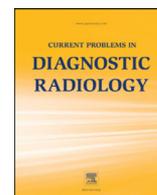




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Multimodality Imaging Review of Anorectal and Perirectal Diseases with Clinical, Histologic, Endoscopic, and Operative Correlation, Part II: Infectious, Inflammatory, Congenital, and Vascular Conditions

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A broad spectrum of pathology affects the rectum, anus, and perineum, and multiple imaging modalities are complementary to physical examination for assessment and treatment planning. In this pictorial essay, correlative imaging, endoscopic, pathologic, and operative images are presented for a range of rectal, perirectal, and perineal disease processes, including infectious/inflammatory, traumatic, congenital/developmental, vascular, and miscellaneous conditions. Key anatomic and surgical concepts are discussed, including radiological information pertinent for surgical planning, and current operative approaches of these anatomic spaces to assist radiologists in comprehensive reporting for gastroenterologists and surgeons.

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Introduction

Acute and chronic conditions affecting the anus, rectum, and perineum are often clinical diagnoses in adults. Imaging may play a role in surgical planning and diagnosis for those entities that are not readily apparent on physical exam. With the expanding use of computed tomography (CT) in the acute setting, such acute anorectal and perineal conditions may be encountered on imaging. Magnetic resonance (MR) imaging offers superior spatial resolution and is used in selected conditions to delineate anatomy and extent of disease, such as anorectal fistula, which may not be readily apparent on physical exam. In pediatrics, imaging plays an obvious role in delineating aberrant anatomy that may need surgical correction.¹⁻³ In all cases, imaging may be used initially to direct proctoscopy, endoscopy, or surgical management or these conditions may be seen incidentally. A review of infectious, inflammatory, and vascular conditions such as perirectal abscess, Fournier gangrene, rectal manifestations of inflammatory bowel disease, vascular malformation, among others, is presented. Congenital anomalies and conditions that present in infants and adults are presented as well. The purpose of this pictorial essay is to describe imaging findings of various anorectal and perineal conditions and provide endoscopic and surgical correlation when applicable.

Infectious and Inflammatory—Anorectal Abscesses and Fistulae

Purulent collections or fistulous tracts can occur in the potential spaces around the rectum and anus (Fig. 1a). Anorectal abscesses and

fistulae represent a spectrum of disease: the two entities may occur independently or in conjunction with each other. The majority (approximately 90%) of anorectal abscesses and fistulae result from obstruction of an anal gland and extend from a crypt at the dentate line between the anal sphincters to the perianal skin (intersphincteric).⁴

Anorectal abscesses usually present as a fluctuant area, painful area apparent on physical exam. However, the immunocompromised or patients with Crohn disease may have atypical presentations that may be indolent and cause a systemic inflammatory response. CT or magnetic resonance imaging (MRI) may be useful in such atypical cases, when there is a suspected fistulous tract, or to assess the extent of abscess (Fig. 1c-d). Caliste et al.⁵ reported a retrospective study with 113 patients with surgically drained perirectal abscess who had CT 48 hours before drainage. They found that CT had an overall sensitivity of 77%. The sensitivity was significantly lower in the immunocompromised.⁵ Surgical drainage is indicated for symptomatic anorectal abscesses.

Anorectal fistulae are most commonly single fistulous tracts with a defined course with or without a dependent abscess. As with abscesses, few cases of anorectal fistulae can have atypical presentations and/or trajectories, particularly in patients with Crohn disease or recurrent fistulae (Fig 2). MRI can be helpful in such cases to define fistulous tracts. Beets-Tan et al.⁶ reported a prospective study that imaged patients who underwent surgical probing and examination of anorectal fistulae in conjunction with preoperative MRIs. MRI findings and descriptions of the fistulae were blinded to the operating surgeon and revealed intraoperative after they made their initial assessment with probing the fistula to define their anatomy. The MRI findings provided additional detail to the surgeon in 21% of the 56 patients in their study. These additional findings were more frequently seen in patients with recurrent fistulae and patients with Crohn disease. Morris et al.⁴ describe a grading system for perianal fistulae: (i) grade 0 implies no fistula, (ii) grade 1 is a simple

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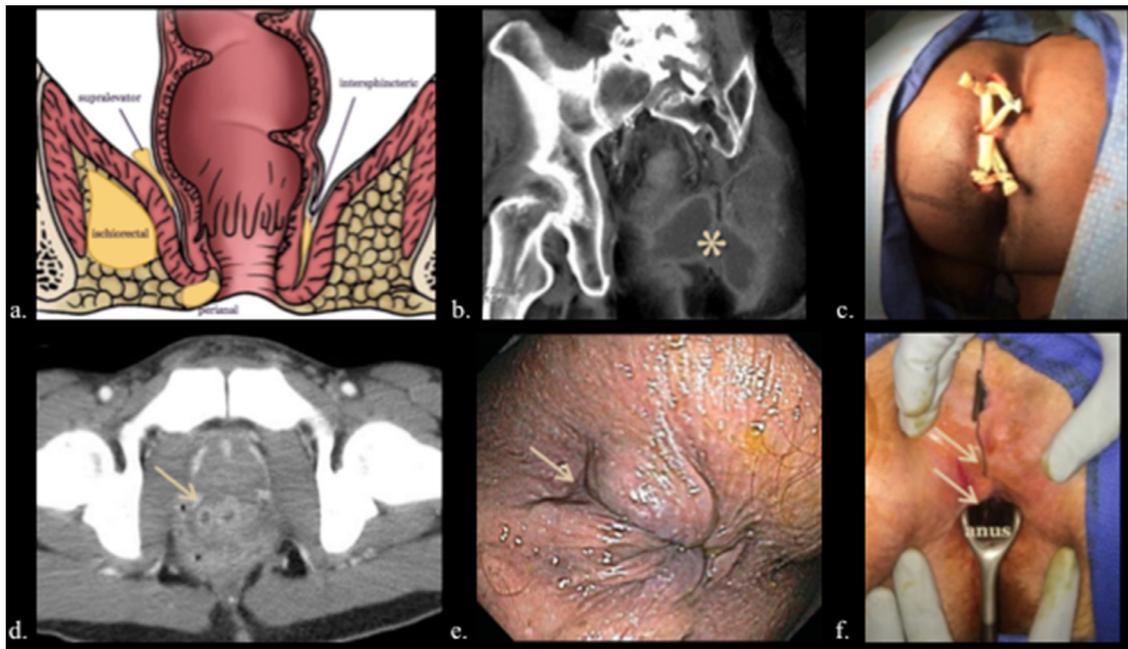


FIG 1. (a) Illustrative diagram of the anorectal spaces where abscesses can accumulate. (b) A 53-year-old male with clinical signs of sepsis. CT demonstrates a large abscess (*) involving the ischioanal fossa and gluteal musculature. (c) Surgical drains in place post abscess incision and drainage. (d-f) Three patients with perianal pain. (d) CT illustrates a small rim enhancing fistulous tract between the right internal and external anal sphincters. (e) A chronic perianal fistula manifests as a "dimple" separate from the anal orifice. (f) A perianal fistula is defined prior to surgery by probing its course (arrows). CT, computed tomography.

linear intersphincteric fistula, (iii) grade 2 is an intersphincteric fistula with intersphincteric abscess or secondary fistulous track, (iv) grade 3 is a trans-sphincteric fistula with no associated abscess or secondary tract, (v) grade 4 is a trans-sphincteric fistula with an abscess or secondary tract, (vi) grade 5 indicates supralelevator and translevator disease.⁴

Asymptomatic, nondraining fistulae can be observed if abscess and sepsis have been excluded. Associated abscesses require drainage. There are a number of surgical approaches. The approach is influenced by the location and length of fistula tract and relationship to anal sphincters, the risks of incontinence and recurrence, and the comfort level of the surgeon with the various techniques. Surgical techniques the treatment of fistulae includes: placement of and serial tightening of setons (Fig 2), opening the tract with fistulotomy,

removing the tract with fistulectomy, flap advancements with anterior fistulae in females, and fibrin glue injection.^{7,8}

Infectious and Inflammatory—Hidradenitis Suppurativa

Hidradenitis suppurativa (HS) is a chronic disorder of the skin affecting predominantly the axilla in both sexes and the perineal and perianal region in females. Once thought to be an infection or inflammatory of apocrine glands, the favored pathogenesis is now follicular occlusion.⁹ Hidradenitis can be a debilitating disorder that requires multiple incision and drainages. Often apparent on physical exam, HS manifests as chronically indurated skin with sinus tracts that may accumulate and drain purulent collections. CT or MRI demonstrates tracts and inflammatory

