The Anorectum

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7.1 Macroscopic Anatomy

7.1.1 Rectum and Anus

The *rectum* is the most distal segment of the large intestine, extending from the sigmoid colon. The junction between the two is marked by the rectosigmoid angle located at the level of the S3 vertebra. It is approximately 15-cm-long in adults, ending distally in the anal canal. The rectum and the sigmoid form, at rest, a right angle (open at the back and whose angulation depends on the contraction of the puborectalis muscle) that will open (straighten) during defecation. The rectum is extraperitoneal in its lower two-thirds and follows the concave curve of the sacrum. In men it is located behind the bladder and the prostate, and in women it is behind the uterus and the vagina (◘ Fig. 7.1).

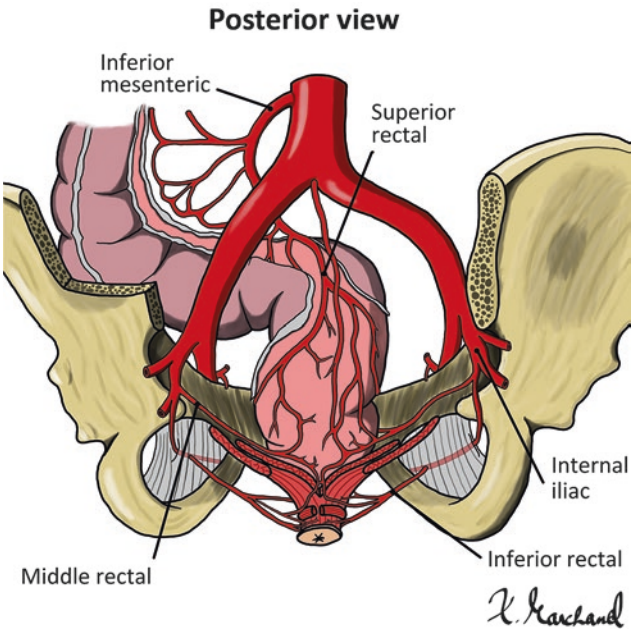
Inside the rectum, three semicircular valves (valves of Houston) are formed by folds of the rectum wall.

The *anal canal* is the terminal portion of the intestinal tract that extends from the lower end of the rectum to the anal orifice at the skin. It is a muscular cylinder about 2- to 4-cm-long that is angled down and toward the back. It is an area of high pressure with a key role in maintaining fecal continence. This high-pressure area is directly related to the presence of two sphincters: the internal anal sphincter and the external anal sphincter.

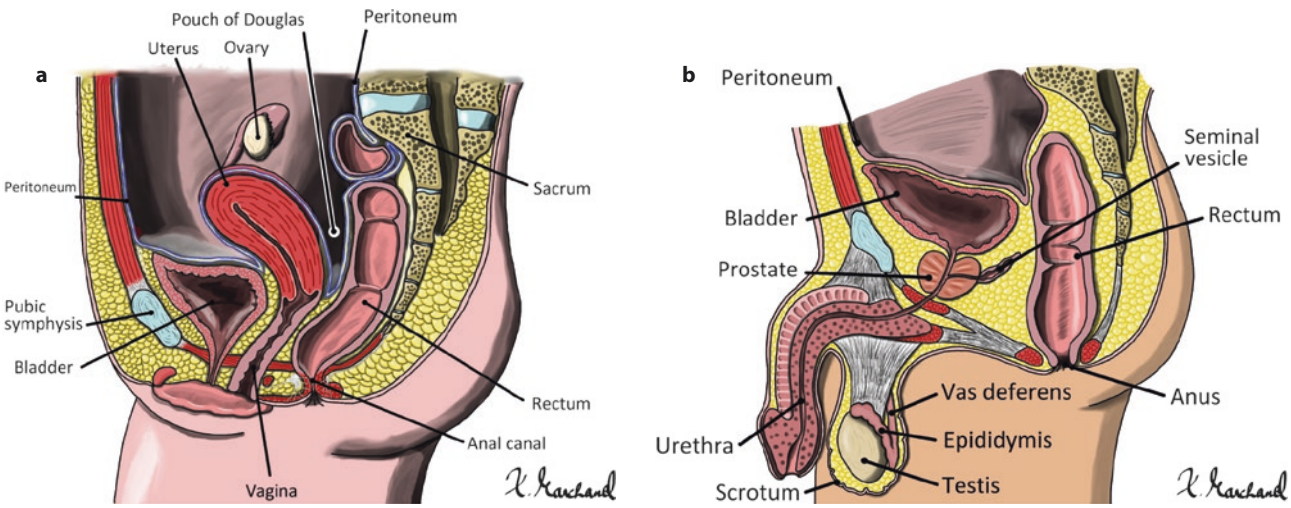
7.1.2 Vascular Supply

Arteries The rectum receives arterial supply from five arteries: the superior, middle (right and left), and inferior (right and left) rectal arteries (also called hemorrhoidal arteries). The superior rectal artery is the main one; arising from the inferior mesenteric artery, it is divided into two branches feeding the posterior and anterior sides of the rectum. The collateral branches penetrate the rectal muscular wall and descend to the anal canal. The rectosigmoid angle, the rectum, and the mucosa of the anal canal are fed by the superior rectal artery (◘ Fig. 7.2). The middle rectal artery originates from the internal iliac artery. It supplies blood to the rectum and the genital area, but it only exists in about one in two cases.

The inferior rectal artery, a branch of the internal pudendal artery (which arises from the internal iliac



◘ **Fig. 7.2** The arteries supplying blood to the rectum and anus (called rectal or hemorrhoidal arteries) originate from a branch of the inferior mesenteric artery for the upper rectum and from branches of iliac arteries for the more distal areas



◘ **Fig. 7.1** Anatomy of the pelvis (lateral view) in woman (**a** on left) and in man (**b** on right)

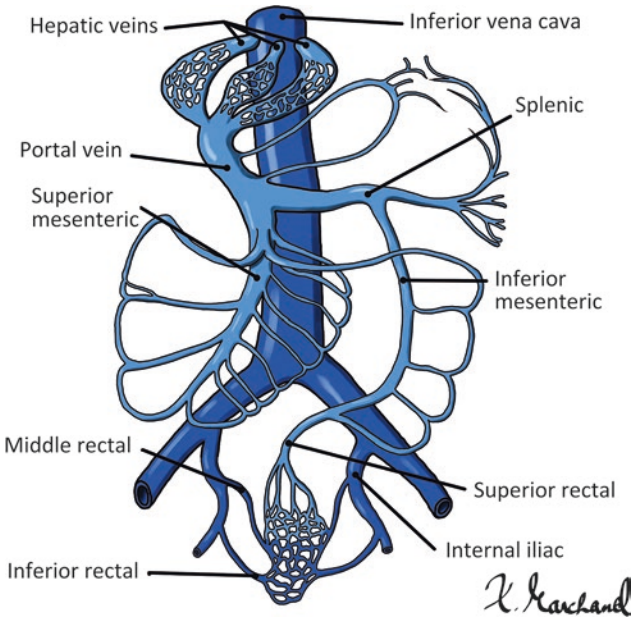


Fig. 7.3 Veins from the lower rectum and anus drain blood into iliac veins to the systemic circulation via the iliac veins. The upper rectum drains into portal system via the lower mesenteric vein to the splenic vein (which joins the upper mesenteric vein to form the portal vein going to the liver)

artery) vascularizes the external and internal sphincters as well as the levator muscle of the anus.

Veins Venous vascularization follows the arterial system (Fig. 7.3). Thus, the superior rectal vein forms, with sigmoid veins, the inferior mesenteric vein which then joins the splenic vein and the portal venous system.

The middle and inferior rectal veins are not always present. They drain the lower part of the rectum and the anal canal to the internal iliac veins connecting to the inferior vena cava.

