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ESSENTIALS A wide range of vascular disorders and vasculitides may affect the gastrointestinal tract. Most are quite uncommon, but presentations are often dramatic with intestinal bleeding or gangrene. Intestinal ischaemia is most commonly due to atherosclerosis or thrombosis causing arterial or venous mesenteric vascular oc- clusion. There are four primary syndromes.
(1) Ischaemic colitis— presents with abdominal pain, nausea, vomiting, and tenderness followed by passage of loose bloody stool. Supportive manage- ment is usually sufficient, but a key challenge is early identification of patients with severe injury who are likely to progress to transmural ulceration and perforation. (2) Acute mesenteric ischaemia—typic- ally presents with sudden abdominal pain, initially without localizing signs such that diagnosis is often delayed. Priorities of management are resuscitation, exclusion of other causes of apparent abdominal catastrophe, and prompt laparotomy to resect ischaemic bowel. (3) Chronic mesenteric ischaemia—most often caused by athero- sclerotic disease and presents with severe and poorly localized cramping abdominal pain after eating. Diagnosis requires evidence of vascular occlusion on imaging, and revascularization is the defini- tive management strategy. (4) Mesenteric venous thrombosis—diag- nosis is most commonly via cross-sectional imaging. The mainstay of treatment is supportive, as well as anticoagulation and a search for predisposing factors. Vasculitides affecting the intestine may be primary (e.g. polyarteritis nodosa, Henoch–Schönlein purpura, and antineutrophil cytoplasmic antibody-associated vasculitis) or secondary (related to underlying infection, exposure to drugs, or a connective tissue disorder such as systemic lupus erythematosus or rheumatoid arthritis). Abdominal symptoms rarely dominate the clinical picture. Vascular lesions of the gastrointestinal tract may present with acute haemorrhage, chronic iron deficiency anaemia, or obstruc- tion. Lesions include angiodysplasias, telangiectasias, haemangi- omas, Dieulafoy lesions, and gastric antral vascular ectasia. These lesions may occur in isolation or as part of a syndrome (e.g. heredi- tary haemorrhagic telangiectasia). Intestinal ischaemia Intestinal ischaemia most commonly occurs

due to arterial or venous mesenteric vessel occlusion from atherosclerosis or embolism. Although uncommon, intestinal ischaemia is associated with high mortality and has an increasing incidence, which is rising in par- allel with an ageing population and the incidence of atherosclerosis. These conditions lead to a range of clinical syndromes and present a diagnostic challenge due to their nonspecific clinical features. Vascular anatomy A basic understanding of the vascular anatomy of the gastrointes- tinal (GI) tract is required (Figs. 15.17.1 and 15.17.2). The main blood supply derives from three branches of the abdominal aorta— the coeliac trunk, the superior mesenteric artery (SMA), and the in- ferior mesenteric artery (IMA). The arteries arising from the coeliac trunk supply the stomach, proximal duodenum, liver, and pancreas. SMA branches supply the distal duodenum, jejunum, and ileum through vascular arcades that terminate in straight end arteries (the vasa recta). Branches of the SMA and IMA anastomose at regular intervals to supply the colon—the right colon by the ileocolic and right colic branches, the transverse colon by the middle colic branch, and the left colon by branches of the IMA. Limited collaterals from the IMA and SMA create a watershed area at the splenic flexure (Griffith's point) which is susceptible to ischaemia during low-flow states. The rectosigmoid junction is another potential watershed area (Sudeck's point). The lower rectum is supplied by both the su- perior rectal artery (a continuation of the IMA) and vessels arising out of the internal iliac artery. This dual blood supply means the lower rectum is often spared from ischaemic episodes. Duodenal venous drainage follows the arterial arcades through to the portal vein. The superior mesenteric vein drains the small bowel and right colon and the inferior mesenteric vein drains the left colon. Pathophysiology Two main mechanisms damage the intestine after vascular occlu- sion. First, cellular dysfunction and necrosis results from direct

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 deprivation of oxygen and nutrients. Second, once oxygenation is restored, reperfusion injury may exacerbate intestinal damage. Reperfusion injury is a complex and incompletely understood in- flammatory response. Reactive oxygen metabolites (including superoxide, hydrogen peroxide, and hydroxyl radicals) and lib- erated proinflammatory intracellular components play key roles. Vasoactive mediators are released into the circulation, intestinal barrier integrity is lost, and bacterial translocation, septic shock, and multiorgan failure may occur. Venous obstruction increases mesenteric venous bed resistance resulting in diminished perfusion, venous congestion, and intestinal wall oedema. Capillary rupture ensues with increasing oedema, and chronically dilated collaterals may bleed. Bowel infarction occurs with complete occlusion of venous return. Various complex and interacting homeostatic mechanisms pro- tect the GI tract from ischaemic injury. Intrinsic processes of splanchnic blood flow autoregulation (including reactive hyper- aemia, pressure–flow autoregulation, and hypoxic vasodilation) help prevent intestinal ischaemia. In addition, collateral vessels tem- porarily dilate and allow for continued perfusion if an abrupt ob- struction to the blood supply occurs. Clinical syndromes Intestinal ischaemia is classified into distinct syndromes based on clinical presentation and the GI tract segment involved. Any one clinical syndrome may have multiple aetiologies (e.g. acute mes- enteric ischaemia can result from both embolic and atherosclerotic disease). Acute ischaemia threatens intestinal viability, whereas the less common chronic ischaemia occurs when compromised blood supply cannot support the functional needs of the GI tract. Ischaemic colitis Ischaemic colitis is the most common form of intestinal ischaemia and the cause of up to 1 in 2000 hospital admissions. The true in- cidence is uncertain as many mild cases go unrecognized due to transient symptoms. Most cases occur in patients aged 60 years or older. Intestinal ischaemia

