CASE REPORT

Gastrointestinal Haemorrhage Secondary to Ectopic Varices at the Site of Previous Surgery

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ABSTRACT
Ectopic varices are uncommon but potentially life-threatening portosystemic venous collaterals. They are high variable and can occur anywhere across the gastrointestinal tract, except for the pathological sites for portoportal or portosystemic varices. Ectopic varices are responsible for 1% to 5% of all gastrointestinal haemorrhage cases, and their bleeding risk is four times that of oesophageal varices. Patients may present acutely with massive upper or lower gastrointestinal bleeding and consequent shock or chronically with anaemia of unknown origin. First-line diagnostic tests include contrast-enhanced computed tomography, gastrointestinal endoscopy, and angiography. Ectopic varices can be an incidental finding on laparotomy and at autopsy. Mortality can be as high as 40% when associated with severe acute haemorrhage. We report three cases of gastrointestinal haemorrhage secondary to ectopic varices at the site of previous surgery.

Key Words: Esophageal and gastric varices; Gastrointestinal hemorrhage

中文摘要

前手術部位的異位靜脈曲張造成的消化道出血

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異位靜脈曲張是並不常見但可能危及生命的門體靜脈側枝循環。除了門靜脈-門靜脈或門靜脈-體循環靜脈曲張病理以外，它們表現各異，可以發生在整個胃腸道的任何地方。異位靜脈曲張引致1%至5%的胃腸道出血病例，其出血風險是食道靜脈曲張的4倍。患者可出現急性上／下消化道大出血，並引至休克，或慢性表現為不明原因的貧血。一線診斷檢查包括對比增強電腦斷層掃描、胃腸內鏡檢查和血管造影。異位靜脈曲張也可以為一種在剖腹手術和屍檢的偶然發現。如有嚴重急性出血其死亡率可高達40%。我們報告了三例在前手術部位的異位靜脈曲張造成的消化道出血。

INTRODUCTION
Ectopic varices are responsible for 1% to 5% of all cases of gastrointestinal haemorrhage, and their bleeding risk is four times that of oesophageal varices.[1-5] Mortality can be as high as 40% when associated with an acute massive haemorrhage.[5-13]

Ectopic varices can be classified as non-occlusive or

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occlusive.[1] Non-occlusive varices are secondary to portal hypertension and increased portosystemic pressure. Occlusive varices are secondary to loco-regional factors such as scarring, adhesions, and altered venous anatomy following major surgical intervention or proximal venous thrombosis. Ectopic varices can be caused by portal hypertension, venous occlusion or thrombosis, familial causes or changes to vascular anatomy secondary to post-surgical adhesions and scarring.[3,9,14] Ectopic varices can also be caused by a combination of liver disease and loco-regional factors, as in two of our cases. The stimulus to angiogenesis at surgical anastamoses or post-surgical adhesions may cause ectopic varices at such sites in patients with portal hypertension.

Just like variceal collaterals at other anatomic sites

(oesophageal or anorectal), ectopic varices are veins with thicker walls and a larger diameter and result in greater wall tension and transmural pressure across the vessel wall. This is one of the major factors associated with the four-time increased risk of haemorrhage in ectopic varices.[15-19]

Haemorrhage from ectopic varices is uncommon. It can be acute and present as fresh peri-rectal or stomal bleeding, melaena, and intra-abdominal haemorrhage. In chronic cases, patients can present with melaena or simply iron deficiency anaemia of unknown origin.