The rectum drains to the portal as well as to the systemic venous system. Since the five rectal veins connect (anastomose) with each other in the submucosa of the lower rectum (forming the hemorrhoidal plexus), anastomoses between portal and vena cava systems result. These anastomoses explain the presence of rectal varicose veins in portal hypertension due to liver cirrhosis.

Lymphatics Lymphatic plexuses of the rectal and anal walls are the starting point of lymphatic vessels that join lymph nodes in the perirectal adipose tissue. Lymphatic drainage follows the arteries in the mesorectum for most of the rectum and ascends to the inferior mesenteric artery. The lower part of the rectum and the ano-perineal region are drained to the inguinal region. Cancer of the rectum has a high risk of lymph node metastasis as soon as the cancer spreads beyond the muscularis mucosae. This characteristic of rectal cancer associated with its double lymphatic drainage (i.e., "portal" and systemic) helps to understand the high risk of locoregional recurrence of rectal cancers after surgery, as well as the increased prevalence of metastatic spread in systemic territories (independent from the portal system) such as the lungs.

7.1.3 Innervation

As in the entire digestive tract, rectal innervation has both intrinsic and extrinsic architecture (Fig. 7.4).

(a) Intrinsic innervation is provided by the enteric nervous system (ENS), which is present all along the intestinal wall (as already described in previous chapters of this book). Sphincter relaxation is mainly ensured by VIP and ATP neurotransmitters.
(b) Extrinsic somatic innervation reaches the striated muscles of the pelvis and consists of the motor bundles (S2, S3, S4) of the pudendal nerve. The pudendal nerve transmits motor function for the external sphincter of the anus and sensation for the perineum.
(c) Extrinsic autonomous innervation depends on parasympathetic and sympathetic fibers, both efferent and afferent, connected to the central nervous system, including the hypothalamus.

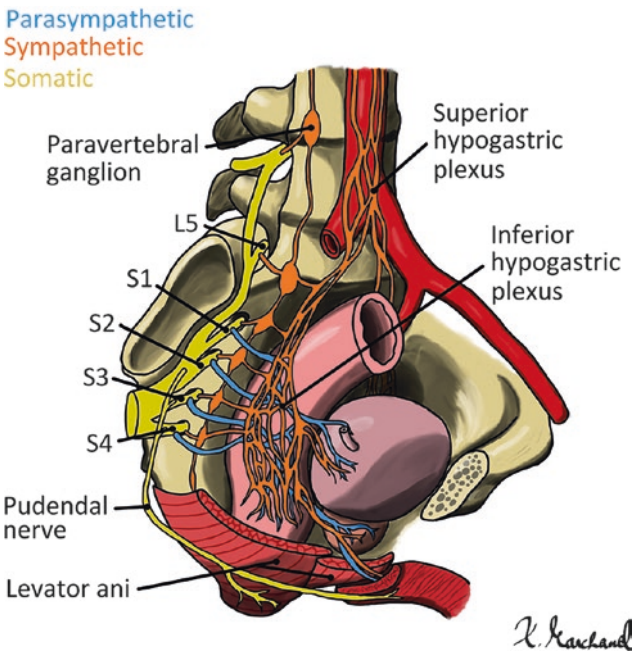


Fig. 7.4 Innervation of the anorectal region: sympathetic fibers from paravertebral ganglia synapse in hypogastric ganglions (superior and inferior) before reaching their target organs. The parasympathetic fibers come from sacral roots S1 to S4. Somatic innervation is provided by the pudendal nerve (a branch of the nerve roots forming, among others, the sciatic nerve)

Parasympathetic fibers Central parasympathetic fibers synapse with cells from the vagal dorsal nucleus on the floor of the fourth ventricle to give rise to the 10th cranial nerve, the pneumogastric or vagus nerve, which travels down along the esophagus until the proximal colon. Parasympathetic fibers also descend into the spinal cord to synapse with sacral roots from S2 to S4 and innervate the distal colon, i.e., the sigmoid and rectum. Sacral parasympathetic fibers, both motor and sensory, play an important role in the defecation (cholinergic parasympathetic stimulation relaxes the internal anal sphincter).

Sympathetic fibers Sympathetic fibers all go down from the brain to the abdomen through the spinal cord and intervertebral nerve roots. Fibers issued from the spinal cord are called "preganglionic" fibers since they all go to nerve ganglia from where the so-called postganglionic fibers emerge to reach the various digestive organs. There are five important neurological ganglia (or plexuses) in the abdomen:

- The celiac ganglion, located close to the aorta at the origin of the celiac trunk artery, receives thoracic splanchnic nerves synapsing to generate postganglionic fibers that innervate the foregut organs (stomach, duodenum, pancreas, etc.). Celiac ganglion block can be done to alleviate pain from pancreatic cancer. Interruption of sympathetic activity can cause diarrhea.
- The superior mesenteric ganglion located at the junction between the aorta and the superior mesenteric artery receives lower thoracic splanchnic nerves (from T6 to T12) to give rise to postganglionic fibers innervating the midgut, i.e., the small bowel and the proximal colon.
- The inferior mesenteric ganglion, at the aortic root of the artery, receives lumbar fibers from L1 to L3 to innervate the left colon.
- The superior hypogastric ganglion, located in front of the aortic bifurcation, receives preganglionic fibers from L4 to L5 and gives postganglionic axons innervating the sigmoid and the rectum.
- The inferior hypogastric (or pelvic) plexus receives sacral fibers from S1 to S3 destined mainly to the anorectal region and pelvic organs such as the prostate, bladder, seminal vesicles, etc. Damage to these pelvic nerves during surgery with rectal dissection can lead to erectile or bladder dysfunction. Alpha-adrenergic sympathetic stimulation contracts the internal anal sphincter, while beta-adrenergic stimulation relaxes it.

7.2 Microscopic Anatomy

7.2.1 Mucosa

Rectal mucosa The mucosa of the rectum is similar to the rest of the colon. The epithelium is columnar and lies on a basal membrane. In the upper part of the crypts, so-called enterocytic absorptive cells and mucus cells (or goblet cells) are predominant, while at the bottom of the crypts, more undifferentiated cells and some endocrine cells are present. Muscularis mucosae separate the mucosa from the submucosa as in the rest of the colon.

Anal mucosa The anus is covered by a squamous epithelium which meets the columnar colonic epithelium (anorectal line or squamocolumnar junction) just above the columns of Morgagni and extends down 4–5 cm to the anal margin (ano-dermal line) where the skin (rich in hair follicles, sweat glands, etc.) begins.

About halfway up the anal canal (2–3 cm from the anal margin), the pectinate or dentate line, is clearly visible. It is made of 10–12 half-moon-shaped valves called Morgagni's columns that cover crypts where channels emerge from glands located in the rectal wall (called Hermann and Desfosses' glands). These glands reside in the inter-sphincter space and are the starting point for abscesses and anal fistulas.

Above the pectinate line, the mucosa takes on a reddish-purple appearance due to the internal hemorrhoidal plexuses located in the submucosa.

Below the pectinate line, a non-keratinized squamous cell type epithelium is present. This 1 cm transitional zone (also called anal pecten) is richly innervated and continues downward to blend with the keratinized skin, gradually changing from being initially smooth to become more pigmented and rich in hairs and sebaceous and sweat glands.

The pectinate line corresponds to the junction of the endoderm and the ectoderm structures during the development of the embryo.