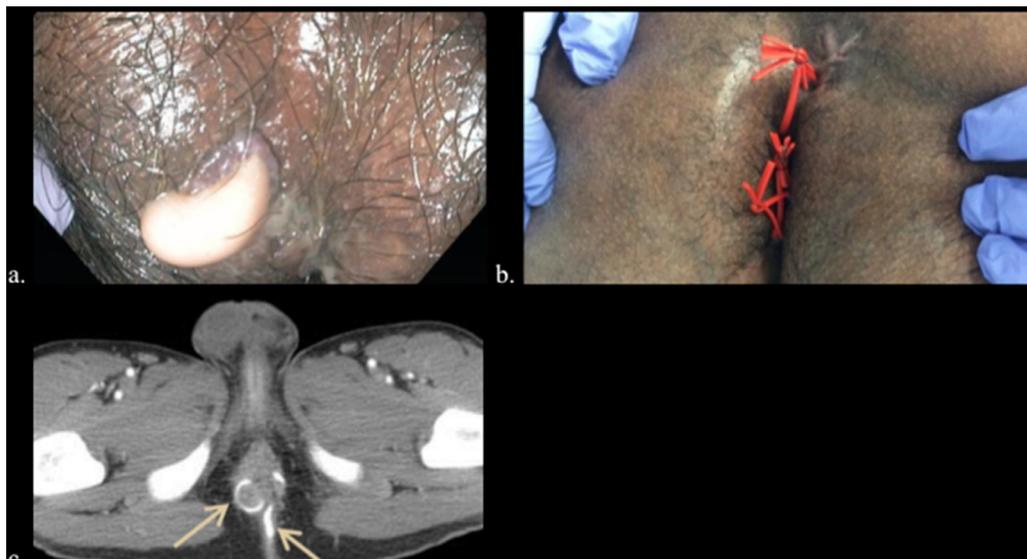


FIG 2. A 27-year-old male with Crohn disease and perianal pain and drainage. (a) A perirectal abscess and perianal fistulae draining purulent material is apparent on physical exam. (b, c) The abscess was surgically drained and setons (arrows) placed in the fistulae.

fluid collections and can evaluate the extent of disease which aids in surgical planning (Fig 3).⁹⁻¹² Treatment begins with counseling and supplementation of hygiene techniques, antibiotics with or without incision and drainage for flairs ups. Some patients benefit from long-term antibiotics. Surgery with wide local excision can be curative or palliative.

Griffin et al.⁹ reported the MR findings of 18 patients with HS. In that study, MR demonstrates multiple subcutaneous tracts in nearly all patients (17 of 18) with the most frequent sites of disease being the intergluteal cleft (16 of 18), perianal area (12 of 18), and perineum (13 of 18). Edema in soft tissue adjacent to areas of heavy disease (most often gluteal) was a common feature and myositis of the gluteus maximus was an infrequent finding (3 of 18). Patients had concurrent Crohn disease in 4 patients and perianal fistulae were present in 4 patients, only 1 of whom had Crohn disease. Given the potential overlap perineal and perianal disease in with HS and perianal Crohn disease, Monnier et al.¹³ reported a retrospective series in which they compared the pelvic MR appearance of 23 patients with HS without concurrent Crohn disease and 46 patients with perianal and/or perineal manifestations of Crohn disease. They found that patients with CD were significantly younger (mean age 38 years vs 49 years for HS), had significantly more fistulae, sphincter involvement, rectal wall thickening, and had a predominance in the perianal region (compared to a perineal or intergluteal cleft predominance in HS). Given this, they proposed on MR that HS could be distinguished from CD by meeting the following three criteria (which had a specificity of 100% in their series for HS): absence of predominant disease in the perianal area, absence of rectal wall thickening, and bilateral disease.

Infectious and Inflammatory—Crohn Disease

Crohn disease is a multisystem chronic inflammatory disease characterized by transmural inflammation of the gastrointestinal tract. Perianal manifestations occur in 10%-20%. Treatment is aimed

at primary prevention, with 5-aminosalicylic acid, immunomodulators, and selected steroid and antibiotic therapy for flare-ups. Surgery is reserved for medically refractive disease, or complications such as bowel obstruction and perforation. Perianal abscesses and fistulae are by far the most common anorectal complications of Crohn disease. Either may occur in up to 60%-80% of patients.^{14,15} Imaging and treatment principle are discussed in the previous section. Other anorectal manifestations include anal and rectal strictures, hemorrhoids, and deep pelvic abscess (Fig 4). Enterography and CT with rectal contrast can be helpful to characterize strictures.¹⁶ MR is helpful in characterizing perianal manifestations. In the previously mentioned study by Monnier et al.¹³ comparing MR feature of perianal CD to that of hidradenitis supportive, patients with CD demonstrated sinus tract in all (100%; 46 of 46) patients, perianal predominance in 80% (37 of 46), frequent (74%; 34 of 46) involvement of the intergluteal cleft, unilateral burden of disease in 74% (34 of 46), and a concurrent abscess in 39% (39 of 46). Management ranges from observation to surgical excision. Deep pelvic abscesses can occur from subacute to chronic fistulae or bowel perforation. These abscesses can often be drained percutaneously, transrectally or through other routes.^{17,18} Select abscess-fistula complexes may benefit from percutaneous catheter cannulation.¹⁹

Infectious and Inflammatory—Ulcerative Colitis

Ulcerative colitis (UC) is an inflammatory state of the colon and rectum that predisposes patients to colonic complications and a greatly increased incidence of colorectal cancer. Whereas the transmural bowel inflammation in Crohn disease may spare the rectum, UC involves the rectum in a continuous mucosal inflammatory state of the colon and rectum. On CT and MR, mural thickening may be slightly greater in Crohn disease compared to UC. The mural thickening will tend to be circumferential and

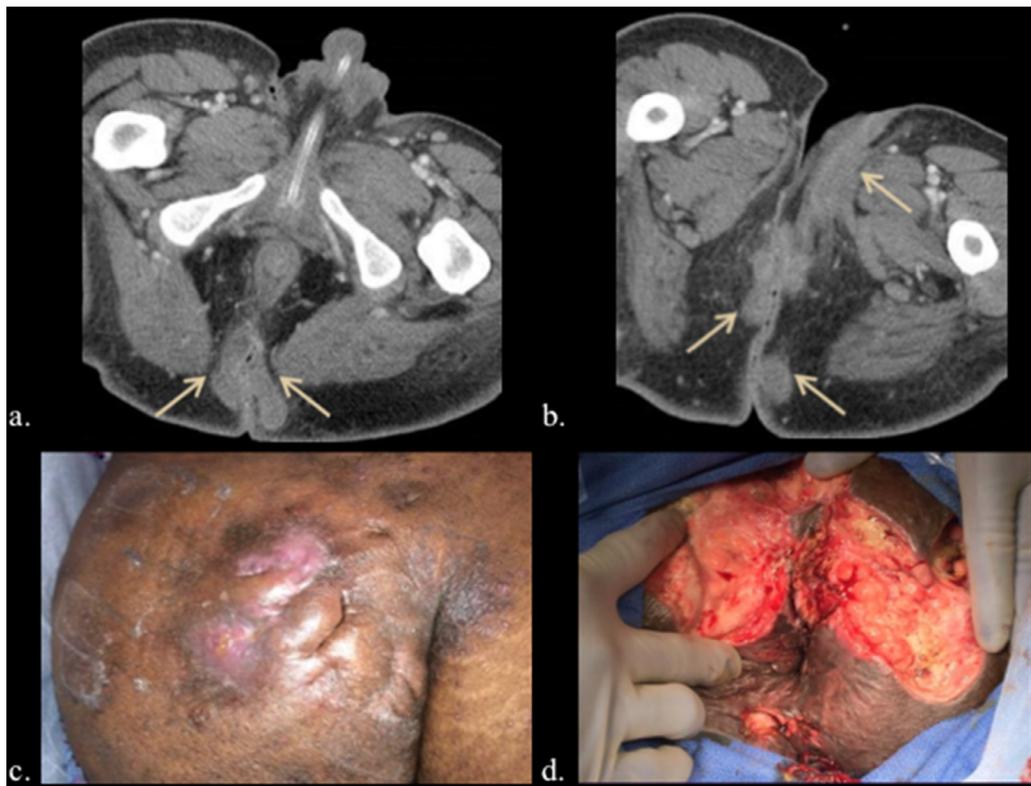


FIG 3. A 26-year-old male with clinical evidence of hidradenitis suppurativa. (a, b) CT demonstrates inflammatory fluid collections in the perianal, ischiorectal, gluteal subcutaneous, and left inguinal regions (arrows). (c, d) Clinical photographs of hidradenitis suppurativa pre-excision and right postexcision of affected tissue.