CASE SERIES
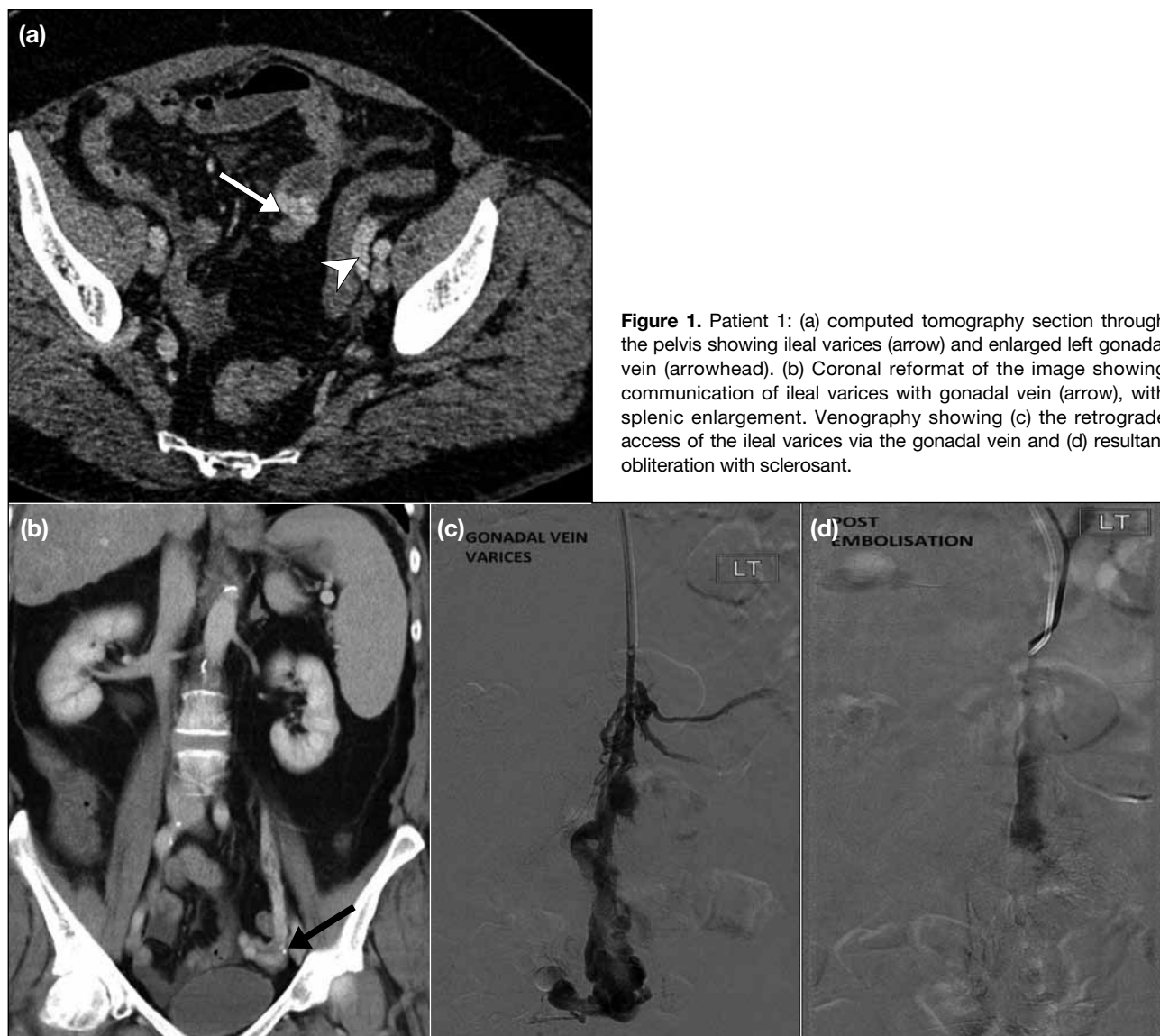### Patient 1
In April 2015, a 74-year-old woman with non-

Figure 1. Patient 1: (a) computed tomography section through the pelvis showing ileal varices (arrow) and enlarged left gonadal vein (arrowhead). (b) Coronal reformat of the image showing communication of ileal varices with gonadal vein (arrow), with splenic enlargement. Venography showing (c) the retrograde access of the ileal varices via the gonadal vein and (d) resultant obliteration with sclerosant.

alcoholic steatohepatitis liver disease presented with repeated episodes of melaena requiring multiple blood transfusions. She had undergone hysterectomy in August 2014. Computed tomography (CT) showed evidence of portal hypertension and ectopic varices at the left pelvis, close to the distal ileum, as a portosystemic communication between peripheral branches of the superior mesenteric vein and the left gonadal vein, presumably having developed through adhesions to the ileum at the site of the pelvic surgery (Figure 1).