7.2.2 Muscularis

(a) Muscles of the rectum The rectal muscularis is made up of a circular inner layer and a longitudinal outer layer different from those of the colon. In the colon, the longitudinal layer does not cover the whole circumference of the colon and is reduced to three strips called taenia coli; in the rectum, the outer layer is continuous as elsewhere in

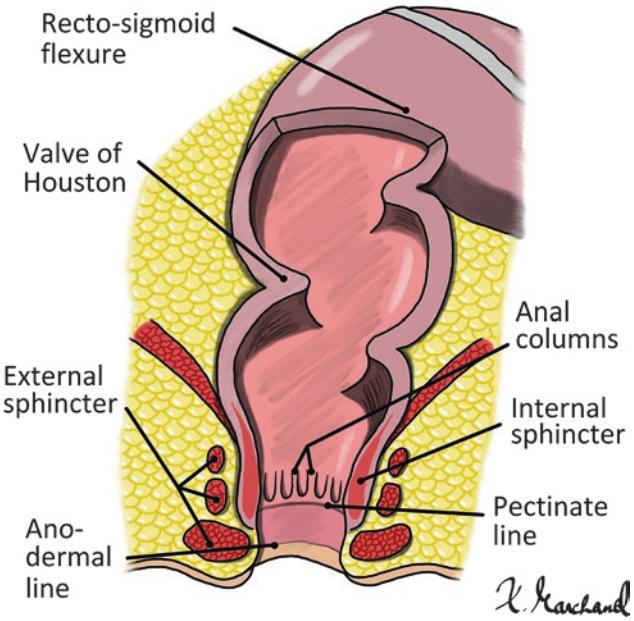


Fig. 7.5 Anus, pectinate line, sphincters, Houston's valves

the digestive tract. The circular and longitudinal muscle layers will, in the part distal to the rectum, merge to form the internal sphincter. The Auerbach's myenteric plexus is located on the outer surface of the circular muscle.

In the rectum, three transverse folds of the mucosa and circular muscles form semilunar valves (valves of Houston) that take up about 1/3 of the lumen. The upper and lower valves are located on the left side wall; the middle valve is on the right. The three valves are located 2–3 cm, 6–7 cm, and 10–11 cm, respectively, above the pectinate line (Fig. 7.5). The function of these valves is uncertain.

(b) External anal sphincter The external anal sphincter is a muscle structure, made of three concentric layers (deep, intermediate, and subcutaneous) of striated muscles which surround the virtual space of the anal canal, forming a ring about 1-cm-thick along the entire length of the canal and specifically at its lower third. The external sphincter is a striated muscle and is under conscious control (e.g., relaxation during defecation and contraction during retention efforts) by the somatic nervous system via the pudendal nerve.

(c) Internal anal sphincter The internal anal sphincter is made of smooth muscle about 4-mm-thick that surround the upper two-thirds of the anal canal. It is an inseparable extension of the internal circular muscles of the rectal wall. It is responsible for the resting tone of the anal canal. It is almost always contracted, relaxing only for brief moments in response to certain stimuli. Sphincter tone is regulated by the autonomic nervous system: lowered by the cholinergic parasympathetic nerves (from S2 to S4) and raised by the alpha-adrenergic sympathetic fibers (from L5).

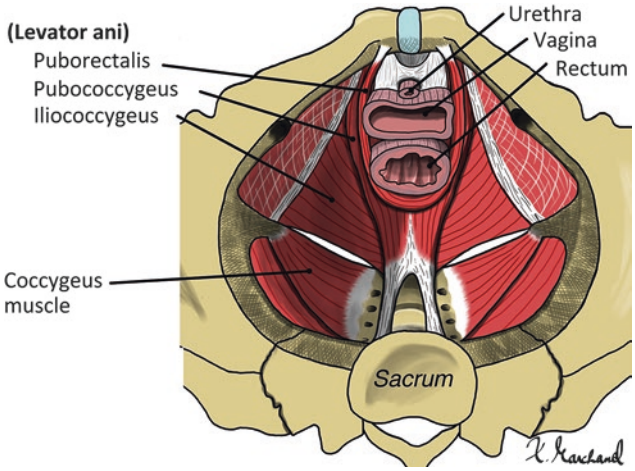


Fig. 7.6 The levator ani is a muscle structure made of three muscles (pubococcygeus, iliococcygeus, and puborectalis) attached in the front at the pubis, to the sacrum at the back, and to the sides of the pelvis to constitute the pelvic floor, which supports the pelvic viscera like a hammock

(d) Levator ani The levator ani is a muscle structure involved in the formation of the pelvic diaphragm, which separates the pelvis into an upper and a lower (perineal) compartment and which forms a thin hammock supporting the pelvic organs (Fig. 7.6). It originates in the front at the pubis and has three muscle components: the pubococcygeus and the iliococcygeus which attach to the sacrum at the back and to the inner surface of the pelvis to support the pelvic viscera (pelvic floor). The puborectalis is the third component muscle which, from the pubis, passes behind the top of the anal canal, in the form of a U-shaped loop that pulls the rectum forward and closes the anorectal angle, thus obstructing the passage of stools and promoting continence. During defecation, the muscle is relaxed to open the anorectal angle, allowing the rectum to be aligned with the axis of the anus and facilitating the expulsion of the rectal contents. The important contribution of the puborectalis muscle to continence is illustrated in Fig. 7.7.

7.2.3 Serosa

The rectum has no serosa in its extraperitoneal segment (the lower two-thirds).

7.3 Embryology

7.3.1 Normal Development

From the primitive digestive tract (derived from the endoderm), three regions (foregut, midgut, hindgut) are formed according to the territories irrigated by the main three arteries of the digestive tract (celiac trunk, superior

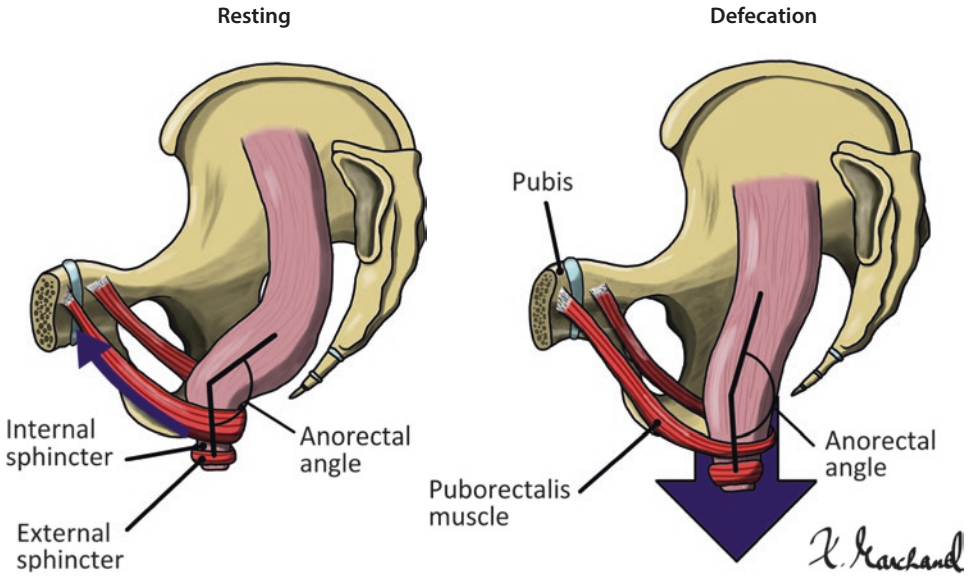


Fig. 7.7 Anus muscles for fecal continence: **a** the internal sphincter provides basal tone for the anal canal (and relaxes during defecation); **b** the external sphincter is made of striated muscles subject to voluntary control (relaxation during defecation or contraction during retention efforts); **c** the puborectalis muscle pulls the anal canal forward and tightens the anorectal angle to help maintain continence (relaxes to open the anorectal angle and facilitate passage of stools during defecation)

mesenteric artery, inferior mesenteric artery). The descending colon and the rectum arise from the hindgut. Between weeks 4 and 6 of embryonic development, from the distal part of the hindgut, the cloaca will divide into an anterior urogenital sinus and the posterior rectum.