commonly complicates aortic valve and abdominal aortic aneurysm repair surgery. Other risk factors in- clude vasculitis, coagulation disorders, and drug use (e.g. cocaine, digoxin, sumatriptans, and nonsteroidal anti-inflammatory agents (NSAIDs)). The colon is more susceptible to ischaemia than the small bowel due to its less well-developed microcirculation. In addition, dis- tension of the colon may impair blood flow. Ischaemic colitis may therefore occur in the segment of intestine immediately proximal to an obstructing lesion or with colonic pseudo-obstruction. Ischaemic colitis may affect any part of the large intestine, al- though the lower rectum is usually spared because of its dual blood supply. The left colon is most frequently affected, especially the watershed areas of the splenic flexure and rectosigmoid junction. However, the right colon is also susceptible to ischaemia because the vasa recta is less well developed. The spectrum of tissue damage is broad, and ranges from mild, superficial, and patchy changes to extensive necrosis. Patients present with abdominal pain, tenderness, nausea, and vomiting, progressing to urgency and bright red or maroon stool. Supportive management is usually sufficient because most cases Middle colic artery Right colic artery Ileal branches Ileocolic artery Superior mesenteric artery Fig. 15.17.1 Anatomy of the superior mesenteric artery. From MacKay GJ, Dorrance HR, Molloy RG, O'Dwyer PJ (eds) (2010). Colorectal surgery (Oxford specialist handbooks in surgery). By permission of Oxford University Press. Middle colic Marginal artery of Drummond Inferior mesenteric Left colic Middle and inferior rectal Superior rectal Superior mesenteric Ileocolic Right colic Appendicular Fig. 15.17.2 Blood supply to the colon. From MacKay GJ, Dorrance HR, Molloy RG, O'Dwyer PJ (eds) (2010). Colorectal surgery (Oxford specialist handbooks in surgery). By permission of Oxford University Press.

 spontaneously resolve. The key challenges for the clinician are to dif- ferentiate ischaemic colitis from inflammatory bowel disease (endo- scopic and histological appearances can be similar) and promptly identify those patients with severe injury who are likely to progress to transmural ulceration and perforation. Initial plain films and laboratory tests are nonspecific but may be diagnostic and are useful in identifying other causes for symp- toms. Colonoscopy with minimal insufflation in an unprepared colon should be organized within 48 h. Findings include oedema, erosions, and ulcers with segmental or circumferential submucosal haemorrhages (Fig. 15.17.3). Planned follow-up colonoscopy is per- formed to ensure improvement. CT and CT angiography often show nonspecific bowel wall thickening and are helpful to exclude other causes, especially if acute mesenteric ischaemia is being considered (Fig. 15.17.4). Supportive treatment is with intravenous fluids, broad-spectrum antibiotics, and bowel rest. Urgent laparotomy may be required if gangrene or perforation occurs. Colonic strictures may develop fol- lowing recovery, although these rarely require intervention.

Acute mesenteric ischaemia

Acute mesenteric ischaemia is a syndrome caused by an abrupt interruption of mesenteric blood flow that leads to intestinal is- chaemia, inflammation, and eventually, infarction. It is relatively rare (approximately 0.1% of hospital admissions). Risk factors in- clude those related to embolic disease (e.g. atrial fibrillation, recent acute myocardial infarction, and valvular heart disease) and athero- sclerotic disease (e.g. increasing age, diabetes, smoking, and high cholesterol). For younger patients, risk factors include underlying thrombophilia, and cocaine, amphetamine, and vasoactive medica- tion use. Mortality remains at 30 to 70% despite improvements in investigation and management. Acute mesenteric ischaemia may be due to embolic, thrombotic, or nonocclusive processes. Emboli cause up to a third of episodes and arise from the heart in 80% of cases, hence the condition is most common in patients with primary cardiac conditions such as atrial fibrillation, bacterial endocarditis, or recent myocardial infarction

with left ventricular thrombus. Uncommon causes include paradox- ical embolism through a patent foramen ovale and embolism from aortic mural thrombi. Emboli lodge preferentially just distal to the origin of the middle colic artery from the SMA. Acute mesenteric arterial thrombosis is caused by plaque rupture in the setting of severe atherosclerosis, most commonly near the origin of the SMA. Extensive compensatory collateral circulation has usually developed due to the gradual nature of atherosclerosis. This collateral formation (and the rich, redundant nature of the in- testinal blood supply) means that at least two major mesenteric ves- sels are generally affected before infarction occurs. Rarely, all three vessels may be occluded without visceral damage. Nonocclusive mesenteric ischaemia is responsible for around 20% of cases of acute mesenteric ischaemia. This occurs due to dispro- portionate splanchnic vasoconstriction resulting from a sustained state of hypoperfusion, most often in settings of elderly patients with severe low-output cardiac failure, patients requiring haemodialysis, and patients with shock requiring intensive care therapy (including those who have had cardiopulmonary bypass surgery). A variety of Fig. 15.17.3 Endoscopic appearances of ischaemic colitis with changes ranging from patchy erythema to frank ulceration in a sharply defined segment of involvement. Reproduced from www.gastrointestinalatlas.com. (a) (b) Fig. 15.17.4 Ischaemic colitis—CT features. Axial (a) and coronal reformatted (b) images demonstrate mural thickening with submucosal oedema (arrows) of the transverse colon and both colonic flexures in a 34-year-old woman with systemic lupus erythematosus. From Levy AD, Mortele KJ, Yeh BM (eds) (2015). Gastrointestinal imaging. By permission of Oxford University Press.