Transjugular intrahepatic portosystemic shunting (TIPS) was performed to reduce portal pressure. Nonetheless, the patient continued to have multiple episodes of

haemorrhage from the ileum. CT and capsule endoscopy confirmed the ileal location of the haemorrhage. Endovascular embolisation of the ectopic varices was performed through the right internal jugular venous access. The gonadal vein was selectively catheterised retrogradely, and the ectopic varices were embolised with sclerosant (sodium tetradecyl sulphate) [Figure 1].

Patient 2

In February 2015, a 54-year-old man underwent a Whipple's resection for obstructive jaundice secondary to pancreatic head carcinoma. At the 6-month follow-up, CT showed recurrence in the pancreatic bed causing stenosis of the proximal portal vein. Standard

Figure 2. Patient 2: (a,b) computed tomography of the upper abdomen showing multiple varices (arrows) related to the choledochoenterostomy in a patient with portal vein thrombosis (arrowheads) caused by recurrence of pancreatic head carcinoma. Venography showing (c) the occluded proximal portal vein with the portal venous system accessed via the right percutaneous approach, (d) multiple portoportal venous collaterals around choledochoenterostomy (arrow) with the catheter manoeuvred beyond the obstruction, and (e) disappearance of the multiple collaterals following portal venous shunting.

chemotherapy was commenced, and 6 months later the patient presented with continuous perirectal bleeding requiring multiple blood transfusions. CT showed major portal venous stenosis and multiple varices around the choledochoenterostomy acting as a portoportal shunt (Figure 2).

Percutaneous portal venous shunting was performed. The right portal vein was accessed via a right lateral, low intercostal percutaneous approach, and a 10 mm x 7 cm Wallstent (Boston Scientific, Natick [MA], USA) was deployed across the stenotic segment. The procedure resulted in reversal of the pressure gradient and no filling of the ectopic veins (Figure 2). TIPS was then performed through the right hepatic vein and a 12 mm x 6 cm Wallstent was used to create the tract. Despite effective reduction of the portosystemic pressure gradient, venography showed persistent filling of the inferior mesenteric vein and communication with left iliac veins via multiple, peristomal, and subcutaneous venous collaterals. The patient continued to have frequent stomal bleeding episodes. He was admitted to the accident and emergency department with major upper gastrointestinal bleeding. He was resuscitated and underwent surgery to tie off the varices and had no further bleeding episodes.

Patient 3

In December 2016, a 51-year-old woman presented with repeated episodes of bleeding from the left lower quadrant stoma. She had undergone an anterior resection for colorectal cancer with left lower quadrant end colostomy 18 months earlier. She was known to have alcoholic liver disease with portal hypertension. CT did not show any active arterial bleeding. Superior mesenteric angiography in the venous phase showed ectopic varices related to the left lower quadrant stoma (Figure 3).

The portal venous system was accessed via the TIPS, and the inferior mesenteric vein was retrogradely catheterised. A coaxial catheter was inserted and manoeuvred into the stomal varices, and the ectopic varices were embolised with sclerosant (1.5 ml of sodium tetradecyl sulphate). The varices were completely obliterated with no contrast filling of systemic veins (Figure 3).