The upper two-thirds of the anal canal also originates from the most distal part of the endodermal hindgut. Its lower third derives from the ectoderm where, facing the endodermal anal canal, an "anal depression" will appear. Endodermal and ectodermal portions of the anal canal are separated by an anal membrane until week 8. In adulthood, the area where this membrane was located can be identified as the pectinate line. The double origin of the anorectum has consequences in its vascularization since the upper part of the rectum is irrigated by abdominal vessels (blood coming from the inferior mesenteric artery and drained to the portal vein), whereas its lower part depends on peripheral internal iliac arteries and veins.

7.3.2 Atresias

Atresias of the anorectal region are present in 1/5000 births. The most frequent abnormalities are the following:

- Failure to separate the distal hindgut (which normally gives the urogenital system in front and the rectum behind). The abnormality observed is then a cloaca (i.e., as in birds where the urinary and fecal waste ends up in a common reservoir).
- Defect of fusion between the ectodermal anus and the endodermal anorectal apparatus. This is called imperforate anus and may present with or without a gap between the anus and the rectum.

The diagnosis of atresia is usually made at birth, when an inspection of the perineum reveals anal imperforation. Other associated malformations must be looked for, such as those affecting the pelvis, vertebrae, as well as urinary, genital, digestive, bone, or muscle systems. The surgical treatment for reconstruction will depend on the anatomical assessment.

7.3.3 Hirschsprung's Disease

Hirschsprung's disease is due to an absence of enteric nervous system plexus ganglions in the distal intestine. The affected length of intestine varies. Its prevalence is 1/5000 births. It is more common in boys. There may be a genetic component and it is also more frequent in trisomy 21.

Pathophysiology is caused by a halt, during the first 12 weeks of the embryonic gut development, in the migration of neuroblasts from the neural crest to the digestive tract. This migration of the precursor cells of the intestinal neurons takes place in the cranio-caudal direction, and the aganglionic zone will thus start at the bottom from the internal anal sphincter and extend proximally over a more or less long segment of the gut. It most often affects the rectum and the distal sigmoid,

but sometimes it can involve the entire colon (or even the small intestine).

Many genetic mutations are associated with Hirschsprung's disease, including those affecting RET gene coding for proteins assisting movements of neural crest cells.

From a functional or clinical point of view, the aganglionic zone is in permanent contraction creating a narrow and rigid intestinal segment acting as an obstacle to the progression of the endoluminal content. The result is an upstream gut distension, hence the name of congenital megacolon which was given to Hirschsprung's disease. Diagnosis is obtained with anorectal manometry testing, since relaxation of the internal anal sphincter upon defecation is always impaired in all patients. The so-called recto-anal inhibitory reflex (relaxation of the internal anal sphincter in response to rectal distension) can easily be observed during anorectal manometry, an examination which can be carried out in newborns. This reflex is innate and can be detected in a newborn baby even under sedation by distending the rectum with a small balloon. In Hirschsprung's patients the inhibitory recto-anal reflex is absent. Rectal biopsy may also be easily performed trans-anally to demonstrate the absence of nerve cells (Meissner's plexus) in the submucosa.

Hirschsprung's disease is often revealed by delayed evacuation of meconium, as the vast majority of newborns evacuate their first meconium within the first 24 h of life. This will be followed by signs of digestive obstruction (vomiting, abdominal distension, etc.). The disease can also cause constipation in children.

7.4 Secretion/Absorption

Secretion and absorption functions of the rectum are identical to those of the colon. In practice, however, stools are already formed by the time they arrive in this short segment of the digestive tract, and the rectum serves primarily as a reservoir (before scheduled and controlled emptying) rather than as an absorbing or secreting organ.

The absorptive function of the rectum can be used when administering medications rectally (suppositories or enemas). Fecal impaction (hardened stool in the rectum) may result from increased fluid absorption during prolonged stool retention in the rectum.

Rectal secretion may be increased during pathological processes such as inflammatory proctitis. Exaggerated rectal secretions result in defecations that are often frequent, but of small volume, that may contain only mucus (and blood) and where stools may be surprisingly soft (not frankly diarrheic or liquid) or even of normal consistency.

7.5 Motility/Sensitivity

The main function of the anorectum is to control the evacuation of fecal material. This motor action is inseparable from anorectal sensitivity, which has the unique feature of being a conscious sensation unlike the rest of the digestive tract. This awareness of the physiology of the anorectal sphere is essential to maintain continence and to allow defecation at appropriate times and places. Any disruption in this control could have consequences on the individual's social life.

7.5.1 Anorectal Physiology

The rectum is physiologically empty. One to three times a day, contractions of the sigmoid push the intestinal content of the colon into the rectum, which triggers the following sequence of events:

(a) *Sensation and need for defecation:* The arrival of stools in the rectal ampulla distends the rectum. Tension receptors in the rectal wall are then stimulated and send information, via the spinal cord, to the cerebral cortex informing the individual about the presence of materials in the rectum and a need for a bowel movement. Rectal hyposensitivity may be a cause of constipation in some patients who do not feel the presence of stools in the rectum and therefore "forget" to defecate.

(b) *Accommodation:* The pressure rise on the rectal wall by stools reaching the rectum is temporary. After few seconds, a phenomenon of accommodation appears, characterized by a relaxation of the parietal tension and therefore a decrease in the intraluminal pressure (at constant volume). This accommodation capacity is very important since it allows defecation to be put off temporarily. Accommodation capacity can be reduced in conditions such as inflammatory or radiation proctitis (causing frequent defecations of small stools), as well as, probably, in IBS (provoking a sensation of incomplete rectal evacuation).

(c) *Recto-anal inhibitory reflex*: In reaction to the pressure rise in the rectum, the internal sphincter will relax, opening the upper part of the anal canal (this sphincter will then regain its baseline tone in 30–60 s). During relaxation of the internal sphincter, some rectal contents may descend into the upper part of the anal canal, allowing the sensory squamous epithelium to perceive the nature of the contents (liquid, gas, or solid). This information travels up through the neurological pathways of the spinal cord to the cerebral cortex and informs the individual on the nature of the contents, which will enable him/her to eventually adapt his/her behavior (e.g.,

evacuation of gas vs. retention of a solid or liquid stool). The recto-anal inhibitory reflex is innate and depends only on the intrinsic nervous system (hence its absence in Hirschsprung's disease).

(d) *Recto-anal excitatory reflex*: Simultaneously with the recto-anal inhibitory reflex relaxing the internal anal sphincter, the excitatory reflex (also induced by rectal distension) contracts the external anal sphincter to close the lower part of the anal canal and prevent leakage of rectal content out of the body. This reflex passes through the spinal cord and is essential for immediate continence; it is not innate and is acquired during childhood (hence the normal physiological incontinence of the child before reaching a certain degree of development).

7.5.2 Defecation

When rectal pressure increases, the individual feels the need to evacuate and, at the same time, is informed about the nature of the rectal contents to be evacuated. If the individual accepts this evacuation, there is then a voluntarily sequence of actions: (1) relaxation of the puborectalis muscle opening the recto-anal angle to align the axis of the rectum in the axis of the anal canal and thus facilitate passage of the luminal content, (2) relaxation of the internal and external sphincters, (3) contractions of the rectal wall promoting rectal emptying, and finally (4) voluntary contractions of the abdominal wall against the diaphragm to increase intra-abdominal pressure to help push the stool outward through the anal orifice.

When defecation is not allowed, reverse phenomena occur and the individual has the ability to close the external sphincter, contract the puborectal muscle, etc. If the rectal contents are not expelled, the phenomenon of rectal accommodation is such that the sensation of needing to defecate regress (until next time the rectal volume increases). Refraining from defecation also induces reduction of proximal colonic motility and backflow of distal content.

Knowing this physiology, we understand the importance of the integrity of the muscular system and of the sensory and motor nervous systems in order to secure fecal continence.

7.6 Inflammation Disorders

7.6.1 Rectal Inflammation (Proctitis, Rectal Ulcers)

(a) Proctitis Inflammation of the rectum is called proctitis (less commonly rectitis). In clinical practice, it defines an inflammation limited to the rectum.