section 15 Gastroenterological disorders 3000 often interrelated factors are implicated, including medications (par- ticularly vasopressors and digoxin), cardiovascular events, and pre- existing atherosclerosis. For elective cardiac surgery patients, risk factors include older patients, renal insufficiency, diuretic therapy, need for intra-aortic balloon pump support, and postoperative high serum lactate. Acute mesenteric ischaemia from any cause may cause a spectrum of intestinal tissue damage ranging from self-limiting and transient superficial injury to life-threatening transmural necrosis. There is wide variability in clinical features, which depend on the extent of is- chaemic damage. The classic presentation is of acute, severe abdom- inal pain, colicky in the early stages but progressing to constant and unremitting, that lasts longer than a few hours. This presentation of overwhelming distress with relatively unimpressive clinical signs makes early diagnosis challenging. Clinical features are nonspecific and include nausea, vomiting, diarrhoea with or without blood, ab- dominal distension, and hyperactive bowel sounds. With progres- sion to transmural infarction, peritonitis and cardiovascular collapse may occur. A preceding history of weeks or months of postprandial abdominal pain, nausea, and weight loss suggests SMA thrombosis. Initial investigations include routine blood tests for evaluation of abdominal pain and coagulation profile, arterial blood gas, lac- tate, and lipase. There are no reliable early serum markers for acute mesenteric ischaemia. Leucocytosis is common, but often only pre- sents late in the disease course. There may be a metabolic acidosis, although early profound vomiting may cause a metabolic alkalosis. Electrocardiogram findings of an arrhythmia might suggest an em- bolic source and help raise the index of suspicion for mesenteric ischaemia. After resuscitation, further initial investigations are per- formed mainly to exclude other courses of severe abdominal pain. Abdominal plain films are often normal early on but may show small-bowel and/or right colon thumbprinting indicating bowel wall oedema (Fig. 15.17.5), nonspecific dilatation of the small bowel with fluid levels, mesenteric vascular gas, or free intraperitoneal air. CT angiography has now become the initial investigation of choice because it can be performed expeditiously and is

less time-consuming and invasive than the historical gold standard of mesenteric angiography. It has high sensitivity and spe- cificity in identifying occlusive SMA disease but is less accurate for nonocclusive mesenteric ischaemia (Fig. 15.17.6). Magnetic resonance angiography (MRA) is often difficult to perform quickly and is limited by lower spatial resolution and slower acquisition times compared to CT angiography. Once gangrene develops (Fig. 15.17.7), the mortality rate of acute mesenteric ischaemia may be as high as 90%, highlighting the im- portance of avoiding delays in proceeding to definitive therapy. Initial management includes aggressive resuscitation (critical to combat further hypotension-related hypoperfusion), hospitaliza- tion, bowel rest, empirical broad-spectrum antibiotics, intensive monitoring, avoidance of vasoconstrictive medications, urgent se-nior surgical review, and prompt laparotomy to resect the ischaemic bowel and restore blood flow. Treatment decisions should ideally involve a multidisciplinary team approach with gastroenterol-ogists, general and vascular surgeons, radiologists, anaesthetists, Fig. 15.17.5 Plain abdominal radiograph of a man presenting with acute abdominal pain and rectal bleeding. There is thumbprinting (arrow) in the region of the splenic flexure, and the small bowel is dilated. Image reproduced with permission from Medscape Drugs & Diseases (https:// emedicine.medscape.com/), Ischemic Colitis Imaging, 2016, available at: https:// emedicine.medscape.com/article/366808-overview. Fig. 15.17.6 Acute arterial occlusive mesenteric ischaemia. CT images during intravenous contrast in early arterial phase in a patient with proximal thrombotic occlusion of the superior mesenteric artery show portal venous gas (arrow on left image), mesenteric venous gas (arrow on right image), pneumatosis intestinalis, and a diffuse lack of bowel wall enhancement in keeping with bowel infarction. From Levy AD, Mortele KJ, Yeh BM (eds) (2015). Gastrointestinal imaging. By permission of Oxford University Press.

 and intensivists. Any sign of peritonism mandates immediate lapar- otomy. Perioperative mesenteric angiography allows for the plan- ning of surgical resection and infusion of vasodilators and has been shown to improve outcomes. Depending on local interventional radiological expertise, aspiration embolectomy, thrombolysis, and endovascular stenting may be alternatives to laparotomy. If there is any doubt over the viability of residual intestine, a repeat lapar- otomy 12 to 24 h later is indicated. With nonocclusive mesenteric ischaemia, infiltration of the SMA with vasodilators during angiog- raphy may help relieve the obstruction. Chronic mesenteric ischaemia Chronic mesenteric ischaemia occurs when the physiological re- quirements of the GI tract cannot be met. It is less common than acute mesenteric ischaemia and accounts for less than 5% of all cases of intestinal ischaemia. Atherosclerotic disease is the cause in over 95% of cases, with the proximal regions of the coeliac artery, SMA, or IMA usually affected (hence the term ‘intestinal angina’). Uncommon causes include mesenteric vasculitis, fibromuscular dysplasia, and radiation. Risk factors are those associated with atherosclerosis. The nonspecific clinical features and lack of specific diagnostic tests make this a challenging diagnosis. The classic presentation is of severe, poorly localized, cramping abdominal pain occurring 20 to 60 min after eating that resolves within a few hours. The postprandial symptomatology may be due to atherosclerosis limiting the higher blood flow requirements, or from a ‘steal’ phenomenon (diversion of blood flow from intestinal to gastric circulation when food enters the stomach). Patients often present with a fear of eating and weight loss. Physical signs such as an abdominal bruit are nonspecific and clinically unhelpful. Common investigations for abdominal pain such as abdominal plain films, CT, and endoscopy are usually normal. CT angiog-raphy, MRA, duplex ultrasonography, or invasive catheter angiog- raphy may identify flow disturbances or occlusion. The diagnosis relies on a combination of clinical features consistent with