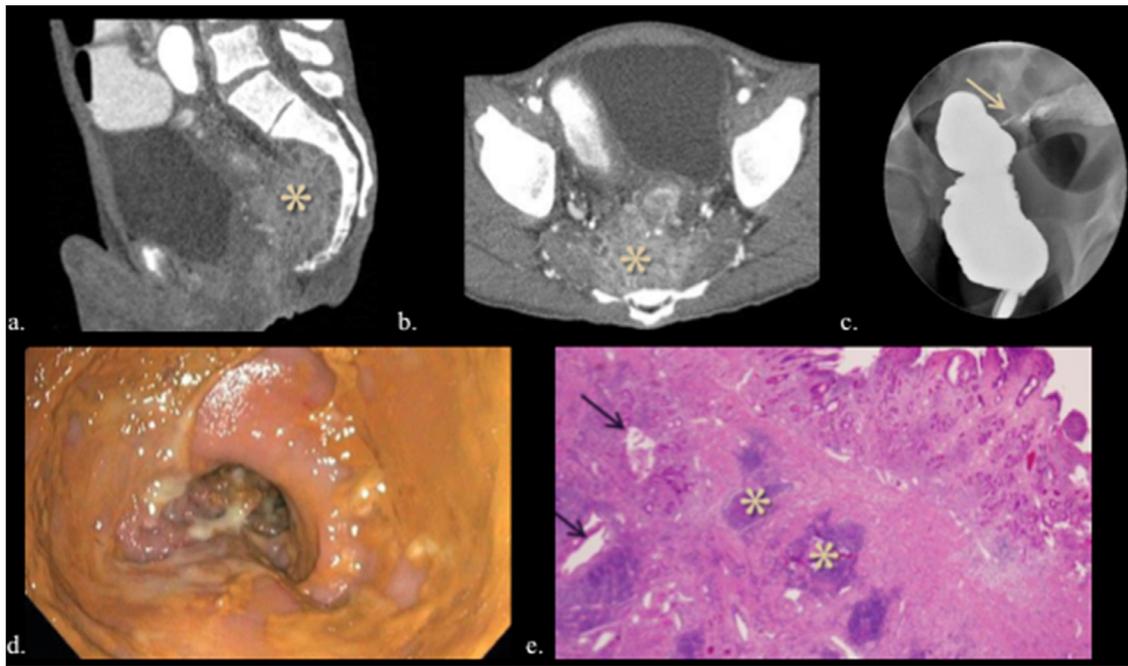


FIG 4. A 33-year-old male with Crohn disease who was initially evaluated after 3 years of abdominal pain and constipation. (a, b) Multiplanar CT images showing extensive presacral and perirectal inflammatory tissue (*). (c) Barium enema demonstrates sinus tracts, and strictures of the ileum and rectosigmoid (arrow). (d) At colonoscopy, the rectal mucosa was friable with purulent exudate (arrows). Disease was unresponsive to immunosuppressive therapy and patient required surgical resection. (e) Specimen demonstrated foci of chronic inflammation, lymphoid aggregates (*), and fistulous tracts (arrow). CT, computed tomography.

diffuse in UC whereas it may be focal, spotty, and eccentric in Crohn disease.¹⁶ In patients with controlled disease, there are typically no clinically significant findings on CT. Gashin et al.²⁰ retrospectively reviewed the CT findings 163 patients with UC who presented to the emergency room, including 110 patients without prior colectomy. CT showed no clinically significant findings in 103 patients (63%). Further, there were only 3 patients (2%) with clinically significant findings directly related to UC (abscess and large bowel obstruction). Other findings, such as small bowel obstruction and ileus, were more common in patients who had a prior colectomy (16 [10%] vs 5 [3%] patients). Barium enema is typically superior to CT in detecting mucosal abnormalities of the colon and rectum (Fig 5). One treatment difference between Crohn disease and UC is that total proctocolectomy is potentially curative in UC—this is in contrast to Crohn disease, where surgery is avoided when possible, due to high rate of recurrence and that disease may occur anywhere along the gastrointestinal tract.^{16,21,22}

Traumatic—Rectovaginal Fistula

Rectovaginal fistulae may result following vaginal trauma during childbirth or secondary to cryptogenic anal gland infection, Crohn disease, or prior radiation exposure. Clinical presentation includes passage of flatus and stool per vagina or frequent urinary tract infections. A number of imaging modalities can demonstrate the fistulous tract, including transvaginal ultrasound (US), fluoroscopic exams, CT, or MRI (Fig 6). Vaginography is more sensitive in detecting rectovaginal fistula compared to water soluble or barium enema lower gastrointestinal studies.^{23,24} In one study, vaginography had a sensitivity of 79%, demonstrating 19 of 24 fistulae compared to 9% of barium enema in 9 of 11 fistulae.²³ Stoker et al.²⁴ reported the endoluminal US and MR findings of 13 patients who had surgically confirmed or refuted ano- or rectovaginal fistulae. US and MR had respective positive predictive values of 100% and 92% and both had sensitivities of 92%. On CT, air will often be seen within the vagina. On CT and pelvic

MR, sagittal reconstructions or sequences are helpful in delineating disruptions in mucosa to demonstrate the fistula. MR offers better visibility, however, more air-filled tracts may make it difficult to delineate the tract. Fluid sensitive sequences may demonstrate a fluid filled tract.^{25,26} Local surgical repair is appropriate for small fistulae with intact sphincters; larger defects may require interposition flaps and fecal diversion via colostomy may be required for some patients.^{24,26}

Infectious and Inflammatory—Fournier Gangrene

Fournier gangrene (FG) is a necrotizing genitourinary infection of the scrotum and/or labia, perineum, and/or ischioanal fat that is fatal if not promptly diagnosed and surgically debrided. It may develop from an area of previous trauma or low-grade soft tissue infection of the anorectal or genitourinary tract. FG is more frequent in diabetics and the immunosuppressed. A true surgical emergency, FG classically presents with disproportionate pain, crepitus, and systemic signs of sepsis and carries a high mortality of up to 50%. Although diagnosis is often clinical, CT or US is occasionally performed for diagnosis, to assess extent, or to plan the surgical approach. CT and US will often show foci of gas along the extent of the infected tissue (Fig 7).^{27,28} However, the absence of soft tissue gas should not be used to exclude necrotizing fasciitis. Wysoki et al.²⁹ reported a retrospective cohort of CT findings of 20 patients with pathologic-proven necrotizing fasciitis, including 10 on the FG spectrum—6 with a scrotal source and 4 with a perineal source. In these 10 patients with genitourinary necrotizing fasciitis, CT showed fascial thickening in all (10 of 10, 100%), soft tissue gas in 8 (80%), and associated abscesses in 5 (50%). In a more recent imaging cohort of 38 patients with surgically proven FG, nearly all patients (95%; 36 of 38) had fascial air, all patients had subcutaneous edema, and abscesses were less frequent (21%–24%; 8 and 9 of 38 patients for the two study radiologists).³⁰ In that series, there were a higher proportion of women (21 [55%] of the 38 patients were female) than typically reported in FG cohorts.

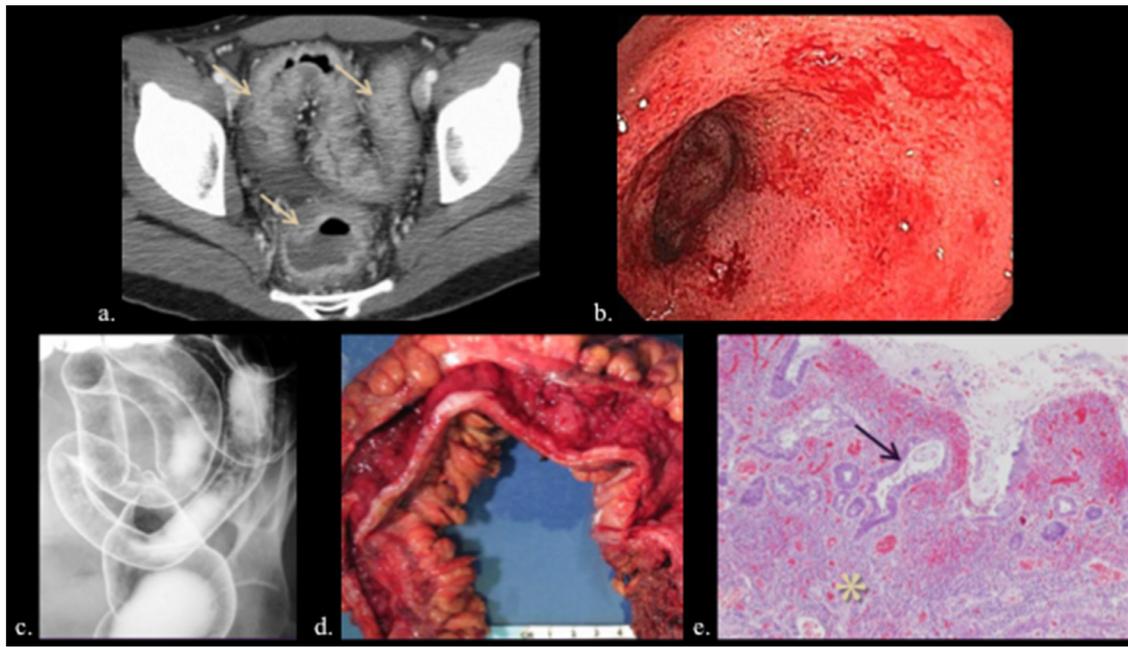


FIG 5. A 29-year-old male with known ulcerative colitis who presented with an acute episode of abdominal pain. (a, b) CT demonstrates contiguous rectosigmoid mucosal thickening and hyper-enhancement (arrows) (a), and colonoscopy confirms gross mucosal hyperemia (b). (c) Barium enema shows the haustral colon with granular mucosa. (d) Proctocolectomy specimen reveals diffuse erythematous friable mucosa, superficial mucosal ulcerations, and pseudopolyps. (e) Marked mucosal inflammation (*), cryptitis, and glandular distortion (arrow) are noted microscopically. CT, computed tomography.