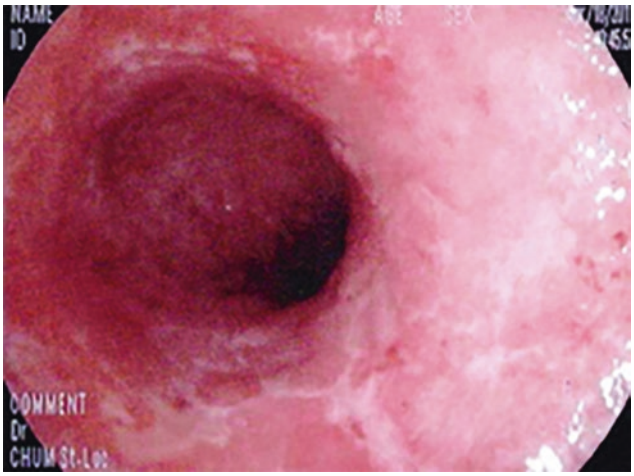


Fig. 7.8 Ulcerative proctitis in endoscopy: the rectal mucosa is erythematous, friable, erosive, covered with mucus or pus. (Photo by P. Poitras)

Symptoms of proctitis include:

- Presence of blood in the stools
- Discharge of mucus or pus, feeling of rectal fullness
- Tenesmus (sensation of frequent urges to have a bowel movement)
- Frequent defecation with false urges, i.e., numerous defecations, often of mucosanguineous secretions with stools in small quantity, which may be either of soft or normal consistency, or even dry and hard as in constipation. In proctitis, there is no classic diarrhea with large amount of liquid stools that could lead to dehydration but rather frequent defecations (q 1 h day or night!) that compromises quality of life. Loss of compliance of the rectal reservoir (caused by inflammation of rectal walls), associated with very distal secretions without any capacity for reabsorption, explains the clinical symptomatology.

The diagnosis of proctitis is obtained by endoscopy (Fig. 7.8) which shows inflammation of the rectum (over a maximum distance of about 15 cm). Depending on the type and severity of the disease, mucosal breaks (erosions, ulcerations, ulcers) can be present.

Types of Proctitis

- *Inflammatory proctitis* may be caused by inflammatory bowel disease (IBD) such as ulcerative colitis (ulcerative proctitis) or Crohn's disease.

When an individual presents with ulcerative proctitis, it is estimated that the disease will remain limited to the rectum in 50–70% of cases; in others, over the following years, it may spread to more proximal areas of the colon and digestive tract.

Ulcerative proctitis is usually treated with local 5ASA or steroid therapies in the form of a suppository (Salofalk® or Pentasa®), foam (Mezera®, Cor-

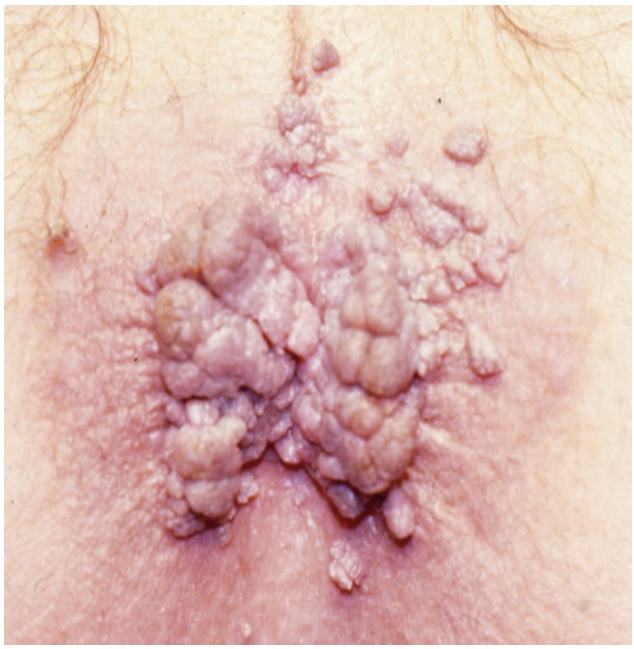


Fig. 7.9 Condyloma (genital warts due to human papilloma virus) of the anus and perianal region. Condyloma within the rectum should be looked for during endoscopic examination. (Photo from D. Bernard)

tifoam®), or enema (Salofalk®, Pentasa®, Cortenema®, Entocort®).

- *Infectious proctitis.* The rectum, during anal intercourse, can be infected by sexually transmitted diseases such as syphilis, gonorrhea, chlamydia (with or without lymphogranuloma venereum), herpes, and papilloma virus (Fig. 7.9).

  Various lesions (diffuse or circumscribed inflammation or ulceration, etc.) can be seen at rectoscopy. Viruses or bacteria can be identified by histological or bacteriological analysis of rectal biopsies. Treatment will depend on the causal organism.
- *Radiation proctitis* is seen mainly after irradiation for prostate cancer. Small caliber vascular damage is seen, rather than inflammatory changes. Rectal bleeding is the most common sign (in some cases, it can be severe enough to justify blood transfusions). Endoscopic treatment (electrocoagulation, etc.) may be required.
- *Chemical proctitis.* Damage to the rectal (and colonic) mucosa can be caused by instillation of enemas containing "irritating" substances such as soap, hydrogen peroxide, ginger, etc. The use of such agents should be discontinued.

(b) Solitary ulcer of the rectum A solitary ulcer of the rectum describes, as its name suggests, an isolated ulcer in an otherwise normal-looking rectum and corresponds to an "ischemic," chronic ulcer associated with a rectal prolapse (to be identified by clinical examination and/or radiological defecography). Histological analysis often reveals proliferation of fibroblasts and smooth muscle fibers in the chorion of the mucosa. Treatment involves correction of the causal prolapse.

The solitary ulcer of the rectum must be differentiated from other unique lesions of the rectum including:

- Cancer of the rectum (to be confirmed by biopsy)
- Crohn's disease (to be confirmed by biopsy and investigation of the digestive tract
- Infectious ulcer such as syphilis, etc. (to be confirmed by histological or microbiological analysis)
- Traumatic ulcer, following the misuse of enema cannulas or other foreign bodies

7.6.2 Anal and Perineal Inflammation (Abscess, Fistulas, Anal Pruritus)

(a) Abscess Anal abscesses are caused by an infection of a gland (Hermann and Desfosses glands) of the anal canal. These glands located in the anorectal wall opening the intestinal lumen at the level of the Morgagni crypts through a main glandular canal. The infection starts in glandular ducts and then proceeds to the inter-sphincteric space. The germ is most often of intestinal origin. In all cases, a primary or internal orifice (which sometimes may be difficult to find) is always present at the level of the pectinate line.

Clinical presentation The clinical picture is dominated by anal or perianal pain. The pain is initially moderate, then increases, and can become severe, insomniant. It is not associated with defecation (as is anal fissure). The pain can radiate to the perineum or the genitals and be associated with painful tenesmus (painful desires to evacuate stools) or dysuria. An infectious syndrome (with fever, etc.) may be present. On perineal examination (Fig. 7.10), the abscess may be visible, consisting of an indurated, inflammatory mass, more or less important, which can erase the plicated folds of the anus; localization is more difficult when the abscess is deeply located, but palpation of the surrounding tissues may reveal abnormal firmness and localized pain. Pus discharge from the primary endoluminal opening may sometimes be noted when examining the anal margin. Digital exam may reveal the same on palpation but, as with endoscopy in this situation, may be difficult because of pain.