chronic mesenteric ischaemia, exclusion of other causes of abdominal pain, and radiological evidence of occlusion in at least two of the three major mesenteric arteries. However, due to the high incidence of asymptomatic mesenteric atherosclerotic disease with collateral for- mation, radiological evidence of obstruction in two or even three mesenteric arteries supports the diagnosis, but is insufficient in itself to prove it. Chronic mesenteric ischaemia may be a harbinger of significant atherosclerosis elsewhere and diagnosis should initiate cardiac in- vestigations. General treatment measures include management of atherosclerosis, including statins, weight loss, exercise, and cessation of smoking. Depending on local expertise and in selected cases with short segments of occlusion, endovascular stenting can have good long-term success rates. Stenotic lesions most commonly occur at the aortic origins of the vessels so definitive management at lapar- otomy with mesenteric reconstruction may be very rewarding. Mesenteric venous thrombosis Mesenteric venous thrombosis is less common than arterial oc- clusion. This condition affects younger people (mean age between 40 and 60 years) and has a male predominance. The superior mesenteric vein is far more commonly involved than the inferior mesenteric vein. Mesenteric venous thrombosis usually occurs in thrombophilias which are inherited (e.g. antithrombin III deficiency and pro- tein C or S deficiency) or acquired (e.g. polycythemia rubra vera, thrombocythaemia, and antiphospholipid antibody syndrome), systemic disorders causing a hypercoagulable state (e.g. cancer and nephrotic syndrome), a local abdominal process (e.g. pancreatitis and postsplenectomy, inflammatory bowel diseases), or stasis (e.g. congestive heart failure and portal hypertension). A clear precipitant cannot be found in 20 to 40% of cases. Presentation may be acute or chronic. The acute presentation has clinical features of acute mesenteric ischaemia and accounts for between 5 and 10% of such presentations. In the chronic presenta- tion, bowel infarction is less common and patients may present with (a) (b) Fig. 15.17.7 Colonic infarction—CT features. (a) Axial image obtained at the pelvic brim shows pneumatosis intestinalis of the ascending (arrow) and descending colon. Both the small bowel and colon are dilated and fluid filled. (b) A scan at the level of the liver demonstrates extensive intrahepatic portal venous gas. From Levy AD, Mortele KJ, Yeh BM (eds) (2015). Gastrointestinal imaging. By permission of Oxford University Press.

 vague abdominal pain, nausea, and vomiting. The condition is often asymptomatic until complications of portal hypertension develop (e.g. varices or ascites). As with acute mesenteric ischaemia, initial investigations are nonspecific. Acid–base abnormalities and leucocytosis may be pre- sent, and plain abdominal films may show dilated loops of bowel, mucosal oedema with thumbprinting, or features of perforation. CT with contrast is usually the imaging modality of choice and is highly accurate in demonstrating thrombi in the superior mesen- teric vein and portal veins. Abdominal Doppler ultrasonography may also be used but is less sensitive for smaller thrombi. Imaging may also show complications of portal hypertension such as varices and ascites. Nowadays, diagnosis seldom requires invasive mesen- teric angiography. Treatment is aimed at preventing bowel infarction by reperfusion of the intestine. For the acute presentation, management includes supportive therapy and immediate anticoagulation, which should generally continue for at least 6 months (or longer if there is an underlying thrombophilia). A systematic search for any underlying predisposing factors should be performed. Patients who pre- sent chronically may require management of portal hypertensive bleeding from varices with banding and/or transjugular intrahepatic portosystemic shunt. Vasculitides affecting the intestine Several vasculitides affect the GI tract, although involvement is usually part of a systemic process and rarely dominates the clinical picture. Vasculitis can affect