Traumatic—Rectal Injuries

The majority (80%) of rectal injuries are caused by gunshot wounds; 10%–15% are related to knife stab wounds, impalement and foreign body insertion, and a small minority occur secondary to pelvic fractures. Primary repair can be attempted for intraperitoneal injuries, whereas extraperitoneal injuries with often require a diverting ostomy. In otherwise healthy individuals, approximately 75% of these traumatic

rectal injuries will improve after the initial surgery, allowing for restoration of bowel continuity. In the trauma setting, CT is the major modality as it can be obtained quickly, assesses the extent of the primary rectal injury, and assesses for associated injuries (Fig 8).^{31–33} CT with water soluble contrast per rectum can be a problem-solving solution for patients with penetrating abdominal injuries, especially those in which a bullet tract involves the rectum. Routine use is not indicated. One prospective study of 200 trauma patients with penetrating

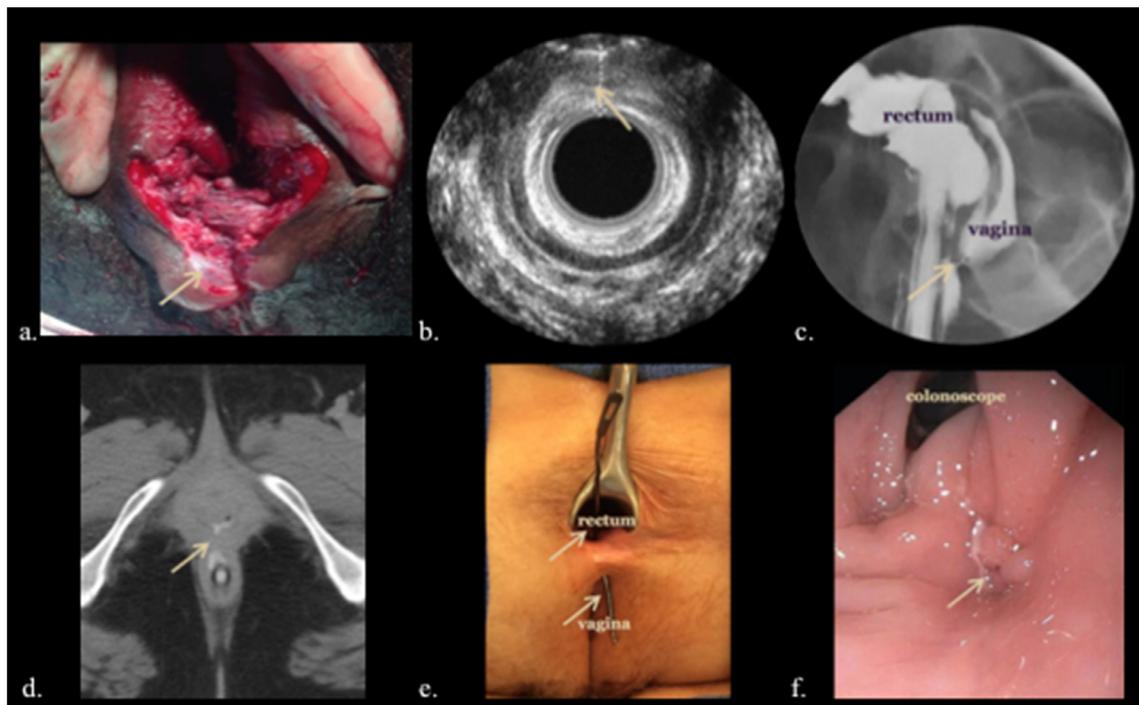


FIG 6. Examples of rectovaginal fistulae. (a, b, c) Perineal body and anal sphincter (arrow) laceration during childbirth (a), chronic disruption of both internal and external anal sphincters (b), and low rectovaginal fistula by barium enema (c). (d–f) CT, operative, and endoscopic depiction of rectovaginal fistula (arrows). Endoscopic appearance is typically that of mucosal puckering or dimpling. CT, computed tomography.

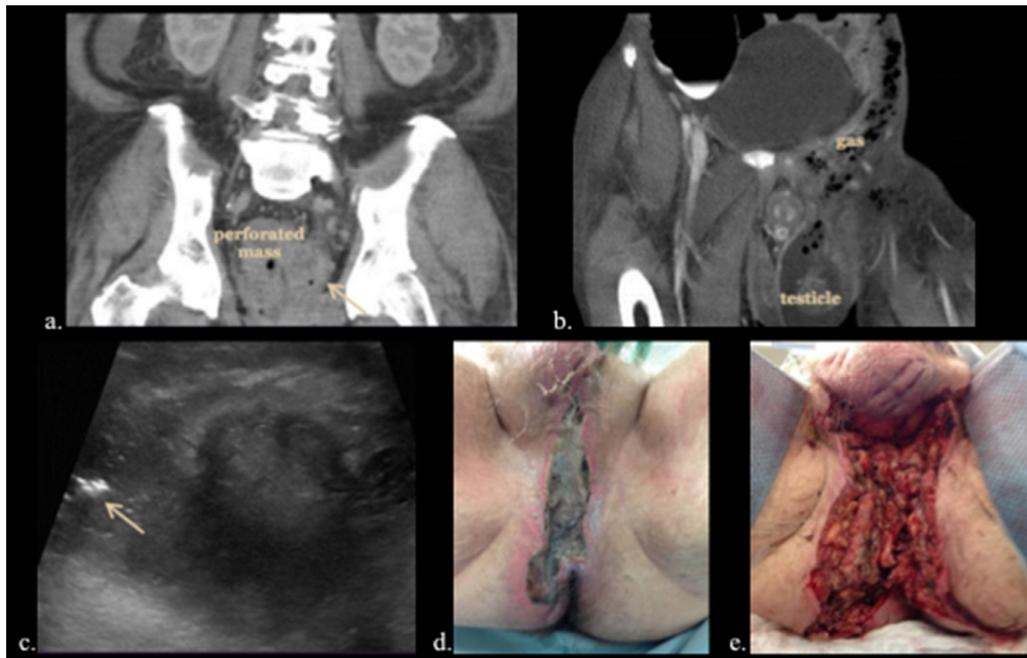


FIG 7. Two male patients presenting with clinical evidence of Fournier gangrene. (a, b) CT images showing soft tissue foci of gas from infections secondary to perforated rectal carcinoma (a, arrow), and of genitourinary origin (b). (c) Superficial ultrasound demonstrates hyperechoic shadowing soft tissue foci consistent with gas (arrow). (d, e) Clinical images illustrating preoperative (d) and postwide surgical debridement (e).

abdominal injuries underwent a “triple-contrast” CT scan (intravenous, per oral, and per rectum) and demonstrated oral or rectal contrast extravasation in only 5 patients (3%).³¹ Treatment varies widely on the modality, location, and extent of the injury. Weinberg et al.³² proposed and validated an anatomic-based surgical approach to penetrating

rectal injuries in which intraperitoneal portions of the rectum are treated with primary repair, accessible extraperitoneal rectal injuries are treated with repair and selective proximal diversion, and inaccessible extraperitoneal injuries to the distal third of rectum are treated with proximal fecal diversion and presacral drainage.³²

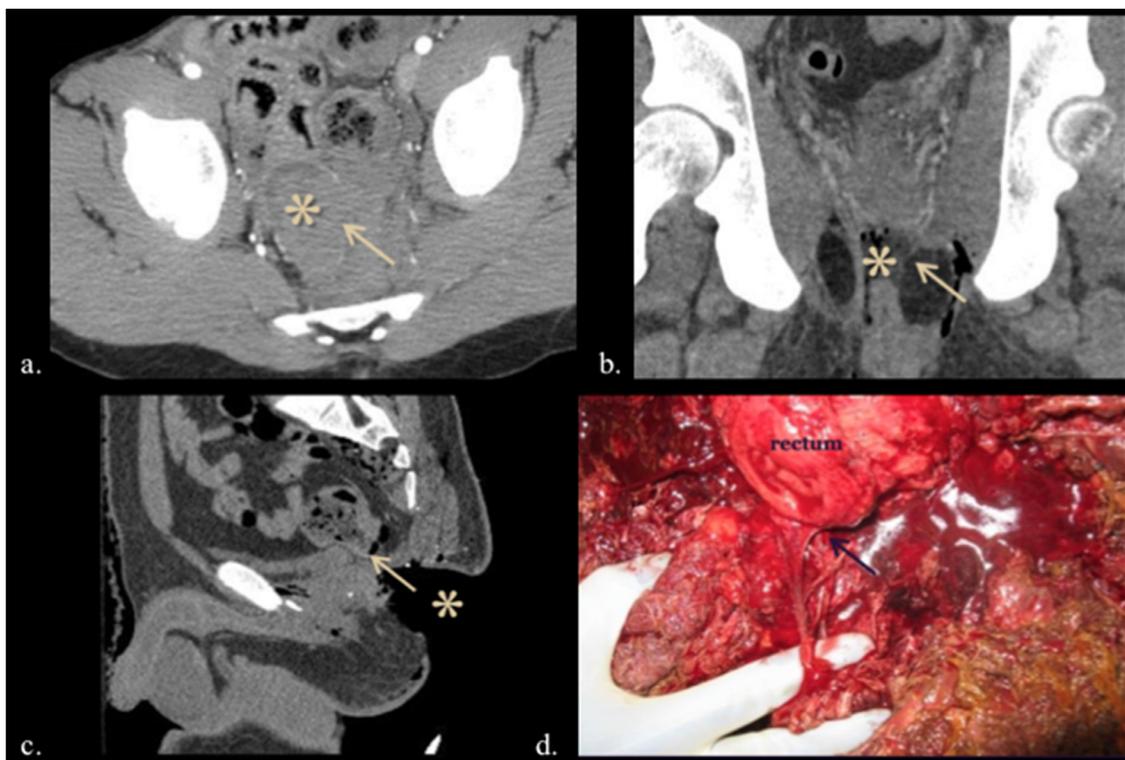


FIG 8. Three different patients with penetrating rectal trauma. (a) A 12-year-old male post gunshot wound. CT demonstrates hematoma (*) in rectal lumen and traversing left rectal wall defect (arrow). (b) A 19-year-old male sustaining impalement anorectal trauma after a 12-foot fall. CT confirms perianal gas (*) and left levator ani defect (arrow). (c, d) A 25-year-old male who sustained blast injury producing a large soft tissue defect (*) and low rectal transection (arrows). All patients underwent diverting colostomy. CT, computed tomography.