Treatment of abscess The ideal treatment is to drain the abscess and treat the associated fistula (see below). Simple drainage of the abscess without addressing the fistula leads to recurrence in 14–40% of cases in randomized

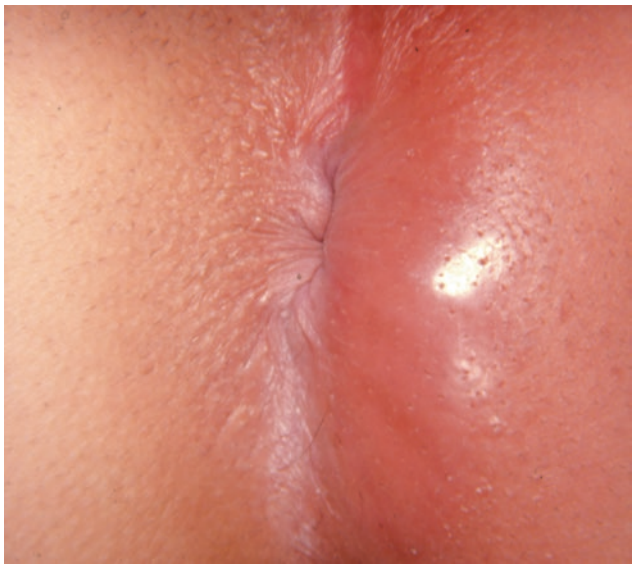


Fig. 7.10 The anal abscess presents as an inflammatory swelling (red, hot, painful, etc.) of the immediate perianal area (here on the right side of the anus). (Photo from D. Bernard)

trials. Treatment should be surgical as often as possible, as antibiotic therapy alone is rarely curative.

Antibiotic treatment of perineal abscess has, curiously enough, rarely been studied in the scientific literature. It is often used (beta-lactams and metronidazole are the most common agents) in presence of systemic infection, significant cellulitis, or in patients with risks of infection extension such as immunocompromised or diabetic patients. Antibiotic treatment (most often ciprofloxacin + metronidazole) also appears to be effective in reducing infection in complicated abscesses of Crohn's disease.

(b) Fistulas A fistula is defined as an abnormal communication between two epitheliums. Most anal fistulas are a complication of an anal abscess which has drained (either spontaneously or through a therapeutic gesture) to the perianal region, spreading along a sinuous path in or through the sphincter apparatus or to the skin (Figs. 7.11, 7.12, and 7.13).

Clinical presentation The fistula may follow the acute phase of an abscess, or it may set in immediately. It is responsible for a flow of serosanguineous or fecal matter to the skin. On clinical examination (Fig. 7.13), there is an external orifice (known as the secondary orifice) located on the skin and allowing secretions to flow out. Palpation of surrounding soft tissues may reveal induration that corresponds to the path of the fistula. There may be several secondary openings, or double fistulas; openings located on both sides of the midline suggest a horseshoe shaped fistula encircling the anus.

Treatment of fistula In all cases, treatment of an anal fistula has two goals: to heal the fistula and to preserve continence. The primary orifice is typically the source of recurrence if the abscess is treated without treating the fistula.

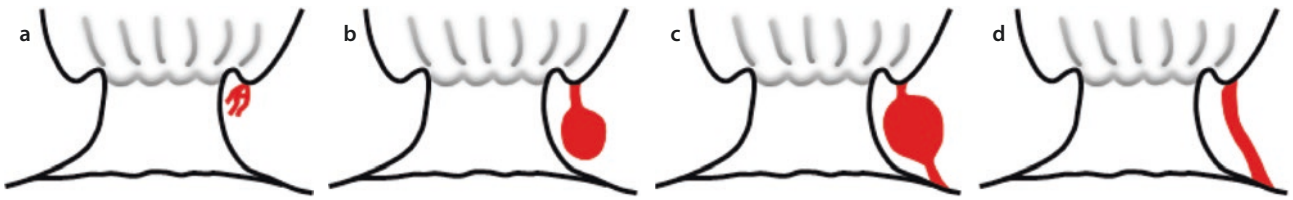


Fig. 7.11 From an internal (primary) opening **a**, an anal gland becomes infected and forms an abscess **b**; the abscess drains by (secondary) opening to the skin **c**, leaving a fistulous path between the internal and external openings **d**

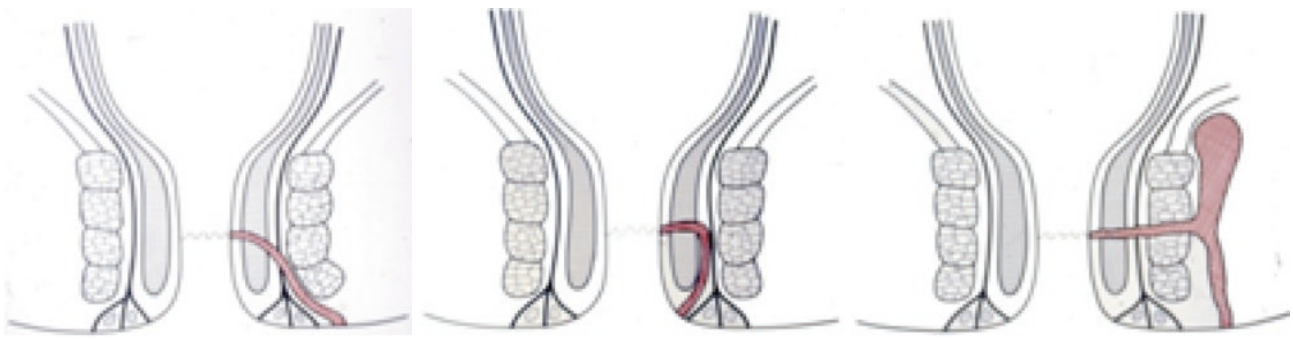


Fig. 7.12 Different fistula paths are possible: sparing the sphincters (two illustrations on the left) or trans-sphincteric (illustration on the right)

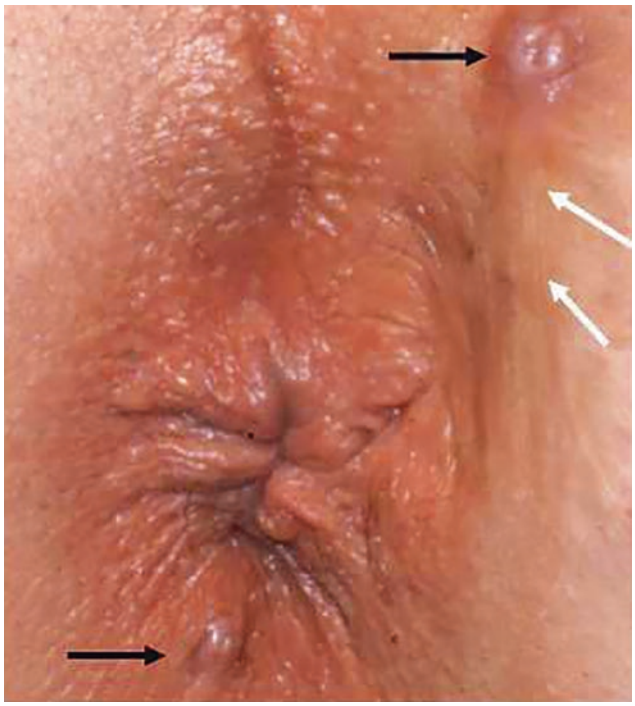


Fig. 7.13 Fistulous openings (black arrows) around the anus; a fistulous path (white arrows) is visible from the anus to the opening. (Photo from D. Bernard)

Conventional treatment is fistulotomy, the techniques of which vary according to the height of the fistula and its path, since treatment must be performed with minimal sphincter trauma. There are also some surgical alternatives whose main goal is to avoid extensive sectioning of the sphincter muscle. In some complex cases, sphincter reconstructions may be required.

(c) Perineal necrotizing fasciitis (or Fournier's gangrene) is an acute bacterial infection with extensive necrotic infection of perineal soft tissues. It is an emergency, both medical and surgical. It most often arises as a complication of an operation in the area (anal surgery, etc.) or a soft tissue trauma in patients with diabetes, alcoholism, immunosuppression, etc.

