from rectal varices was shown to be significantly higher in patients with rectal varices of advanced form and/or with a positive "red-colour" sign [31].

Diagnosis

Table 4 provides a list of the recognized presentations of patients with bleeding EcV. Fortunately, "luminal EcV", that is, those occurring within the gastrointestinal tract lumen, are more common, usually easier to detect and manifest earlier than the "non-luminal EcV", that is, those elsewhere in the abdominal cavity or the pelvis.

Patients with duodenal varices may present with hae-matemesis, or massive lower gastrointestinal bleeding [32]. In addition, EcV located distal to the duodenum present mostly with melaena or haematochezia [33]. Moreover, the presence of EcV should be considered in all patients with PHT and gastrointestinal bleeding if both upper and lower gastrointestinal endoscopies failed to show a definite source of bleeding.

Many other cases are discovered accidentally, especially those presenting with anaemia of obscure origin, melaena or positive faecal occult blood testing. EcV can also be detected during routine endoscopy, diagnostic transfemoral or transhepatic angiography [23], Technetium TC-99m red blood cell scintigraphy [34], video capsule endoscopy [3], computed tomography (CT) angiography [35], multislice helical CT [36], CT-enteroclysis [37], contrast-enhanced 3D magnetic resonance angiography [38], endoscopic ultrasound (EUS) [39], laparoscopy or laparotomy (Table 5). In addition, colour Doppler-flow imaging has been used to demonstrate gallbladder varices in three cases of extrahepatic portal venous obstruction (EPVO) and obstructive jaundice [40]. Occasionally, the presence of EcV can be confirmed only at autopsy [41]. It should be noted that EcV can appear as filling defects in barium studies of the colon or small bowel and may be misdiag-nosed as polyps or tumours [21]. This emphasizes the importance of endoscopic and angiographic modalities.

Table 5 Methods of detection of EcV

Oesophagogastroduodenoscopy
Push and DBE
Wireless video capsule endoscopy
Colonoscopy
EUS
Colour Doppler-flow imaging
Multislice helical CT
CT-enteroclysis
Contrast-enhanced 3D magnetic resonance angiography
TC-99m red blood cell scintigraphy
Multidimensional-CT
Angiography and CT-angiography
During laparotomy or laparoscopy
At autopsy

Varices in the abdominal wall usually rupture externally and can easily be diagnosed, whereas those located around the falciform ligament of the liver, in the diaphragm, in the rectovesical region, or on the splenic ligament may rupture into the peritoneal cavity causing internal bleeding and can be fatal [42, 43]. Diagnosis of cases with internal bleeding needs a high index of clinical suspicion and often presents with rapid accumulation of ascites associated with signs of hypovolaemic shock and a reduction in the haematocrit value. The diagnosis is further indicated by CT scan, which detects retroperitoneal varices and intra-abdominal bleeding or collections, and can be confirmed by the detection of a heavily bloodstained ascitic fluid tap.

Various techniques can be used to reveal the extent of portosystemic collaterals, the direction of flow and the pressure in the portal venous system. These include splenic portography, umbilical vein catheterization, percutaneous transhepatic portography and transjugular transhepatic portography at the time of or after transjugular intrahepatic portosystemic shunt (TIPS) insertion [44–46]. The last three modalities allow direct and selective access to the main tributaries of the portal vein, including the coronary-azygous collaterals, which represent a dominant mechanism in patients with PHT [47].

Role of capsule endoscopy (PillCam)

About 8.1% of patients with PHT who have undergone capsule endoscopy have small-bowel varices [3]. This number reflects a prevalence that is generally higher than expected and points to the role of capsule endoscopy as a non-invasive tool in the workup of patients with obscure gastrointestinal bleeding, including those with chronic liver diseases. Also, a case report by Fix et al. [48] has shown the significance of PillCam in diagnosing obscure gastrointestinal bleeding from mesenteric varices. It has also been found to be beneficial in detecting small-bowel varices in a patient after Whipple's operation [49]. Furthermore, PillCam as an alternative to upper endoscopy has been tested in 32 patients with liver cirrhosis for the detection of OG varices and portal hypertensive gastropathy [50]. In one patient, PillCam detected small varices that were not seen at endoscopy, and the overall concordance between both modalities was 96.9 and 90.6% for the diagnosis of varices and gastropathy, respectively, without any adverse effects related to PillCam [50]. A large-scale trial is underway to validate and expand these findings.