Congenital and/or Developmental—Imperforate Anus

Imperforate anus is a spectrum of disorders. The rectum and anus may end in a blind-ending pouch or may fistulize cutaneously or to urinary, reproductive areas. The rectum may end in a blind pouch or have fistulae with other organs. Approximately 95% will have an associated rectal fistula to the perineum, vagina, urethra, or bladder. Prenatal diagnosis is uncommon, 16% by one source.³⁴

Anorectal malformations result from abnormal development of the urorectal septum, a mesenchymal plane of tissue that separate the developing ventral and dorsal hindgut, during development. The ventral portion of the urorectal septum becomes the urogenital sinus, dorsal portion becomes the rectum and anus, and the most distal portion of the urorectal septum becomes the perineum. Associated abnormalities may affect genitourinary, skeletal, and cardiac systems and sacral defects are associated with high lesions.³⁴

There are two classification systems, the Wingspread and Krickenbeck systems. A detailed discussion is beyond that of the current work, but general principles of the classification systems include identifying anorectal malformation as low, intermediate, or high identifying and describing associated fistulae.³⁴ Fluoroscopic voiding cystourethrogram and water soluble enema studies can demonstrate fistulae between the bladder and rectum or other rectal communications. MR can be helpful to delineate the abnormal anatomy (Fig 9).³⁴ Identification of the primary and associated abnormalities is key, as it will guide the pediatric surgeon if early operative manipulation would be helpful. Management is determined by presence and severity of associated anomalies and level of anorectal defect. Primary repair may be performed for lesions at the coccygeal level. Higher and more complicated lesions require fecal diversion and staged repairs.³⁴

Congenital and/or Developmental—Hirschsprung Disease

Hirschsprung disease (HD) is the most common cause of neonatal colonic obstruction. Affecting approximately 1 in 5000 live births the pathogenesis of HD involves failure of ganglion cell precursor migration into the hindgut during 5th–12th week. A spectrum of severity, HD typically manifest as a functional partial colonic obstruction. The length of the aganglionic segment length varies and approximately 80% are rectosigmoid. Most present shortly after birth due to failure to pass meconium, abdominal distention, and vomiting. Delayed diagnosis can present as severe constipation and megacolon and risk of colonic perforation.³⁵

Lower gastrointestinal fluoroscopic studies are the most common modality used to diagnose HD (Fig 10). Water soluble contrast

enemas have been shown to have similar sensitivities to barium. Although the rate of colonic perforation is infrequent, water soluble contrast is preferred if there is a clinical suspicion of perforation. In one study of 38 neonates and infants with bowel dysfunction, HD was the most frequent diagnosis (47% or 18 of 38) followed by functional megacolon and meconium plug syndrome (29% or 11 of 38 and 18% or 7 of 38, respectively).³⁵

Milder forms of HD and adult hypoganglionosis may go undiagnosed in infancy and childhood and present later in adulthood, typically with years of refractory constipation. Kim et al.³⁶ reviewed 10 cases of histologically proven HD and adult hypoganglionosis and identified colonic caliper transition zones in the descending colon in 5 of 10 (50%) or upper rectum and/or rectosigmoid junction in 5 of 10 (50%). In neonates, infants, and adults definitive therapy consists of surgical resection of the aganglionic segment and anastomosis of normally functioning bowel, often in staged operations to allow for growth and development.

Congenital and/or Developmental—Retrorectal Cysts

Retrorectal cysts is a broad term used to describe cystic lesion, mostly congenital, that are located posterior to the rectum and anterior to the sacrum. These rare lesions most commonly affect middle-aged females who may have mass effect-related symptoms such as lower back pain, anorectal pain, and constipation. These cysts can fistulize, leading to anal fistulae. Retrorectal cysts include epidermoid cysts, dermoid cysts, tailgut cysts, and rectal duplication cysts.^{37,38} The content and composition of epidermoid and dermoid cysts are the same as those encountered elsewhere, respectively simple uniloculated fluid and more complex content for dermoid cysts. Tailgut and rectal duplication cysts are lined with intestinal mucosa.

Retrorectal cysts are often misdiagnosed, often as a fistula-in-ano on clinical exam or as an abscess on cross-sectional imaging (Fig 11). Singer et al.³⁹ reported a retrospective surgical series of retrorectal cysts in 7 patients, all of whom were initially misdiagnosed, underwent a mean of 4.7 operations or procedures to diagnose or treat their disease, and all patients went to resection. The most common misdiagnosis was as an abscess in which 4 patients underwent unsuccessful prior percutaneous or surgical drainage of these retrorectal cysts.

Tailgut cysts, also known as retrorectal cystic hamartomas, may undergo malignant degeneration, therefore are more likely to undergo surgical resection compared to other retrorectal cysts. Imaging findings on CT and MR are nonspecific and there is a paucity of imaging based studies. MR can give better internal cyst

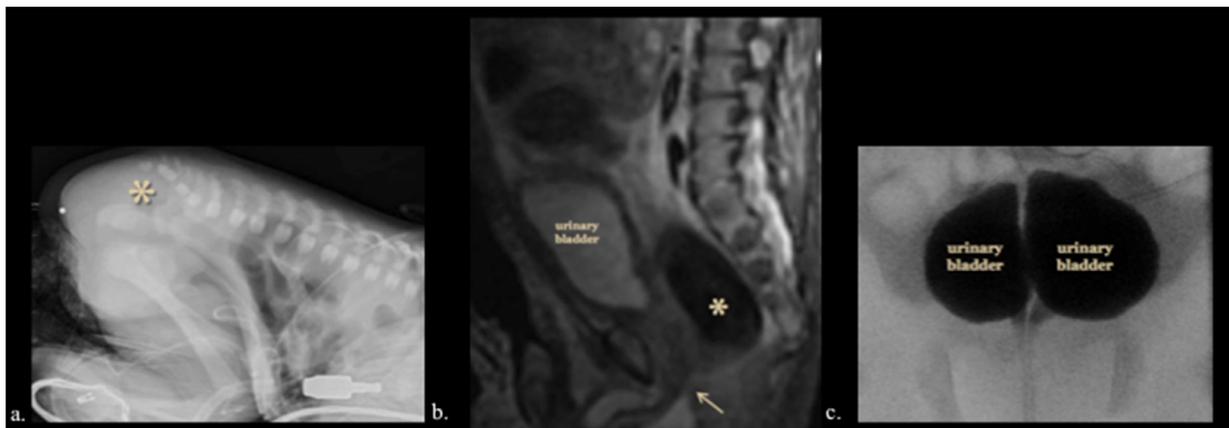


FIG 9. Prone jack-knife lateral view (a) and sagittal T2 lumbar MR (b) of a male newborn with clinically imperforate anus. The rectal air column is seen deep to the coccyx (*) compatible with a low defect (arrow). MR of the spine was negative for tethered cord, although urinary bladder and urethral duplication was identified on voiding cystourethrogram (c). MR, magnetic resonance.