Germs involved are usually bacteria from the normal anorectal flora (aerobic or anaerobic, including *Escherichia coli*, *Streptococcus*, *Staphylococcus*, *Proteus*, *Bacteroides fragilis*, or clostridiums). The first clinical signs are a localized, constant, and intense perineal pain associated with a septic syndrome that quickly becomes severe. Clinical examination reveals necrotic skin damage. On palpation, a crepitation can be felt, indicating subcutaneous emphysema by air-producing bacteria. The infection may spread rapidly throughout the perineum and upward to the abdomen or lumbar regions or downward to the thighs.

Medical treatment of necrotizing fasciitis combines broad-spectrum antibiotic therapy (a third-generation cephalosporin combined with an aminoside and metronidazole) and surgical treatment with extensive (often mutilating) debridement, instituted rapidly and which may need to be repeated to remove all necrotic tissues. Colostomy is often necessary, and some people use hyperbaric oxygen therapy to treat this gangrene whose mortality, despite treatment, has been estimated at 16%.

(d) Pilonidal disease The pilonidal sinus is a pseudocystic cavity of the subcutaneous tissue that communicates with the skin through a duct that opens into small orifices on the midline of the gluteal cleft (butt crack). This cavity does not have hair follicles but contains hairs; it can become infected and produce an abscess that will fistulize to the skin. It is not strictly speaking a digestive nor an anorectal pathology.

Pathogenesis The congenital theory has long been predominant and considered these cavities as embryological scars. However, the acquired theory is now generally accepted, especially since the description of Jeep Disease during the Second World War, when nearly 79,000 Jeep drivers of the US Army were hospitalized for pilonidal disease, which is therefore considered an inflammatory reaction secondary to the penetration of a hair in the gluteal furrow with secondary suppuration promoted by various phenomena such as friction (prolonged sitting, etc.).

Clinical An inflammatory and painful swelling is present in the gluteal fold. The swelling may resolve spontaneously or may progress to release pus and pilar content (corresponding to the fistulization of the abscess). The drainage may flow through one of the median orifices, or sometimes through lateral fistulization orifices, and may occur chronically or repeatedly.

Treatment Treatment of a pilonidal sinus is surgical to prevent recurrence.

(e) Hidradenitis suppurativa (HS, Verneuil's disease, acne inversa) is a skin disease that can be fistulizing and may be mistaken for perineal Crohn's disease. In fact, it is a hyperkeratinization of hair follicles with a secondary infectious process (by bacterial colonization).

HS is a rare infection (prevalence of 0.3%) and more frequent in women than in men (4/1), as well as in Crohn's disease.

Clinical presentation HS lesions are located mainly in underarms, under the breasts and groin areas.

The elementary lesion consists of a hypodermal nodule which is generally hard on palpation, well-limited, and mobile with deep planes. This nodule may be isolated or associated with several nodules forming growths and/or scars on the skin. Most often, one of these nodules will open to the skin with a seropurulent discharge.

The disease will evolve with subcutaneous extension and the appearance of new localizations forming new nodules with more and more suppurative orifices communicating with each other. These cutaneous suppurations and fistulas may be suggestive, when occurring in perianal area, of fistulizing Crohn's disease.

Treatment Antibiotics do not treat the disease but may be helpful to prevent progression of infection. The treatment is surgical and based on resection of the lesions with the widest and earliest possible excision. TNF inhibitors (e.g., adalimumab) have a positive effect on HS lesions and are a more and more popular treatment.

(f) Anal pruritus Itching sensation in the anal or perianal region that causes the patient to scratch (generally with traumatic erosions that promote licking of the skin, which itself is a source of pruritus) is a common reason for consultation. Its prevalence is poorly known, but it is thought to affect about 1–5% of the population, with a male prevalence of 4 to 1.

Causes for anal pruritus are numerous:

- Infectious: *Candida albicans* (favored by antibiotics, diabetes), bacteria (group A beta hemolytic *Streptococcus*, *Corynebacterium minutissimum*), virus (human papillomavirus, herpes), and parasites (pinworm) are causes of skin infection with anal itching.
- Inflammatory: Contact dermatitis or eczema most often related to toiletries (e.g., soap or other hygiene products) or to fecal contamination (sticky stools, lack of hygiene, etc.) is common. Psoriasis should be considered in presence of red, squamous lesions, often with lesions at distance in other locations. Shiny whitish papules in women may be due to lichen sclerosus present in the anal area but also in the vulva.
- Tumoral: Bowen's disease (squamous cell carcinoma of the epidermis) and Paget's disease (intraepithelial adenocarcinoma of the epidermis) are possible. Resistant to topical steroids, these lesions are usually infiltrative, keratotic, and inflammatory.
- Proctologic: Conditions with anorectal suppurations such as fistulas, abscesses, Verneuil's disease, pilonidal cyst, anal fissure, and rectal prolapse may promote pruritus. Hemorrhoidal prolapse or oozing can cause pruritus, as can skin tags due to difficulty in maintaining good hygiene. Hemorrhoids may be responsible for 20% of itching in adults.
- Idiopathic: a quarter of cases remain unexplained. A psychological origin (with neurodermatitis) is frequently suspected. Excessive or aggressive hygiene of the perianal area may also cause or maintain pruritus in some patients.

Management of pruritus Extensive investigations are rarely necessary at first consultation. A good clinical examination should be able to identify the main causes; a "Scotch tape" test for pinworms and/or swabs for bacteriological, mycological, or virological examinations can be considered.

Treatment of anal pruritus Hygiene rules are of primary importance and should insist on keeping the perianal area clean and dry. Cleanliness should be best achieved without soap or antiseptic products and only with water. Wiping should be done with a soft paper, ideally slightly moistened and without rubbing. Drying should be done with a towel without rubbing. Creams and intra-channel cleaning after defecation should be avoided. No diet regimen is known to be effective.

Medicated treatment depends on the cause. Antifungal ointments or creams (nystatin, miconazole, etc.) or antibiotics (metronidazole, bacitracin, etc.) can be applied locally in case of superinfection. Test treatment against pinworms can be undertaken (mebendazole 100 mg, two doses 14 days apart). Some authors have proposed type 1 antihistamines for predominantly nocturnal pruritus. When acute dermatitis is present, dermatologic advice may be required, and a small dose of a topical steroid (hydrocortisone 0.1%) may be used; it is, however, best to avoid the long-term use of these creams, which in the long run lead to skin atrophy and can make the condition even more difficult to treat. Recently, capsaicin ointment (topical analgesic) has been reported to be effective.

In children, perianal erythema is a frequent reason for consultation; it may involve specific pediatric conditions:

- Perianal *Streptococcus* A dermatitis is a lesion starting from the anus and extending into the perianal region for 2–3 cm. The erythema is bright red with clear borders, sometimes associated with edema or oozing. Local swabbing with a bacteriological culture swab confirms the diagnosis. The treatment consists of antibiotic therapy directed against *Streptococcus* A.
- Diaper psoriasis is characterized by an intense, varnished, sometimes dry and scaly erythema, which is very limited in infants with diapers. Superinfection or maceration often makes the diagnosis difficult. The existence of distant psoriasis lesions and of family history helps in the diagnosis. It is probably a reaction triggered by any other cause of diaper rash

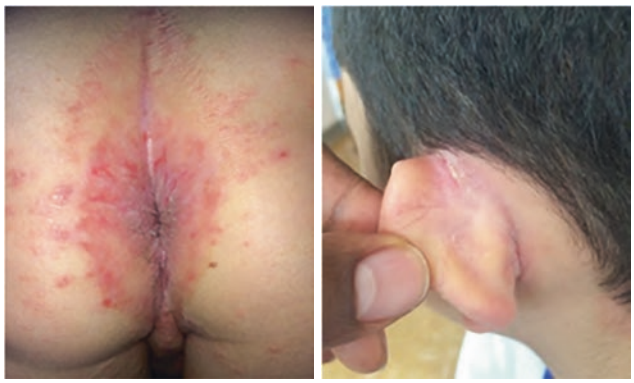


Fig. 7.14 Perianal and retroauricular psoriasis lesions. (Photos from P. Jantchou)

- in children with a psoriasis trait. The treatment is based on topical steroids and skin emollients.
- Anti-TNF induced psoriasis is a complication increasingly observed in children treated with these biologic drugs (infliximab, etc.). Perianal involvement is manifested by an intense, dry, scaly, pruritic erythema, sometimes associated with micro-ulcerations (scratching lesions). Extra-perineal lesions, particularly in retroauricular areas, may be present (Fig. 7.14).