any splanchnic blood vessel, including arteries, arterioles, veins, venules, and the vasa recta. Inflammation of these vessels drives regional ischaemia, resulting in disruption of blood flow and eventually ischaemic damage to the intestine or other organs such as the pancreas and liver. Overall, the most common forms of intestinal vasculitis en- countered are those associated with polyarteritis nodosa, systemic lupus erythematosus, rheumatoid arthritis, and Henoch–Schönlein purpura. Intestinal involvement can lead to the full spectrum of tissue damage, from self-limiting superficial injury to perforation. Intestinal vasculitis may occur through a primary autoimmune process (‘primary’ vasculitis) or as part of a systemic process such as an underlying connective tissue disorder, infection, malig- nancy, or exposure to drugs (‘secondary’ vasculitis). This chapter will focus on the approach to patients with vasculitis affecting the intestine: the classification, pathophysiology, clinical features, and treatment of specific vasculitides are covered elsewhere (see Chapters 19.11.6, 19.11.7, 19.11.8, 19.11.9, and 19.11.10). Clinical presentation Patients with known systemic vasculitis or connective
tissue disease Most patients with GI involvement of primary or secondary vascu- litis have other clinical features related to the underlying systemic vasculitis or connective tissue disease. Indeed, diagnosis of intestinal vasculitis commonly relies on the systemic features and laboratory findings of the underlying illness, rather than the often nonspecific abdominal complications. In reality, most patients will already have been diagnosed with vasculitis, and the challenge for the clinician is to promptly identify GI involvement when present. Involvement of multiple specialty teams and interdisciplinary meetings are often required. The clinical features reflect the underlying vessels involved and depend on the extent and severity of disease in the affected intes- tine. Patients can present with any of the features of mesenteric ischaemia. The acute presentation of GI vasculitis is indistinguish- able from acute mesenteric ischaemia, including severe abdominal pain, a relative lack of clinical signs, and progression to peritonitis and shock, but vasculitis accounts for only around 2% of all causes of intestinal infarction. The chronic presentation is indistinguish- able from chronic mesenteric ischaemia due to atherosclerosis, with intestinal angina, fear of eating, and progressive weight loss. Vasculitis of the intestine can lead to protein-losing enteropathy and subsequent cachexia, peripheral oedema, and ascites. Nonspecific symptoms of nausea, vomiting, weight loss, abdominal pain, and diarrhoea are often present. GI bleeding can result from ischaemic damage or rupture of a mesenteric aneurysm. Patients may uncom- monly present with small-bowel obstruction from stricturing dis- ease, which can be difficult to distinguish from NSAID-induced enteropathy or Crohn’s disease. More severe disease is suggested by intussusception, perforation, or infarction. Ischaemic hepatitis, oe- sophagitis, gastritis, pancreatitis, appendicitis, and cholecystitis may also occur. In patients with a pre-existing diagnosis of systemic vasculitis or connective tissue disorder, presentation with symptoms in keeping with acute or chronic mesenteric ischaemia, but without an obvious source of embolism, should prompt further investigation for GI involvement. Patients not known to have systemic disease In the absence of an existing diagnosis of vasculitis or connective tissue disease, when should clinicians consider vasculitis of the GI tract as a cause for abdominal symptoms? Unexplained abdom- inal pain, diarrhoea, bleeding, weight loss, and anorexia with or without fevers should prompt the usual investigations for these symptoms, including laboratory tests, abdominal imaging, and en- doscopy. If these investigations fail to reveal a diagnosis, the less common differentials such as occult malignancy, occult infection, or intestinal vasculitis should be entertained. Abdominal symptoms occurring with other features of systemic vasculitis or connective tissue disease may provide the critical clinical clue. Occasionally, clinicians may encounter a patient who has had multiple courses of empiric corticosteroids for relief of GI symptoms without careful exclusion of

intestinal vasculitis. Primary vasculitides Any systemic vasculitis can theoretically affect the mesenteric circulation. There is variable GI tract involvement depending on the specific primary vasculitis involved. Table 15.17.1 outlines the various vasculitides, the frequency of GI tract involvement, and associated GI features. Vasculitides affecting only the large to medium-sized arteries such as giant cell arteritis and Takayasu's ar- teritis have low rates of clinically significant GI tract involvement due to the marked redundancy of the mesenteric circulation. In general, GI involvement in primary vasculitis indicates more severe disease and poorer prognosis.

 In two types of vasculitis, intestinal involvement is reflected in the American College of Rheumatology (ACR) diagnostic criteria. ACR criteria for polyarteritis nodosa include weight loss, which is prob- ably due to common involvement of the superior mesenteric artery. ACR criteria for Henoch–Schönlein purpura include postprandial abdominal pain or bowel ischaemia, usually associated with bloody diarrhoea. Rare syndromes of localized small-bowel vasculitis have been reported. Cryptogenic, multifocal ulcerating stenosing enteritis (CMUSE) is a rare localized vasculitis that leads to small-bowel ulcers, strictures, and obstructive symptoms. After bone marrow transplantation, there are case reports of jejunal vasculitis causing protein-losing enteropathy. Secondary vasculitis related to collagen vascular diseases Mesenteric vasculitis should be considered in patients with collagen vascular diseases and GI symptoms (Table 15.17.2). Patients with these conditions often have GI symptoms such as abdominal pain, diarrhoea, dyspepsia, dysphagia, and gastro-oesophageal reflux, but it is important to note that most GI symptoms in these patients are due to causes other than mesenteric vasculitis. In fact, possibly the most common cause of GI symptoms and pathology in patients with collagen vascular disorders is as a complication of treatment. NSAIDs are commonly used and lead to many GI manifestations including peptic ulcer disease, enteropathy, intestinal strictures, and secondary vasculitis. Prednisolone, also commonly used to treat these conditions, may contribute to peptic ulcer disease or GI infections. GI symptoms may result from a process related to the primary collagen vascular disorder itself. For example, dysmotility in sys- temic sclerosis due to overproduction of collagen and subsequent fibrosis may cause dysphagia, delayed gastric emptying, colonic in- ertia, or small intestinal stasis with bacterial overgrowth. Mesenteric vasculitis in systemic lupus erythematosus is relatively uncommon (2–15%) but has a high mortality rate. The Systemic