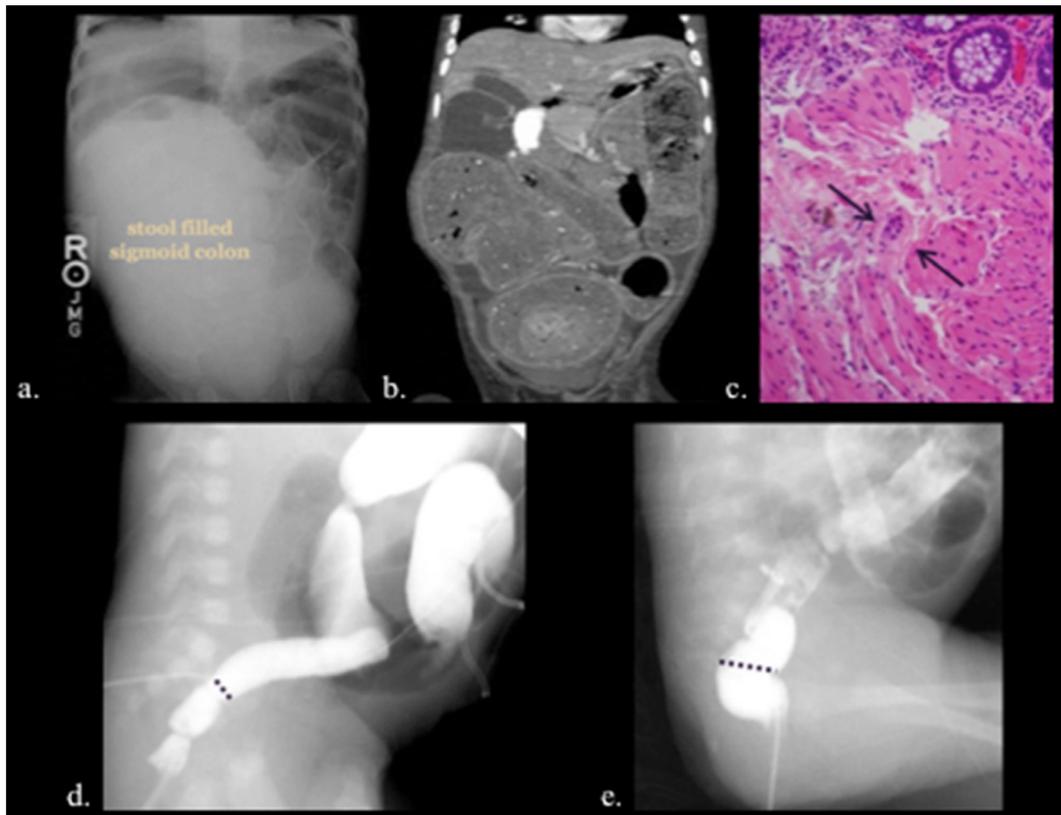


FIG 10. (a-c) A 11-month-old male with abdominal distention and failure to thrive subsequently diagnosed with Hirschsprung disease. Abdominal radiograph (a) and coronal CT (b) demonstrate massive distention of a stool filled colon. Multilevel suction mucosal biopsies revealed rectal aganglionosis, and ganglion cells (arrows) at mid-descending colon (c). (d) Newborn with failure to pass meconium and nondistensible rectum (dashed line) on contrast enema compatible with Hirschsprung disease (d). The rectum is normally the most distensible portion of the left colon, as seen in the normal enema comparison (e). CT, computed tomography.

characteristics, especially on T2 weighted images and fluid sensitive sequences, but again, one cannot make a specific diagnosis as features overlap.⁴⁰

Rectal duplication accounts for <10% of gastrointestinal duplication cysts. Complications include infection, bleeding, and malignant degeneration. In one historic pediatric surgical series, 5 of 96 patients with alimentary tract cysts were rectal duplications cysts. All presented at less than 6 months of age and 4 of the 5 were resected.⁴¹ Rectal duplications cysts can be asymptomatic in childhood and present later in life as an adult with pelvic pain and obstructive symptoms.⁴² For all retrorectal cysts which require resection, treatment is surgical with or without coccygectomy with a parasacrococcygeal approach for low rectal lesions and combined abdominoperineal approach for high lesions.

Vascular—Hemorrhoids and Rectal Varices

Hemorrhoids are normal arterIALIZED submucosal sinusoids that act as cushions in the anal canal and contribute to continence. Abnormal dilatation of these subepithelial vascular cushions above the dentate line produces internal hemorrhoids and below the dentate line external hemorrhoids. Symptoms may include bleeding, pain, pruritus, and prolapse. Treatments range from topical ointments for external hemorrhoids, to sclerotherapy, coagulation, and banding for internal hemorrhoids, to surgical hemorrhoidectomy.^{7,8} External hemorrhoids distal to the dentate line have sensory innervation, therefore require anesthesia (locoregional or general) prior to manipulation. In contrast, internal hemorrhoids do not have sensory innervation and can be manipulated analogous to polyps without

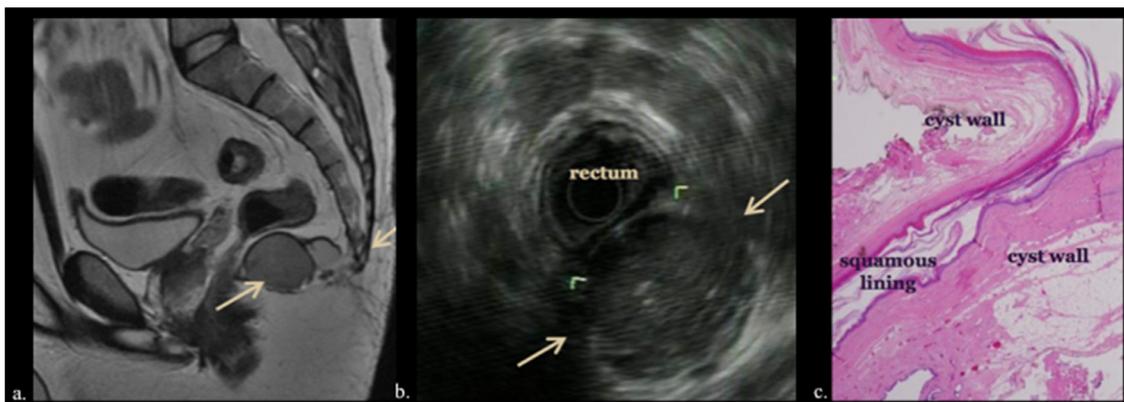


FIG 11. Retrorectal epidermoid cyst. (a-c) Sagittal T2 MR and endoanal ultrasound of a 35-year-old male presenting with blood per rectum and lower abdominal pain. Multiloculated mixed signal retrorectal lesion (arrows) is avascular with low level internal echoes on ultrasound. The cyst wall is lined by squames compatible with epidermoid.

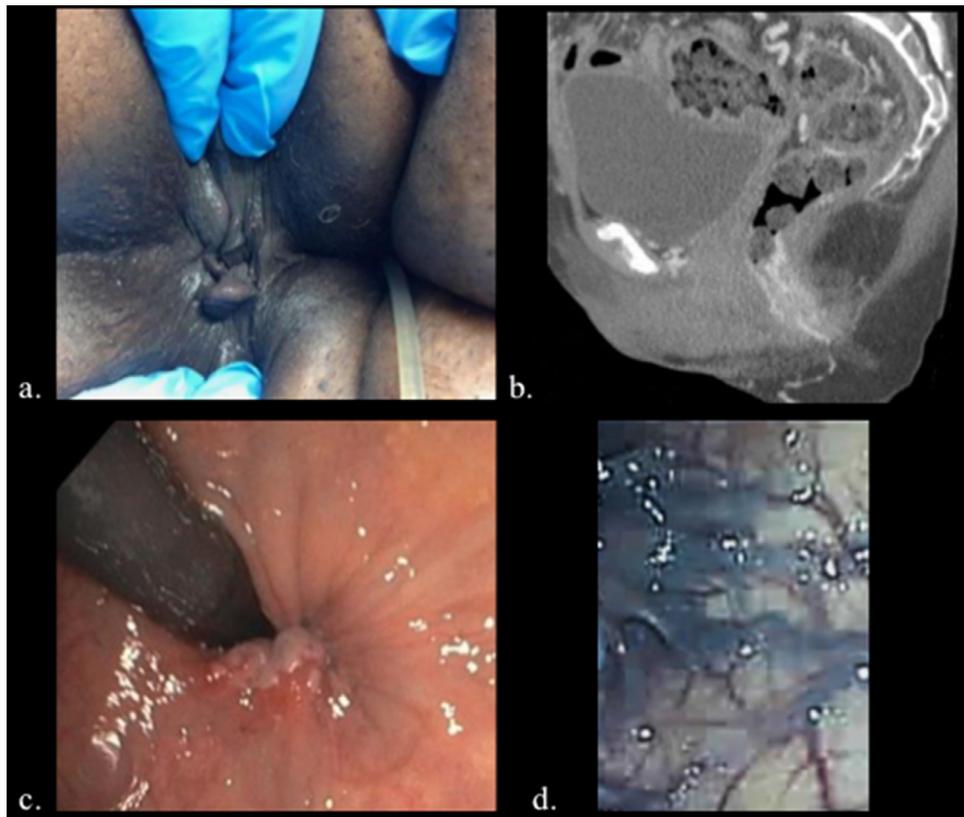


FIG 12. (a, b) Clinical image (a) and CT (b) of external hemorrhoids. (c, d) Endoscopic images of internal hemorrhoids. CT, computed tomography.