DISCUSSION

Portal hypertension secondary to liver cirrhosis causes increased pressure within the portal circulation. This can cause portosystemic varices in usual (gastro-oesophageal, caput medusa, anorectal) and ectopic locations. If the shunts are large enough they can compete with the portal venous flow itself and reduce flow in the splanchnic venous circulation with potential thrombosis. Conversely, portal vein thrombosis as a primary pathology can lead to ectopic portoportal and
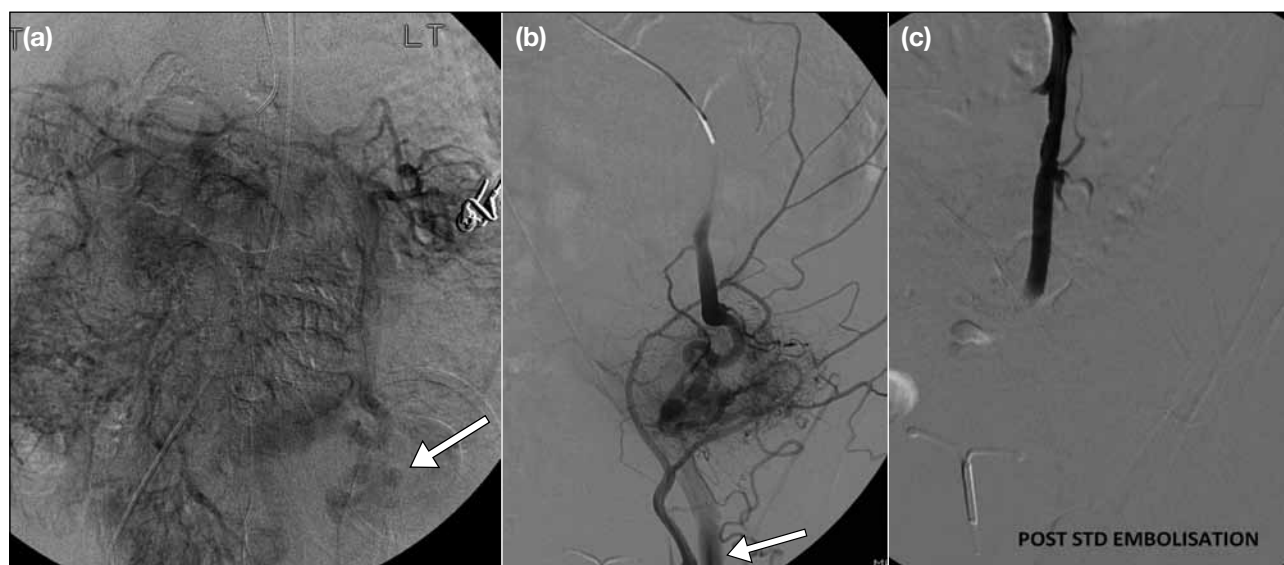

Figure 3. Patient 3: (a) the venous phase of superior mesenteric angiography showing portosystemic varices arising from the inferior mesenteric vein around left lower quadrant colostomy (arrow). (b) Retrograde catheterisation of the inferior mesenteric vein with multiple peristomal varices draining via cutaneous veins to the left external iliac vein (arrow). (c) Obliteration of varices after embolisation with sclerosant.

portosystemic flow.

In many patients, the venous outflow alteration and obstruction secondary to surgical intervention is the most likely loco-regional factor attributed to dilated venous channels bypassing the site of local scarring and adhesions. This process can be independent of portal hypertension or can be secondary to surgery as part of portal hypertension management. One example is surgical resection and anastomosis for ulcerative colitis associated with primary sclerosing cholangitis. Such patients can develop colonic, ileal, jejunal, or parastomal varices depending on the type of surgical procedure.[15] Similarly, jejunal varices can be secondary to portal vein resection or stenosis secondary to pancreatoduodenectomy and radiotherapy.[2,16]