Table 15.17.1 Gastrointestinal involvement of primary vasculitides

Disease	GI tract features	Approximate frequency of GI involvement
Polyarteritis nodosa	Small bowel and gallbladder most commonly affected. Rarely complicated by perforation and infarction. Association with hepatitis B. Other: cholecystitis, appendicitis, pancreatitis	30–60%
Henoch–Schönlein purpura	May present with acute abdomen without typical palpable purpura. GI bleeding is common. Small bowel most frequently involved. Rarely complicated by intussusception, perforation, and infarction. Other: gallbladder hydrops, pancreatitis	50–70% (5–20% at disease presentation)
Antineutrophil cytoplasmic antibody associated: Granulomatosis with polyangiitis (formerly known as Wegener's granulomatosis)	GI tract ulceration (oesophageal, intestinal). Less commonly: infarction, cholecystitis, pancreatitis	5–20%
Eosinophilic granulomatosis with polyangiitis (formerly known as Churg–Strauss syndrome)		30–45%
Microscopic polyangiitis		5–10%
Degos's disease	Peritonitis secondary to bowel perforation is the most common cause of death	50%
Behçet's disease	Ileocaecal region most frequently involved. Also can affect oesophagus, transverse and ascending colon. Can mimic inflammatory bowel disease with mucosal ulceration, and may also have similar extraintestinal findings. Other: pancreatitis	5–25%
Kawasaki's disease	5% have acute abdomen at initial presentation. May	

present with paralytic ileus, gallbladder hydrops, appendicitis, or intestinal ischaemia 5–10% Buerger’s disease – Case reports Giant cell arteritis – Case reports Takayasu’s arteritis Mesenteric vessels involved in up to 12% on angiography but only rarely symptomatic Case reports Cryoglobulinaemic vasculitis Association with hepatitis C Case reports Table 15.17.2 Gastrointestinal manifestations and mesenteric vasculitis in collagen vascular disorders Disease Approximate frequency
of mesenteric vasculitis Common GI symptoms (from disease or side effects of medications) Rheumatoid arthritis 0.5–1% Heartburn, dysphagia, diarrhoea Systemic lupus erythematosus 0.2–10% Abdominal pain, nausea, vomiting, anorexia in 30–50%. Relatively common: heartburn and diarrhoea. Less frequent: dysphagia, abdominal pain, distension due to ascites, and GI bleeding Systemic sclerosis Rare Most frequent: progressive dysphagia. Heartburn, stricture formation, poor gastric emptying, bacterial overgrowth causing diarrhoea and malabsorption, progressive constipation due to impaired colonic motility Sjögren’s syndrome Rare Symptoms relatively rare: abdominal pain, nausea, constipation, diarrhoea, and malabsorption

section 15 Gastroenterological disorders 3004 Lupus International Collaborating Clinics (SLICC)/ACR Damage Index includes scores for mesenteric insufficiency and intestinal infarction or resection. Acute mesenteric ischaemia in systemic lupus erythematosus may be caused by either thrombosis related to antiphospholipid antibodies or mesenteric vasculitis. This distinc- tion is important as treatment will be with anticoagulation rather than immunosuppression. Investigation It is preferable to investigate vasculitis affecting the GI tract prior to specific treatment being instituted, although this is sometimes not possible. Acute presentation with severe abdominal pain should be investigated as described previously. Investigations including inflammatory markers, serological markers of autoimmunity, and angiography may be helpful. The erythrocyte sedimentation rate is almost always elevated in active vasculitis and, although it lacks specificity, may be used as an initial screening test and to monitor progress. In the presence of mesenteric ischaemia, serum lactate is often raised, although this is neither sensitive nor specific enough to be used for diagnosis. Other systemic features of vasculitis may be found with careful clinical examination and basic investigations such as chest radiography and urinalysis. Screening with mesenteric angiography or CT angiography is the most revealing test and should be performed if there is significant clinical concern. However, the findings of different vasculitides can overlap considerably and hence this test rarely determines the spe- cific diagnosis in isolation. Up to two-thirds of cases of polyarteritis nodosa have recognizable aneurysms of mesenteric and renal vessels on mesenteric angiography (Fig. 15.17.8). Although multiple mesen- teric circulation microaneurysms are a characteristic of polyarteritis nodosa, these can be found in other vasculitides. Furthermore, fibromuscular dysplasia may have similar angiographic appearances. CT angiography can help exclude thrombotic or embolic causes of mesenteric ischaemia. Up to 12% of patients with Takayasu’s arteritis (medium and large vessels affected) will have findings of mesenteric involvement on angiography, but most will not have symptomatic intestinal vasculitis. In systemic lupus erythematosus, CT angi- ography may show bowel wall thickening, dilated loops, ascites, lymphadenopathy, and dilated mesenteric veins. Intestinal tissue biopsy has a low sensitivity in diagnosing GI tract vasculitis and tissue diagnosis can usually be made from alternative, more accessible sites. Management Treatment of vasculitis affecting the GI tract depends on the underlying systemic disease. General treatment measures such as bowel rest, total parenteral nutrition, antibiotics, and corticoster- oids are often required. Corticosteroids at high doses are effective for all vasculitides. Other treatments such as cyclophosphamide, azathioprine,