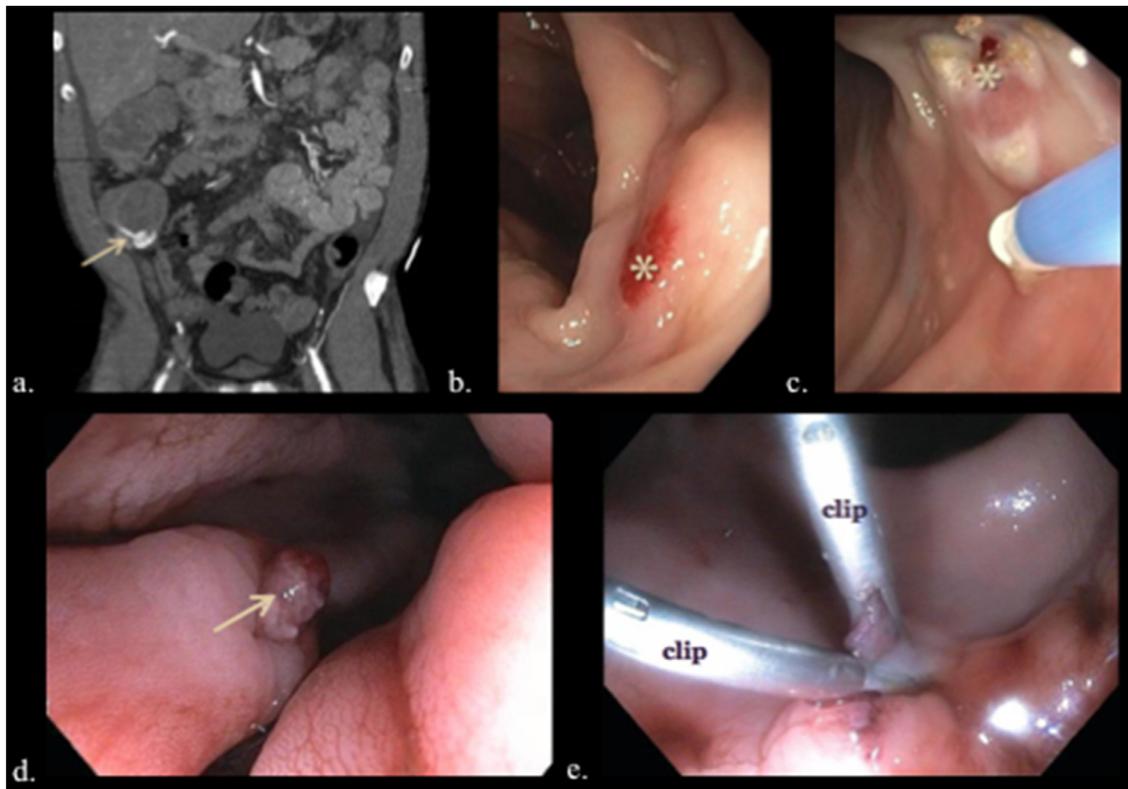


FIG 13. (a-c) A 58-year-old male with bright red blood per rectum. Active cecal bleeding on CT (arrow) was caused by angioectasia. Endoscopic rectal angioectasia (*) pre-coagulation and post-coagulation therapy. (d, e) A 44-year-old male with acute onset of painless hematochezia. Protruding rectal Dieulafoy lesion (arrow) was treated with hemostatic clip placement. CT, computed tomography.

anesthesia. Imaging is certainly not need to diagnose hemorrhoids, however, it is important to know the imaging features as simple hemorrhoids have been mistaken for malignancy on cross-sectional imaging and vice versa.^{43,44}

Rectal varices and hemorrhoids are two separate entities, however their imaging appearance is similar in appearance though different in location, thus discussed together. Rectal varices are abnormally dilated veins in and about the rectum as a sequela of portal hypertension and the portal tributaries to the hemorrhoidal plexus. The hemorrhoidal plexus is made of branches from the portal system and iliac veins. The superior hemorrhoidal vein is a branch of the inferior mesenteric vein and the middle and inferior venous plexus is

tributary of the internal iliac vein.⁴⁵ Rectal varices are distinguished from hemorrhoids as they originate more than 4 cm from the anal verge and are not contiguous with the anal columns or pectinate line. Rectal varices are very prevalent in patients with cirrhosis and portal hypertension, estimates range from 38% to 94%. Clinically significant bleeding from rectal varices is rare (<1%-5%). Treatment includes aims focused at improving portal hypertension, such as transjugular intrahepatic portosystemic shunt. Various endoscopic manipulations can effectively treat the rare clinically significant lower gastrointestinal bleed.⁴⁶

On imaging, hemorrhoids may appear as large serpiginous veins in the anal canal (Fig 12). More proximal serpiginous veins in the rectal

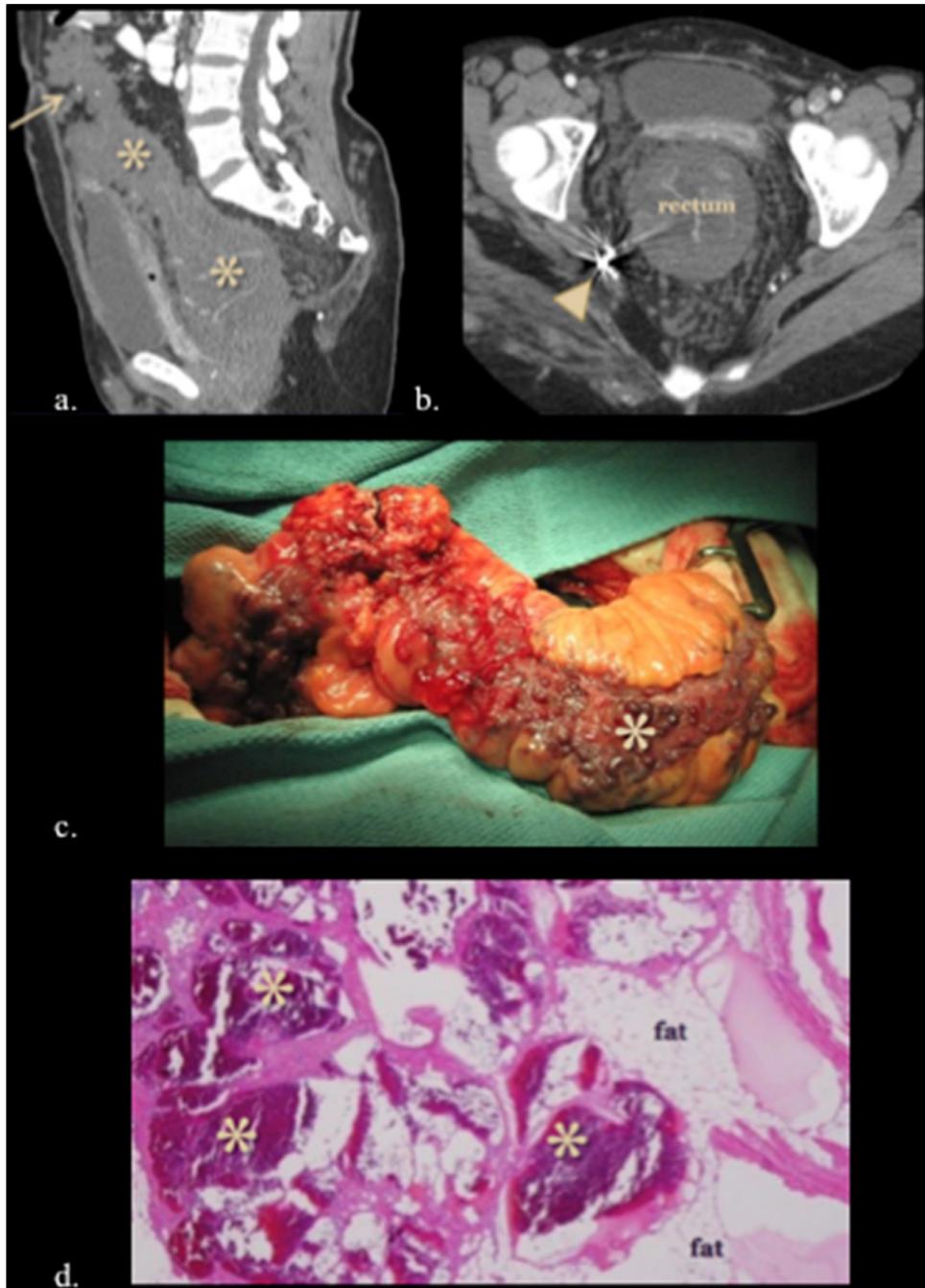


FIG 14. Klippel-Trenaunay-Weber syndrome. (a and b) A 26-year-old female with past history of right lower extremity vascular malformation postendovascular treatment, and recurrent lower gastrointestinal bleeding. CT illustrates marked circumferential colorectal mural thickening (*), mesenteric varices and associated phleboliths (arrow). Note embolic coil (arrowhead), in addition to mesorectal and gluteal varicosities. (c and d) Dilated vascular channels (*) involve the entirety of the colonic wall grossly and microscopically consistent with angiomatosis. CT, computed tomography.

lumen or surrounding it are the imaging features of rectal varices. Thrombosed hemorrhoids will typically be larger and more apparent and will not enhance or poorly enhance during a venous phase of contrast. Rectal varices may enhance during a venous phase. Active bleeding from either hemorrhoids or rectal varices will very rarely be visualized as active extravasation.⁴⁷

Vascular—Vascular Malformation

There are a number of causes of lower gastrointestinal bleeding due to vascular malformation, which may be arteriovenous, venous, or capillary. Angioectasia, Dieulafoy lesions, and Klippel-Trenaunay syndrome are three representative entities that may cause clinically significant bleeding in the rectum. Regardless of the etiology diagnosis can be made by CT angiography, red blood cell scintigraphy, and catheter angiography. Endoscopic hemostasis may be achieved through clipping, epinephrine injection, banding, or coagulation.⁴⁸⁻⁵⁰

Angioectasia, or angiodysplasia, is a common cause of occult gastrointestinal bleeding. Their pathogenesis is debated, but one of the most cited mechanisms is that with aging, intermediate submucosal venous obstruction leads to arteriovenous collaterals leading to further engorgement of submucosal veins and eventually protrusion into the lumen. CT angiography may reveal nodular hypertrophied vessels within the submucosa and active extravasation. Angioectasia can occur in the rectum, but is more common in the cecum and ascending colon (Fig. 13a-c). Endoscopy may reveal a small flat red, fern-like mucosal lesion with a central vessel.⁴⁹⁻⁵¹

Dieulafoy lesions, in contrast, represent a tortuous exposed nontapering submucosal artery that penetrates intraluminally over time.