Investigation

Contrast-enhanced CT has revolutionised the diagnosis, identification, planning, and management of variceal haemorrhage. With the advent of the 64-slice CT scanner, even actively bleeding, unstable patients can be readily imaged in a single breath hold. Contrast-enhanced CT in arterial and portovenous phases allows detection of minute-degree of blood loss as little as 0.5 ml/sec.[20,21] The software allows desired slice thickness from the raw volume data and reconstruction into coronal and sagittal planes and provides accurate anatomical detail for interventional, endoscopic, or surgical planning. CT is useful in the diagnosis of underlying pathology. It can effectively spot liver cirrhosis, secondary signs of portal hypertension or loco-regional surgical causes. One particular advantage of CT over all other modalities is detection of extra luminal varices, in particular at unusual locations such as the site of surgical adhesions, parastomal, surgical anastomoses, or mesentery.

Capsule endoscopy, endoscopy, and endoscopic ultrasonography are effective for diagnosing luminal varices or identifying the site of intraluminal haemorrhage. Capsule endoscopy is useful in detection of obscure luminal varices in patients with liver cirrhosis.[22] Endoscopy should be the first-line investigation tool in patients with known liver cirrhosis. Nonetheless, the usefulness of these modalities is limited in cases of ectopic, extra-luminal venous collaterals.

Upper and lower gastrointestinal endoscopy and contrast-enhanced CT are the mainstay of diagnostic tests in the acute setting. In more stable and chronic presentations, other diagnostic options such as capsule endoscopy and red cell nuclear scanning, contrast-enhanced magnetic resonance imaging, and laparoscopy can also be used. Laparotomy is rarely used as a primary investigative method.

The methods and modalities used in diagnosis vary, depending on the type and location of varices and the manner of presentation (acute or chronic), the variety of diagnostic modalities at hand, and the resources available to provide interventional radiology and endoscopy services.

Treatment

Management of haemorrhage from ectopic varices requires a high index of clinical suspicion, the correct diagnostic tests, and a multidisciplinary team of interventional radiologists, endoscopists, and surgeons. CT, angiography, and endoscopy can accurately locate the varices and help classify them as luminal or extra-luminal. In acute, life-threatening luminal haemorrhage or intra-peritoneal bleeding, priority goes to immediate resuscitation that includes intravenous access, intravenous colloids and blood products, and immediate transfer of the patient to a specialist centre.

After haemodynamic stabilisation, secondary medical management with vasoactive drugs (octreotide and vasopressin) injected intravenously or endoscopically for accessible varices (i.e. gastroduodenal or rectal) is recommended.[23,24] Nonetheless, locoregional endoscopic injection of vasoactive medication may not be feasible in ectopic variceal haemorrhage secondary to post-surgical scarring due to their difficult or extra-luminal position.

Endoscopy (Sclerosant and Band Ligation)

Endoscopy enables immediate treatment of the accessible luminal varices with sclerotherapy or band ligation.[25-27] Use of sclerosant in large rectal varices is equivocal and the combined use of band ligation and local injection has been reported.[25-28] Various sclerosing and thrombotic agents including cyanoacrylate glue, thrombin, and argon laser have been reported to effectively control haemorrhage.[29-32]

Transjugular Intrahepatic Portosystemic Shunting

TIPS should be the first-line management of non-occlusive varices secondary to portal hypertension.[33] Creation of a shunt between the portal and hepatic vein

can reduce the porto-systemic pressure gradient. In two of our patients, TIPS was performed to reduce the portosytemic gradient. In the third patient, shunting of the stenosed portal vein was required to reduce portal venous pressure and filling of portoportal collaterals. TIPS has been reported to effectively control ectopic varices.[33,34] Nonetheless, concurrent or subsequent selective embolisation of the bleeding varices may be needed in cases of unsatisfactory control of bleeding by TIPS alone or as a bridge to liver transplant. TIPS is a complex procedure and its risks include failure and liver damage causing active bleeding. Even with successful intervention, patients are at risk of hepatic encephalopathy. Thus, patient selection, pre-procedure antibiotics, rehydration, and anaesthetic support are imperative.