methotrexate, immunoglobulins, and biologics de- pend on the specific diagnosis. Early involvement of senior surgical colleagues is warranted when patients present acutely as with acute mesenteric ischaemia. Patients with stricturing disease may also re- quire surgical resection. Vascular lesions of the intestine Vascular lesions of the intestine are not uncommon and may be con- genital or acquired. They may present at any age, although some le- sions are more likely in certain age groups (e.g. caecal angiodysplasias in elderly patients). This section focuses on primary vascular lesions of the intestine and does not cover the many disorders which may secondarily affect blood vessels of the GI tract, such as peptic ulcers, diverticular disease, aortoenteric fistulas, malignancy, and portal hypertensive bleeding. The clinical presentation of vascular lesions of the intestine is varied and includes acute, sometimes life-threatening haemorrhage, chronic iron deficiency anaemia, and obstruction. If the lesions are part of an underlying disorder, there may be other associated clin- ical features (e.g. epistaxis in patients with hereditary haemorrhagic telangiectasia). Investigation is often difficult, especially when lesions are not ac- cessible via gastroscopy or colonoscopy. In such cases, video capsule endoscopy, CT angiography, and MRA can help identify the location of haemorrhage. Double-balloon enteroscopy, radiographic embol- ization, or surgery may be required to treat lesions inaccessible by routine gastroscopy, enteroscopy, or colonoscopy. In acute haemorrhage, urgent endoscopy is required, with haemo- stasis achieved through a combination of one or more strategies, including injection of adrenaline, banding, clipping, and application of heat or nonthermal coagulation methods such as argon plasma coagulation. If endoscopic treatment is unsuccessful, surgical over- sewing or resection is required. Angiodysplasias Angiodysplasias (alternative names include angioectasias, vascular dysplasias, and vascular ectasias) are acquired arteriovenous mal- formations. They are usually small degenerative lesions of previous healthy blood vessels and histologically comprise dilated vessels Fig. 15.17.8 Arteriography showing multiple renal artery stenoses and microaneurysms (arrows) with an inferior kidney pole infarction in a patient with polyarteritis nodosa. From Ball GV, Fessler BJ, Bridges, Jr. SL (eds) (2014). Oxford textbook of vasculitis, 3rd edition. By permission of Oxford University Press.

in the mucosa and submucosa. They may be singular or multiple and can occur throughout the GI tract. Up to 80% are found in the caecum and ascending colon, around 15% are in the jejunum and ileum, and the remainder are elsewhere. Angiodysplasias are the most common GI tract vascular abnor- mality, present in around 1% of asymptomatic individuals. There is an increasing prevalence with age, and in patients over 60 years they are second only to diverticular disease as the most common cause of lower GI bleeding. Small-bowel angiodysplasias may account for up to 40% of occult GI bleeding. Bleeding from angiodysplasias is more common in patients on antiplatelet agents or therapeutic anticoagulation, and those who have conditions with a bleeding diathesis such as chronic renal in- sufficiency and end-stage liver disease. There is a higher rate of GI bleeding from angiodysplasias in the presence of aortic stenosis (Heyde’s syndrome), although the cause of this remains uncertain. Presentation may be with iron deficiency anaemia or acute lower GI haemorrhage, particularly from right-sided colonic lesions. Direct visualization of the intestinal mucosa with endoscopy or video capsule endoscopy are the investigations of choice. The classic appearance is of one or more small (<10 mm), cherry red, arborizing lesion(s) (Fig. 15.17.9). In the acute setting, CT, MRA, or standard angiography may demonstrate a bleeding point (Fig. 15.17.10). If incidentally found on endoscopy, there is no indication to treat these lesions. If, however, the lesion is thought to be the cause of symptoms, treatment with direct application of heat or nonthermal coagulation

is often sufficient. Argon plasma coagulation is gaining favour as the modality of choice. Some patients may require mul- tiple endoscopic procedures to treat difficult lesions. Local resection or arterial embolization may be necessary if lesions are inaccessible via endoscopy, too widespread, or resistant to repeat endoscopic therapy. Arterial embolization of the small bowel carries a substan- tial risk of intestinal ischaemia. Pharmacological treatments with thalidomide or octreotide are reserved for transfusion-dependent and treatment-resistant cases. Telangiectasias Telangiectasias are dilated venules and most commonly occur in the GI tract as part of hereditary haemorrhagic telangiectasia (also known as Osler–Weber–Rendu disease). Hereditary haemorrhagic telangiectasia is an uncommon, multisystem, autosomal dominant disease with telangiectasias and arteriovenous malformations affecting the nasal passages, skin, GI tract, lungs, liver, and brain (Fig. 15.17.11). After epistaxis, GI haemorrhage is the most frequent form of bleeding, occurring in up to one-third of these patients. Gut telangiectasias are most commonly found in the stomach and prox- imal small bowel. Presentation may be with iron deficiency anaemia Fig. 15.17.9 Small-bowel enteroscopy showing angiodysplasia. From Jolly E, Fry A, Chaudhry A (eds) (2016). Training in medicine. By permission of Oxford University Press. (a) (b) (c) Fig. 15.17.10 (a) Angiodysplastic lesion in the caecum: superior mesenteric angiogram in a 53-year-old man with anaemia for 20 years (no lesion found at previous operations). Vascular lake in caecum (arrowed). (b) Angiodysplastic lesion in the caecum: superior mesenteric angiogram in a 53-year-old man with anaemia for 20 years (no lesion found at previous operations). Capillary phase, showing early filling vein arising from lesion. (c) Angiodysplastic lesion in the caecum: superior mesenteric angiogram in a 53-year-old man with anaemia for 20 years (no lesion found at previous operations). Injected specimen magnified ×30. Courtesy of Dr D J Allison, Royal Postgraduate Medical School; previously published in Br J Hosp Med (1980), 23, 358.