Role of DBE

At ileocolonoscopy, 18% of patients with liver cirrhosis and PHT have ileal varices [51]. DBE has the potential to visualize the whole small bowel, take biopsy specimens, and perform all necessary endoscopic interventions [52]. Hekmat et al. [53] have recently demonstrated a successful obliteration of a jejunal varix by using N-butyl-2-cyanoacrylate (Histoacryl) in a lesion found ∼240 cm from the ligament of Treitz. This signifies the therapeutic benefit of DBE over capsule endoscopy in small-bowel EcV. The widespread use of this modality in the future would help detect such varices and allow therapeutic intervention, and hence reduce rebleeding, transfusions, and may be mortality.

Management of bleeding EcV

The management of EcV is difficult and may require a multidisciplinary team of endoscopists, hepatologists, surgeons and interventional radiologists (Table 6). It is very difficult to draw guidelines for the treatment of EcV because of the diversity of their location, presentation, complications and diagnostic and therapeutic options. However, the optimal therapeutic modality depends mainly on the location of varices, patient's condition, the locally available expertise and facilities, and the cause of PHT. Management essentially includes prompt resuscitation, immediate workup to localize the site/source of bleeding, followed by application of the suitable treatment modality or immediate transfer to tertiary referral centres. A proposed systematic approach for any patient presenting with bleeding from possible EcV is shown in Fig. 3.

Table 6 Therapeutic options for EcV

Endoscopic
  Band ligation
  Injection sclerotherapy
  Argon plasma coagulation
Medical: β-blockers
Interventional radiology
  Embolization
  TIPS ± embolization
  B-RTO ± TIO
Surgical

B-RTO, balloon-occluded retrograde transvenous obliteration; TIO, transiliocolic obliteration; TIPS; transjugular intrahepatic portosystemic stent shunt
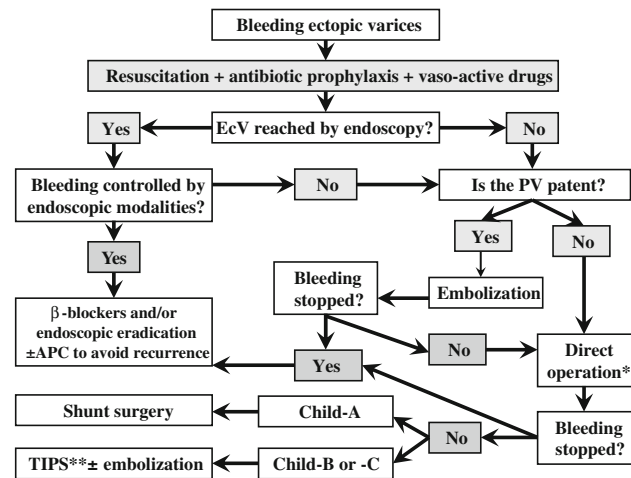

Fig. 3 Schematic of the management of bleeding EcV. APC, argon plasma coagulation; EcV, ectopic varices; PV, portal vein; TIPS, transjugular intrahepatic portosystemic shunt. * A direct operation or local devascularization of the EcV is a useful procedure even if portal vein is not patent or the patients have Child-Pugh B or C cirrhosis. ** Use TIPS with caution in patients with Child-Pugh C cirrhosis and weigh the benefit of stopping bleeding against the risk of encephalopathy and deterioration in liver function

Initial management

Similar to acute OG variceal bleeding, clinical assessment, resuscitation, haemodynamic stabilization, antibiotic prophylaxis and referral to specialist centres should be started as soon as possible in patients with suspected EcV bleeding.