Embolisation
Interventional radiology techniques enable steerable microcatheters, wires, and detachable coils to access difficult, peripheral areas of the circulation. Various embolic materials like sodium tetradecyl sulphate, gel foam, alcohol, and cyanoacrylate can be used, depending on the size of vessel and speed of blood flow. The immediate success in control of haemorrhage with coil embolisation has been reported to be up to 94%.[35,36]

Previously, the endpoint of embolisation was occlusion of the proximal feeding vein from the portal venous side (the high pressure gradient component). With technological advancements, super-selective embolisation at or near the site of active extravasation can be carried out. Endovascular embolisation can be performed in conjunction with TIPS or other endoscopic treatments to achieve satisfactory outcome. In two of our patients, initial TIPS failed to completely stop bleeding and subsequent selective ectopic variceal embolisation was performed. This supports the theory that bleeding in ectopic varices secondary to post-surgical scarring is a result of a combination of local anatomic features and portal hypertension.[1-3] In one patient, the varices were selectively catheterised using a transjugular approach through the existing TIPS into the portal circulation. In another patient, the varices were accessed via the systemic side of the portosystemic shunt.

Percutaneous coil embolisation can be very challenging in patients with portal venous thrombosis or occlusion. In a retrospective case series of 14 patients (12 of them had previous abdominal surgery) who underwent percutaneous coil embolisation for ectopic varices, re-bleeding occurred within the first 72 hours in two patients and after 23 days to 27 months in nine patients.[37,38]

Medical Treatment
The role of β-blockers in the primary and secondary prophylaxis of bleeding from gastro-oesophageal varices is well established. Nonetheless, the use of β-blockers or nitrates in the long-term management of patients with ectopic varices is not.

Surgical Treatment
Surgery is the last resort if endoscopic modalities or interventional embolisation in combination with TIPS fail to control bleeding. Surgical options include laparoscopic procedures, selective devascularisation techniques, or open exploration depending upon the site of varices.[38]

Balloon-occluded Retrograde Transvenous Embolisation
This technique is extremely demanding due to the highly variable venous anatomy and requires thorough knowledge of gastric variceal anatomy. A transfemoral or transjugular approach can be used. In the transjugular approach, a balloon occlusion, coaxial catheter system is manoeuvred into the gastrorenal shunt and gastric varix. After occlusion balloon venography, sclerosant is injected to obliterate the gastric varix.

CONCLUSION
Patients who present with acute gastrointestinal haemorrhage should be investigated with endoscopy and CT to identify the source of bleeding. The importance of effective resuscitation with fluids, colloids, blood products, and vasoactive drugs cannot be overstated. Endoscopy is the therapeutic modality of choice for luminal varices in patients with known liver cirrhosis and portal hypertension. In ectopic varices secondary to post-surgical scarring (which are extra-luminal), interventional radiological techniques such as TIPS and selective embolisation of the bleeding varices are appropriate. Both can be performed individually or combined concurrently or consecutively depending on the underlying pathology and anatomy.

REFERENCES
1. Saad WE, Lippert A, Saad NE, Caldwell S. Ectopic varices: anatomical classification, hemodynamic classification, and hemodynamic-based management. Tech Vasc Interv Radiol. 2013;16:158-75. **cross**ref

2. Helmy A, Al Kahtani K, Al Fadda M. Updates in the pathogenesis, diagnosis and management of ectopic varices. Hepatol Int. 2008;2:322-34. **cross ref**

3. Hashimoto N, Akhoshi T, Yoshida D, Kinjo N, Konishi K, Uehara H, et al. The efficacy of balloon-occluded retrograde transvenous obliteration on small intestinal variceal bleeding. Surgery. 2010;148:145-50. **cross ref**

4. Kochar N, Tripathi D, McAvoy NC, Ireland H, Redhead DN, Hayes PC. Bleeding ectopic varices in cirrhosis: the role of transjugular intrahepatic portosystemic stent shunts. Aliment Pharmacol Ther. 2008;28:294-303. **cross ref**

5. Onozato Y, Kakizaki S, Iizuka H, Mori K, Takizawa D, Ohyama T, et al. Ectopic varices rupture in the gastroduodenal anastomosis successfully treated with N-butyl- 2-cyanoacrylate injection. Acta Med Okayama. 2007;61:361-5.