 or overt GI bleeding. Typically, there are numerous lesions making endoscopic therapy difficult. Often regular cauterization is required. Haemangiomas Intestinal haemangiomas are uncommon, benign vascular lesions that occur as single or multiple lesions anywhere in the GI tract. They most commonly occur in the small bowel, and in particular the jejunum. The rectosigmoid is the most common colonic site. When occurring as multiple lesions, haemangiomas may occur as part of a syndrome (Table 15.17.3). Patients may present at any age with oc- cult or overt GI haemorrhage, obstruction, or intussusception. Haemangiomas have solid characteristics of tumours and are classified as cavernous, capillary, or mixed. The capillary subtype is a proliferation of capillaries surrounded by a lining of endothelial (a) (b) (c) Fig. 15.17.11 Hereditary haemorrhagic telangiectasia. (a) Characteristic tiny dilated vascular lesions on the cheek in a middle-aged woman. (b) Red maculopapular lesions on tongue and lower lip in a 55-year-old woman. (c) Endoscopic image of right nasal cavity showing telangiectasias on the anterior septum and inferior turbinate in a 59-year-old man. From Mulliken JB, Burrows PE, Fishman SJ (eds) (2013). Mulliken and Young’s vascular anomalies: hemangiomas and malformations. By permission of Oxford University Press. Table 15.17.3 Syndromes associated with GI vascular lesions Syndrome Vascular lesion in the GI tract Most common location of GI involvement Turner’s syndrome Telangiectasia Angiodysplasias Haemangiomas Small bowel Hereditary haemorrhagic telangiectasia Telangiectasias Stomach and proximal small bowel Blue rubber bleb nevus syndrome Haemangiomas Small bowel Klippel–Trenaunay and Parkes–Weber syndromes Haemangiomas Distal colon Systemic sclerosis and CREST Telangiectasias Anywhere throughout the GI tract; may have GAVE in the stomach Peutz–Jeghers syndrome Hamartomatous polyposis Jejunal and ileal

15.17 Vascular disorders of the gastrointestinal tract 3007 cells and occurs as single lesions, predominantly in the perianal skin but also the appendix and small bowel. Cavernous haemangiomas comprise large blood-filled cavities lined by endothelial cells, and present the greatest clinical risk as they are generally larger, have a higher propensity to bleed, and may consume platelets leading to thrombocytopenia. The endoscopic appearance of the cavernous type is as a sessile, polypoid lesion which can be distinguished from adenomatous polyps by their bluish colour, poorly defined margins, and typically submucosal location. Endoscopic therapy or angiographic embolization may arrest bleeding. Dieulafoy's lesions A Dieulafoy's lesion represents a small submucosal defect overlying a large-calibre and tortuous arteriole. It is the reported cause of 1 to 2% of cases of acute GI bleeding. Diagnosis can be a challenge because identification often relies on stigmata of recent bleeding or active bleeding during endoscopy (Fig. 15.17.12), and 'missed' Dieulafoy's lesions are likely to be the cause of many cases of un- identified GI bleeding. Most lesions occur in the upper stomach but they may occur anywhere along the GI tract. Advances in endoscopy have dramatically reduced mortality from these lesions, with pro- cedures successful in over 90% of cases. Gastric antral vascular ectasia Gastric antral vascular ectasia (GAVE) is an incompletely under- stood condition that causes up to 4% of cases of GI bleeding. The characteristic endoscopic appearance is of erythematous stripes along the antral rugal folds which represent ectatic vessels (thus the alternative name, 'watermelon stomach') (Fig. 15.17.13). GAVE is associated with several comorbid conditions such as primary biliary cholangitis, systemic sclerosis and renal failure. The treatment of choice is endoscopic cautery, usually with argon plasma coagulation and often requiring multiple treatments. Pharmacological treat- ments such as oral oestrogen and progesterone, octreotide, tranex- amic acid, and thalidomide have been reported in small series and should be reserved for transfusion-dependent patients. Resistant disease may require surgical antrectomy. FURTHER READING Bala M, et al. (2017). Acute mesenteric ischemia: guidelines of the World Society of Emergency Surgery. World J Emerg Surg, 12, 38. Garcia-Porrua C, et al. (2006). Localised vasculitis of the gastrointes- tinal tract. Seminar Arthritis Rheum, 35, 403–6. Lara LF, et al. (2010). Dieulafoy lesions of the GI tract: localization and therapeutic outcomes. Dig Dis Sci, 55, 3436–41. Oglat A, Quigley EM (2017). Colonic ischemia: usual and unusual presentations and their management. Curr Opin Gastroenterol, 33, 34–40. Thomas A, et al. (2018). An analysis of the clinical, laboratory, and histological features of striped, punctate, and nodular gastric antral vascular ectasia. Dig Dis Soc, 63, 966–73. van Dijk LJ, van Petersen AS, Moelker A (2017). Vascular imaging of the mesenteric vasculature. Best Pract Res Clin Gastroenterol, 31, 3–14. Fig. 15.17.12 Endoscopic image of a spurting Dieulafoy's lesion. Reproduced from www.gastrointestinalatlas.com. Fig. 15.17.13 GAVE. Note the linear aggregates of red markings in the antrum and the absence of an underlying mosaic-like pattern. From Hauser SC (ed) (2014). Mayo Clinic gastroenterology and hepatology board review, 5th edition. By permission of Oxford University Press.

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