The use of vasoactive drugs such as octreotide and terlipressin to reduce splanchnic blood flow and variceal pressure may be of benefit as in patients with bleeding from OG varices [54, 55]. The role of these vasoactive drugs in the control of bleeding from EcV has not been addressed.

All subjects with suspected variceal bleeding should undergo emergency upper endoscopy as a first-line investigation. If it fails to show the source of upper gastrointestinal bleeding, colonoscopy after a rapid colonic purge (3 litres of polyethylene glycol solution delivered via a nasogastric tube) should be the second step of investigation because in a series of 22% of French patients with

EcV, bleeding was reported from the colon or rectum [8]. Colonic and rectal varices appear as serpiginous, dilated bluish vessels projecting into the lumen. However, if panendoscopy fails to identify and localize a variceal or a non-variceal source of bleeding, capsule endoscopy, DBE, transfemoral angiography or TC-99m red blood cell scintigraphy or other modalities, as mentioned above, would be the third step of the investigation.

Endoscopic treatment

The use of endoscopic modalities has been reported both in the setting of controlling acute bleeding from EcV and in the secondary prophylaxis. Therefore, depending on the currently available data, primary prophylaxis of EcV bleeding should not be recommended.

Band ligation

Only a few reports are available regarding the successful use of band ligation of EcV mainly in the rectum and duodenum [56–60]. It has also been reported that band ligation, together with balloon-occluded retrograde transvenous obliteration (B-RTO), is beneficial in the management of bleeding duodenal varices [59]. However, if the entire varix cannot be banded, there might be a high risk of developing a wide defect in the varix, especially after sloughing, rendering the banding technique unsafe for large EcV. It has been recommended that banding can be done only if the varix diameter does not exceed the diameter of the endoscope [4].

Injection sclerotherapy

Most EcV are within reach of standard endoscopy [11] or DBE [54]. Several studies have reported successful treatment of duodenal and rectal varices with sclerosant injection [61, 62]. The response to injection sclerotherapy can be unsatisfactory, especially with rectal and colonic varices, probably due to excessive dilution of the sclerosant in large varices beyond a concentration adequate to obliterate the varix. Therefore, a combination of sclerotherapy with banding in such a situation can be attempted [63]. Also, successful control of bleeding from duodenal, jejunal, colonic and rectal varices after injecting with cyanoacrylate, thrombin or any other combination of sclerosants has also been reported [53, 64–67].

Argon plasma coagulation

The use of argon plasma coagulation (APC) as a supplemental treatment in the eradication and prevention of

recurring oesophageal variceal after band ligation has been shown to be both effective and safe, and better than band ligation alone [68–70]. APC of EcV has been reported to be effective in stopping bleeding and eradicating ileocolonic anastomotic varices in a 65-year-old male patient [71]. The use of APC in preventing recurrence of endoscopically accessible EcV should be tested in more patients after initial eradication with other modalities such as band ligation or sclerotherapy.

Embolization therapy

Embolization, via intervention radiological techniques, of steel coils, thrombin, gel foam, collagen or autologous blood clot [72], either alone or in combination with band ligation or TIPS, is an effective short-term therapy for bleeding OG varices. The goal of this therapy is not to occlude the bleeding site itself but to occlude the feeding vein (i.e., the vein on the portal venous side) to the EcV. Steel coils are the preferred embolic materials because they lead to a permanent focal occlusion and are available in a variety of sizes, and allow occlusion of large veins without difficulty. The rate of bleeding control has been reported to reach 94% [73, 74]. Unfortunately, embolization does not decompress the portal venous system, resulting in high 1-year rebleeding rates [73]. The feeding veins can be reached via percutaneous transhepatic or transjugular routes.