6. Fleming RJ, Seaman WB. Roentgenographic demonstration of unusual extra-esophageal varices. Am J Roentegenol Radium Ther Nucl Med. 1968;103:281-90. **cross ref**

7. Amin R, Alexis R, Korzis J. Fatal ruptured duodenal varix: a case report and review of literature. Am J Gastroenterol. 1985;80:13-8.

8. Kanagawa H, Mima S, Kouyama H, Gotoh K, Uchida T, Okuda K. Treatment of gastric fundal varices by balloon-occluded retrograde transvenous obliteration. J Gastroenterol Hepatol. 1996;11:51-8. **cross ref**

9. Shimizu T, Okabayashi Y, Andoh Y. The duodenal varices: report of a case and review of 70 Japanese cases. Mie Med J. 1992;42:215-21.

10. Hashizume M, Tanoue K, Ohta M, Ueno K, Sugimachi K, Kashiwagi M, et al. Vascular anatomy of duodenal varices: angiographic and histopathological assessments. Am J Gastroenterol. 1993;88:1942-5.

11. Itzchak Y, Glickman MG. Duodenal varices in extrahepatic portal obstruction. Radiology. 1977;124:619-24. **cross ref**

12. Wang CS, Jeng LB, Chen MF. Duodenal variceal bleeding—successfully treated by mesocaval shunt after failure of sclerotherapy. Hepatogastroenterology. 1995;42:59-61.

13. Khougeer F, Morrow C, Jordan P. Duodenal varices as a cause of massive upper gastrointestinal bleeding. Surgery. 1987;102:548-52.

14. Takamura K, Miyake H, Mori H, Terashima Y, Ando T, Fujii M, et al. Balloon occluded retrograde transvenous obliteration and percutaneous transhepatic obliteration for ruptured duodenal varices after operation for rectal cancer with multiple liver metastasis: report of a case. J Med Invest. 2005;52:212-7. **cross ref**

15. Wiesner RH, LaRusso NF, Dozois RR, Beaver SJ. Peristomal varices after proctocolectomy in patients with primary sclerosing cholangitis. Gastroenterology. 1986;90:316-22. **cross ref**

16. Sakai M, Nakao A, Kaneko T, Takeda S, Inoue S, Yagi Y, et al. Transhepatic portal venous angioplasty with stenting for bleeding jejunal varices. Hepatogastroenterology. 2005;52:749-52.

17. Salam AA, Goldman M, Smith D Jr, Hill HL. Gastric, intestinal, and gallbladder varices: hemodynamic and therapeutic considerations. South Med J. 1979;72:402-8. **cross ref**

18. Wilson SE, Stone RT, Christie JP, Passaro E Jr. Massive lower gastrointestinal bleeding from intestinal varices. Arch Surg. 1979;114:1158-61. **cross ref**

19. Groszmann RJ. Reassessing portal venous pressure measurements. Gastroenterology. 1984;86:1611-4.

20. Weishaupt D, Pfammatter T, Hilfiker PR, Wolfensberger U, Marincek B. Detecting bleeding duodenal varices with multislice helical CT. AJR Am J Roentgenol. 2002;178:399-401. **cross ref**

21. Jain TP, Gulati MS, Makharia GK, Paul SB. Case of the season: detection of duodenal varices by CT enteroclysis. Semin Roentgenol. 2005;40:204-6. **cross ref**

22. De Palma GD, Rega M, Masone S, Persico F, Siciliano S, Patrone F, et al. Mucosal abnormalities of the small bowel in patients with cirrhosis and portal hypertension: a capsule endoscopy study. Gastrointest Endosc. 2005;62:529-34. **cross ref**