Accounting for 1%-2% of gastrointestinal bleeding and are much more common in the foregut than the small and large intestine, but can present in the rectum (Fig. 13d and e). Dieulafoy lesions can be a diagnostic dilemma due to their intermittent and intense nature of their bleeding. As nearly half of these lesions can be missed at initial endoscopy, CT angiography, tagged red cell blood scintigraphy, or catheter angiography can potentially reveal the lesion.⁵⁰

Klippel-Trenaunay syndrome is a rare congenital disorder that includes a triad of a cutaneous capillary malformation (port wine stain), venous varicosities or malformations, and soft tissue and bony extremity overgrowth. Two of the three are need for diagnosis. Capillary malformation is the most common manifestation and unilateral extremity hypertrophy is the least common. When a true arteriovenous malformation is present, the syndrome can be termed Klippel-Trenaunay-Weber or Parkes Weber syndrome. Gastrointestinal vascular abnormalities can produce bleeding in 1%-12.5%, most commonly from the colon and rectum (Fig 14).⁵¹

Miscellaneous—Pelvic Lipomatosis

Pelvic lipomatosis is a rare benign proliferation of pelvic adipose tissue which may compress and distort pelvic viscera, including the rectum. The majority of patients are male and present with vague chronic abdominopelvic pain or low urinary tract symptoms. Proliferation of unencapsulated fat with surrounding fibrosis and inflammatory tissue can distort the course of the distal ureters or urinary bladder, leading to low urinary tract symptoms and hydronephrosis. Additionally, extrinsic compression upon pelvic organs can manifest chronic pelvic pain. On imaging, CT and MR will show an abundance

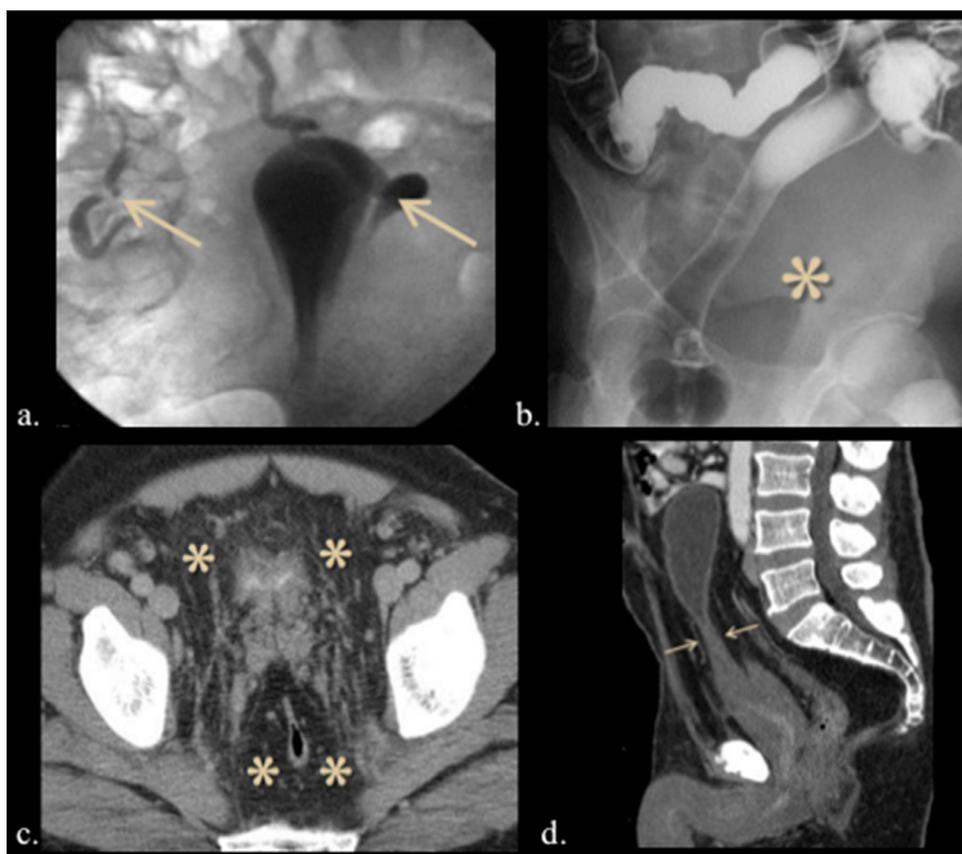


FIG 15. Two patients with pelvic lipomatosis. (a-c) A 53-year-old male undergoing excretory urography for evaluation of hematuria. Pear shape urinary bladder configuration, lateral deviation of the ureters (arrows) and extrinsic compression of the rectum on subsequent barium enema are confirmed as secondary to expansion of nonencapsulated pelvic fat by CT (*). (d) Sagittal CT of similar process in an asymptomatic male undergoing staging CT for colorectal carcinoma. Note the compression and distortion of the bladder (arrows). CT, computed tomography.



FIG 16. (a and b) Dynamic imaging with defecography or MRI may be used to assess patients with disordered defecation. Rectocele (*), transient rectorectal intussusception (RR), and herniation of sigmoidocele (arrow) are illustrated. (c) Rectal prolapse on CT. Note vertical orientation of anorectal junction (dashed line) and protrusion of anus below the pubococcygeal line at rest on CT. CT, computed tomography; MRI, magnetic resonance imaging.

of perivesical and perirectal fat (Fig 15).⁵² The largest series to date is a radiologic series by Zhang et al.⁵² in which they retrospectively reviewed the CT and MR findings of 32 clinically proven cases of pelvic lipomatosis and compared them to normal controls, correlating radiologic findings with both pelvis lipomatosis and severity of disease. They also define different bladder shapes and found that symmetric compression of the bladder of its most caudal aspect (that what they described as banana and pear shaped bladders) had a high specificity for pelvic lipomatosis compared to controls, although sensitivity was low (40%). The majority of patients had some degree of bladder wall thickening. Approximately half were associated with solitary or multiple bladder diverticula. Many (22 of 32) patients showed fibrous tissue about the proliferated adipose tissue with a speckled appearance on T1WI, what they described as “salt and pepper.”⁵² Treatment is often related to severity of urinary tract symptoms. Up to 40% may experience urinary obstruction requiring ureteral reimplantation or urinary diversion.⁵³

Miscellaneous—Rectocele, Intussusception and Prolapse

Dynamic imaging with traditional fluoroscopic or MR defecography is used to assess patients with constipation or fecal incontinence (Fig 16). Enteroceles, rectoceles, transient rectal intussusception, and rectal prolapse are common pathologic conditions. Enterocoele is a herniation of the peritoneal sac into the rectovaginal or rectovesicular space, impairing defecation. Rectoceles are bulges of the rectal wall, most often anterior wall, due to laxity or inadequate support about the anal canal. Rectocele can be a normal finding with no symptoms. One study of healthy volunteers demonstrated rectoceles in 17 of the 21 (81%) female volunteers compared to fewer (3 of 24, 13%) in male volunteers.⁵⁴ Rectocele increase in frequency with parity and may be a sequela of obstetric trauma in addition to normal increasing pelvic floor muscle laxity. Clinically significant rectoceles are related to their size and occur almost exclusively in females. Size greater than 2 cm has been proposed to be a clinically significant size. In the aforementioned healthy volunteer study, only 1 of the 20 incidental rectoceles was larger than 2 cm.⁵⁴ Painless bulging of the anterior rectal wall into the posterior vagina occurs secondary to loss of rectovaginal septal support and additional pelvic compartment defects are common. Defecography will reveal this bulge in real time during defecation.⁵⁵ Rectoceles are common findings in asymptomatic patients. Larger size is associated with increased frequency of symptoms. Large rectoceles may be asymptomatic but obstructed defecation can be treated with surgical repair.

Transient rectal intussusception and external rectal prolapse are spectra with prolapse being the extreme. Transient intussusception without prolapse involves part or all of the rectum wall temporarily invaginating descending toward or into the anal canal. External rectal prolapse is when this intussusception traverses past the anal verge and may be limited to mucosa alone or involve all rectal wall

components. Prolapse is most common in females over the age of 60 years. Defecography will characterize intussusception in real time during defecation.⁵⁵ Therapy for rectal prolapse is surgery such as rectopexy and resection of redundant rectosigmoid.

Conclusion

Many conditions may affect anorectal and perirectal areas and knowledge of common and rare anorectal entities will help the radiologist develop an appropriate differential, delineating incidental findings, and adequately protocoling problem-focused studies. Imaging modalities offer information complementary to physical examination for optimal assessment and treatment planning. Many of these entities, such as hemorrhoids, will be clinically apparent, but knowledge of their appearance will help the radiologist in recognizing different entities, such as thrombosed hemorrhoids and rectal varices. In many cases, CT and MR can assess the extent, complications, and associations of these infectious, inflammatory, and other conditions. Knowledge of expected appearance, characteristic findings, clinical association, and management strategies can help prompt a prospective diagnosis in challenging rectal lesions and conditions.

Conflicts of Interest

All authors claim no conflicts of interest or disclosures.

Conference Presentation

Concepts and some cases from this review were presented as an educational exhibit at the 2014 RSNA meeting—Naeem S, Heldmann M, Dubose A, Thomas-Ogunniyi J, Sangster G, Cole P. Rectal, Perirectal and Perineal Diseases—a Multi-modality Pictorial Review with Histological, Endoscopic and Operative Correlation. Radiological Society of North America 2014 Scientific Assembly and Annual Meeting; Chicago IL. Abstract Available at: <http://archive.rsna.org/2014/14019566.html>.

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