Percutaneous embolization of bleeding EcV has been reported in the literature as case reports or case series [75–78]. The transhepatic approach is usually preferred, especially in patients with active bleeding, because access to the portal venous system is faster than the transjugular approach. On the other hand, the transhepatic approach may be difficult in patients with portal vein occlusion, but is usually preferred if a subsequent TIPS is planned after variceal embolization [79–81]. Macedo et al. presented a retrospective review of 14 patients, 12 of whom had an abdominal surgery 2–38 years earlier, who underwent transhepatic embolization of coils into the draining veins for the management of bleeding EcV. Rebleeding occurred within 72 h in two patients, and recurrent bleeding from 23 days to 27 months after initial intervention was identified in nine patients. Therefore, it can be concluded that percutaneous coil embolization is a simple and safe treatment for bleeding EcV; however, recurrent bleeding is frequent and repeated intervention is often required. TIPS usually offers good control of bleeding at the expense of a more complex procedure and increased risk of encephalopathy [72]. Okasaki et al. [82] have recently reported a case of obliteration of bleeding giant rectal varices with the use of modified percutaneous transhepatic obliteration with sclerosant.

Medical treatment

The role of β-blockers in the primary and secondary prophylaxis of bleeding from OG varices is well established. However, no solid data exist regarding the use of β-blockers or nitrates in the long-term management of patients with EcV. Although an earlier study in 1986 had shown that β-blocker therapy was not effective in the management of peristomal varices [22], a recent report of three cases had shown their effectiveness [83]. Another case report suggests that β-blockers can be effective as secondary prophylaxis of duodenal variceal rebleeding, especially after simple oversewing of duodenal variceal veins [84]. It seems logical that this modality could be tried in patients with EcV, especially after the implementation of emergency endoscopic or surgical modalities or when patients cannot undergo a surgical shunt [85].

Surgical treatment

If endoscopic modalities or interventional embolization fail to control bleeding from EcV, creating a TIPS or proceeding with surgery is the optimal option depending on the availability of expertise, liver function and the cause of PHT. Direct surgery or local devascularization of the EcV is a useful minimally invasive procedure that does not take much time, does not involve resection of long segments of the small intestine and can be done even if the portal vein is not patent or if the patient has Child B or C cirrhosis [84, 86].

Surgery is preferred both in patients with Child-Pugh class A cirrhosis and in patients with an EPVO. Other surgical interventions reported to control EcV include simple oversewing of the duodenal varices through a duodenotomy [87], duodenal dearterialization and stapling [79], circumferential-stapled anoplasty [88] and double selective shunting [89].

The non-selective portosystemic shunts, such as mesocaval, portocaval or central splenorenal shunt, adequately decompress the portal circulation, but because they are considered major surgical procedures, they are not commonly used nowadays. Also, non-conventional portosystemic shunts using large collateral vessels may be performed in patients with EPVO, for example, children with extensive thrombosis of the portal venous system [90].

Transjugular intrahepatic portosystemic shunt

Many recent publications have addressed the role of TIPS in the successful control of bleeding EcV caused by intrahepatic PHT unresponsive to conservative or endoscopic management [17, 40, 91–102]. The largest four

series involved 12, 9, 24 and 21 patients who received TIPS for bleeding EcV [40, 100, 101, 103]. Although TIPS offers a highly effective modality for controlling bleed, the long-term survival of patients is mainly dependent on their liver function. The high efficacy of TIPS has to be balanced against the potential for increased encephalopathy and the procedure-related morbidity.

TIPS together with variceal embolization has the advantage of being effective, minimally invasive, can be performed in one session, does not preclude subsequent liver transplantation, and therefore may be used during the acute situation both as a bridge to transplantation and as the definitive therapy in patients unfit for surgery.

Balloon-occluded retrograde transvenous obliteration

B-RTO is a recent therapeutic technique for the management of patients with large GV. The technique is valuable in patients who bleed at lower portal pressures, patients with hepatic encephalopathy, and when the portal vein is not patent. Recently, B-RTO—alone or in combination with transiliocolic obliteration of vein—has also been shown to be effective in controlling bleeding of duodenal varices that failed endoscopic therapy in poor surgical candidates, without complications or recurrence for a period of up to 3 years [104–108]. However, its use is currently limited because of its low availability and lack of technical expertise, and caution is required in patients with large OV. Most of the available literature addressing this method originates from the Far East, mainly Japan, and the early results are promising. B-RTO might be a difficult procedure, but it works well if the doctors are familiar with the procedure.