23. Besson I, Ingrand P, Person B, Boutroux D, Heresbach D, Bernard P, et al. Sclerotherapy with or without octreotide for acute variceal bleeding. N Engl J Med. 1995;333:555-60. **cross ref**

24. Silvain C, Carpentier S, Sautereau D, Czernichow B, Metreau JM, Fort E, et al. Terlipressin plus transdermal nitroglycerin vs. octreotide in the control of acute bleeding from esophageal varices: a multicenter randomized trial. Hepatology. 1993;18:61-5.

25. Lebrec D, Benhamou JP. Ectopic varices in portal hypertension. Clin Gastroenterol. 1985;14:105-21.

26. Firoozi B, Gamagaris Z, Weinshel EH, Bini EJ. Endoscopic band ligation of bleeding rectal varices. Dig Dis Sci. 2002;47:1502-5. **cross ref**

27. Bosch A, Marsano L, Varilek GW. Successful obliteration of duodenal varices after endoscopic ligation. Dig Dis Sci. 2003;48:1809-12. **cross ref**

28. Ikeda K, Konishi Y, Nakamura T, Nabeshima M, Yamamoto S, Migihashi R, et al. Rectal varices successfully treated by endoscopic injection sclerotherapy after careful hemodynamic evaluation: a case report. Gastrointest Endosc. 2001;54:788-91. **cross ref**

29. Sans M, Llach J, Bordas JM, Andreu V, Reverter JC, Bosch J, et al. Thrombin and ethanolamine injection therapy in arresting uncontrolled bleeding from duodenal varices. Endoscopy. 1996;28:403. **cross ref**

30. Bhasin DK, Sharma BC, Sriram PV, Makharia G, Singh K. Endoscopic management of bleeding ectopic varices with histoacryl. HPB Surg. 1999;11:171-3. **cross ref**

31. Schafer TW, Binmoeller KF. Argon plasma coagulation for the treatment of colonic varices. Endoscopy. 2002;34:661-3. **cross ref**

32. Chen WC, Hou MC, Lin HC, Chang FY, Lee SD. An endoscopic injection with N-butyl-2-cyanoacrylate used for colonic variceal bleeding: a case report and review of the literature. Am J Gastroenterol. 2000;95:540-2. **cross ref**

33. Tripathi D, Jalan R. Transjugular intrahepatic portosystemic stent-shunt in the management of gastric and ectopic varices. Eur J Gastroenterol Hepatol. 2006;18:1155-60. **cross ref**

34. Vidal V, Joly L, Perreault P, Bouchard L, Lafortune M, Pomier-Layrargues G. Usefulness of transjugular intrahepatic portosystemic shunt in the management of bleeding ectopic varices in cirrhotic patients. Cardiovasc Intervent Radiol. 2006;29:216-9. **cross ref**

35. Ohnishi K, Takayasu K, Takashi M , Suzuki N, Hirashima T, Hara T, et al. Transhepatic obliteration of esophageal varices using stainless coils combined with hypertonic glucose and gelfoam. J Clin Gastroenterol. 1985;7:200-7. **cross ref**

36. Smith-Laing G, Scott J, Long RG, Dick R, Sherlock S. Role of percutaneous transhepatic obliteration of varices in the management of hemorrhage from gastroesophageal varices. Gastroenterology. 1981;80:1031-6.

37. Macedo TA, Andrews JC, Kamath PS. Ectopic varices in the gastrointestinal tract: short- and long-term outcomes of percutaneous therapy. Cardiovasc Intervent Radiol. 2005;28:178-84. **cross ref**

38. Hsieh JS, Wang WM, Perng DS, Huang CJ, Wang JY, Huang TJ. Modified devascularization surgery for isolated gastric varices assessed by endoscopic ultrasonography. Surg Endosc. 2004;18:666-71. **cross ref**