Special types of EcV

Intra-abdominal varices

Varices in the abdominal wall usually rupture externally and therefore are easily recognized and treated with local compression and surgical ligation. Many cases of veins around the falciform ligament of the liver that rupture into the peritoneal cavity have been reported and are characterized by a rapid increase in ascites, particularly in the presence of abdominal pain, and associated with a decrease in the haematocrit value. Management of this situation is difficult because transhepatic obliteration of the varices is relatively contraindicated because of the accompanying ascites. Other modes of therapy include transjugular, intrahepatic embolization of the portal venous system, TIPS placement, and emergency surgical ligation of the bleeding varix, but this results in high mortality [109].

Stomal varices

The term "ectopic stomal varices" refers to abnormally dilated veins that have developed in the stomal mucosa. Stomal varices are commonly seen in patients with ileostomies after proctocolectomy for inflammatory bowel disease associated with primary sclerosing cholangitis and PHT [24]. Stomal varices are recognized by the purplish hue around the stoma. As the patient is usually aware of the bleed early in the clinical course, he or she may be taught how to control the bleeding with pressure. Therefore, mortality from bleeding stomal varices is relatively low (3–4%) [92]. A case report from Korea described the use of 2D reformatted and 3D volume rendered images by multidimensional-CT in a patient with an episode of acute bleeding from the colonic stoma [110]. Local measures, such as epinephrine-soaked gauze, pressure dressings, suture ligation, gel foam or refashioning the stoma, have all been applied successfully in controlling the initial bleeding episode [93, 111]. Injection sclerotherapy has also been tried, but it may cause mucosal ulceration, stomal orifice stricture and peristomal skin necrosis [112]. In patients with uncontrolled or recurrent bleeding, a portosystemic procedure should be contemplated. TIPS—alone on in combination with embolization therapy—is preferred to a surgical portosystemic shunt because patients with stomal varices may be candidates for liver transplantation [111, 113–116].

Biliary tract varices

The development of biliary tract varices is a rare complication of PHT and sometimes is referred to as "pseudocholangiocarcinoma" or "pseudosclerosing cholangitis" or "portal hypertensive bilopathy". In a series of 42 patients with EPVO, gallbladder varices were detected by Doppler ultrasonography in 11 patients and choledochal varices in 9 patients [117]. Another study showed varices around the gallbladder and/or the biliary channels in 30% of patients [118]. Biliary varices are usually asymptomatic, often discovered accidentally, in the absence of other signs of PHT [119], but can lead to silent fatal outcomes [120]. Occurrence of varices around the common bile duct can result in extrahepatic biliary obstruction, cholangitis and haemobilia. The appearances of a biliary tree on endoscopic retrograde cholangiopancreatography may mimic those of primary sclerosing cholangitis [121]. Also, EUS can serve to diagnose biliary varices in patients with EPVO and jaundice. Although biliary varices are mainly asymptomatic, they may cause obstructive jaundice when they are located in the wall of the common bile duct. EUS can also detect unrecognized malignant tumours in patients with EPVO of undetermined origin [39]. Smith et al. [122]

reported a case of severe gastrointestinal bleeding due to jejunal-biliary anastomotic varices. The anastomotic site was the location of the pressure gradient, from the high-pressure small-bowel varices to the low-pressure biliary tract. This was successfully treated by disconnection of the anastomosis and bile duct repair. Therapeutic modalities of biliary varices include surgical portosystemic shunt and endoscopic, radiological or even surgical biliary drainage procedures.

Ano-rectal varices

The misdiagnosis of ano-rectal varices as haemorrhoids can sometimes have devastating consequences. Therefore, it is important to differentiate anal varices from haemorrhoids. The former collapse with digital pressure, whereas the latter do not. Rectal varices are enlarged portosystemic collateral veins, which develop in patients with PHT as a pathway for portal venous blood flow from the superior haemorrhoidal veins (portal) to and through the middle and inferior haemorrhoidal veins (systemic). Massive bleeding from ano-rectal varices has been reported, but such reports are infrequent [123–125]. On the other hand, haemorrhoids represent a common cause of rectal bleeding, but their prevalence in patients with PHT is not higher than the general population [126]. Weinshel et al. [127] have published an excellent review of the differentiation, pathogenesis and management of these two entities.

Ectopic IGV type 2

GV may be primary or secondary in origin. The former refers to those varices that are present at the initial endoscopic examination in a patient who never underwent variceal sclerotherapy or band ligation for OV, whereas the latter refers to GV that develop after endoscopic therapy for OV. Many classification systems have been proposed for GV on the basis of their location, size, colour, form and relation to OV. However, the most widely used classification system is that proposed by Sarin et al. [128, 129] and recommended by the Baveno III consensus working group [130], and categorizes GV on the basis of their location in the stomach and their relationship with OV (Table 7).

IGV2 are present either in the body or on the antrum of the stomach or upper duodenum. Sarin et al. [131] have described the prevalence, natural history and clinical significance of these varices in a prospective study involving 1,128 patients with PHT. Of these patients, 53 (4.7%) had IGV2. These IGV2 were predominantly (84.9%) secondary in origin and located in the antrum (53%), duodenum (32%) or at both sites (11%), and rarely in the corpus or fundus (4%). Bleeding due to IGV2 was seen only in three (5.7%) patients during a mean follow-up of 36.3 ± 12.1 months and could be successfully managed with endoscopic ligation or obliteration [131].

Recommended future research

Because of the low incidence of EcV and the lack of valid reproduced RCTs addressing this complication, we recommend the establishment of an international Web-based EcV case registry through which a more precise epidemiologic, diagnostic, prognostic, prophylactic and therapeutic data can be obtained. Major referral centres should be encouraged to participate and register their cases and subsequently be involved in multicentre RCTs concentrating on the management of such patients. Also, comparative haemodynamic studies between patients with G-O varices and those with EcV are warranted.

Concluding remarks

- Most of the available data about EcV are obtained from case reports or short series of cases.
- EcV are being recognized more frequently nowadays because of the continuous advances in the endoscopic and imaging techniques.
- Ectopic variceal bleeding is reported in patients with internal haemorrhage, hypovolaemic shock, occult gastrointestinal bleeding, anaemia or bleeding from the gut of obscure origin, especially in patients with PHT, following abdominal surgical procedures, in cases

Table 7 Classification of GV[a]

IGV (GV occurring in the absence of OV)		G-OV (GV extending from OV into the stomach)	
Type 1 (IGV1)	Type 2 (IGV2)	Type 1 (G-OV1)	Type 2 (G-OV2)
Varices located in the fundus that are often tortuous and complex in shape	EcV in the antrum, corpus and around the pylorus	Varices continuous with OV and extending along the lesser curve for about 2–5 cm below the G-O junction	Often long, tortuous, varices extending from the oesophagus below the G-O junction towards the fundus
8%	2%	74%	16%

G-O, gastro-oesophageal; G-OV, gastro-oesophageal varices; GV, gastric varices; IGV, isolated gastric varices; OV; oesophageal varices

[a] Adapted from references [120]–[122]

with extrahepatic venous obstruction or thrombosis, or anomalies of the portal venous system.

- It is difficult to recommend general guidelines for the management of EcV and each case should be dealt with depending on the site and severity of bleeding and according to available expertise and facilities.

- Patients with EcV bleeding must be considered difficult, should be managed in specialized canters, and have to be approached with a multidisciplinary team involving intensivists, gastroenterologists, surgeons and interventional radiologists.

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