7.7 Tumor Disorders

7.7.1 Rectum Cancer

- *Adenocarcinoma* of the rectum is a form of colorectal cancer (discussed in ▶ Chap. 4). Symptoms of rectal adenocarcinoma include rectal bleeding, stools of reduced diameter (pencil-shaped), dyschezia, or pelvic discomfort.

Any of these symptoms should prompt a digital rectal examination, since many rectal tumors are located distally enough to be felt on digital palpation. Endoscopy with biopsies will confirm the diagnosis (Fig. 7.15).

Progression of rectal adenocarcinoma may lead to metastasis through the lymphatic and venous drainage systems of the rectum, which can lead to locoregional spread to pelvic and inguinal lymph nodes, or more distantly to the liver but also to the lungs due to venous drainage bypassing the portal circulation.

Treatment of adenocarcinoma of the rectum is surgical and almost always combines preoperative (neoadjuvant) radiotherapy and chemotherapy. Surgical therapy is complex because of the desire to fully and uncompromisingly resect the tumor while preserving normal bowel function (i.e., keeping the anal sphincter intact and avoiding a permanent colostomy), as well as avoiding damage to other pelvic mechanisms (including erection, urinary function, etc.). Colorectal and even colo-anal anastomosis techniques, combined with radiotherapy, now make it possible, in a large number of cases, to avoid proctectomy with definitive colostomy that was previously performed in these cases.

- *Neuroendocrine tumors* (*NET*) can be found in the rectum. They are most often nonsecreting.

7.7.2 Anus Cancer

Squamous cell (epidermoid) carcinoma of the anus is very frequently associated with the human papillomavirus (HPV) responsible for condylomas (and cervical cancers in women) and is therefore more similar to genital neoplasia than to other digestive cancers. Among the more than 40 known types of HPV, types 16 and 18 are more frequently associated with neoplasm. Epider-

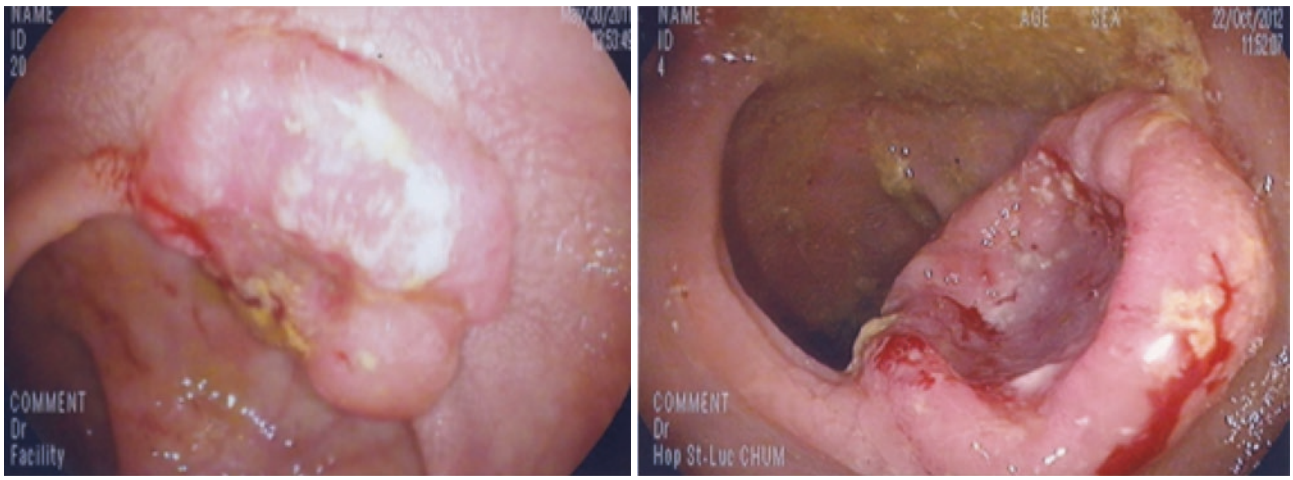


Fig. 7.15 Malignant polyps seen during rectal endoscopy. (Photos by P. Poitras)

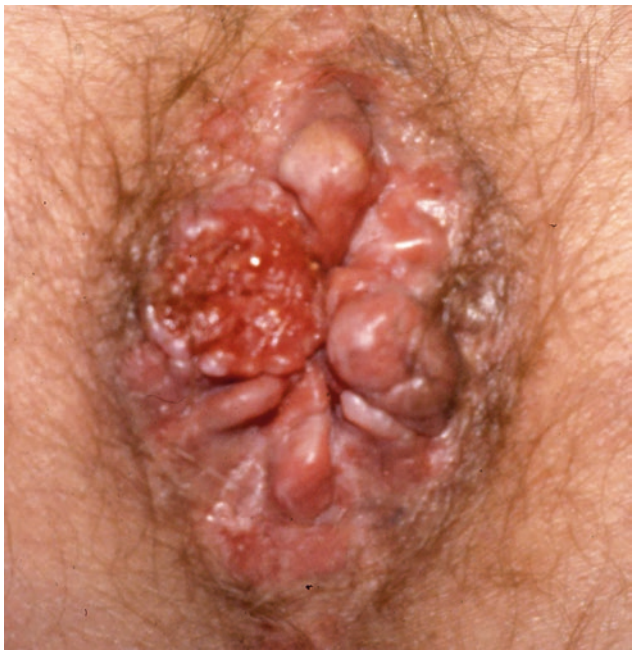


Fig. 7.16 Epidermoid neoplasia of the anus presented here as an ulcerated lesion. (Photo from D. Bernard)

moid cancer of the anus (Fig. 7.16) can reach the anal canal as well as the perianal skin region. It presents with rectal bleeding (40% of patients) and sensation of anal pain or mass (30%) and also may be found in asymptomatic subjects.

Tumors of the anal canal were previously treated by proctectomy and colostomy. Treatment with radiotherapy and chemotherapy is now preferred with a 5-year survival of 70–90%.

Perianal cancer, which is rarer, is most often presented with an itchy or eczematous lesion. Perianal tumors are considered and treated as skin lesions.

Bowen's disease is a dysplasia of the epidermis without infiltration of the dermis and corresponds to in situ squamous cell neoplasia related to HPV virus. It may present as an erythematous plaque suggestive of psoriasis and has a good prognosis.

Paget's disease (extramammary) is an intraepithelial adenocarcinoma that originates from an anal gland and most commonly presents as an eczematous plaque of the anal margin. Its prognosis is poor considering its invasive and/or metastatic potential.

Cloacogenic carcinoma is a rare (2% of epidermoid lesions) tumor originating from a persistent remnant of the embryological cloacal membrane located in the transitional zone 1 cm above the dentate line.

Melanoma is rare (1% of anal tumors) but can occur.

7.8 Function Disorders

7.8.1 Proctalgia Fugax

Proctalgia fugax is a painful syndrome in the anus or lower rectum. It is a cramp-like pain that appears suddenly, at first of maximum intensity, and lasts for few seconds or minutes (less than 5 min in 90% of people), and then disappears quickly without leaving any residual discomfort between attacks. These paroxysmal pains are recurrent (more than five attacks/year in 50% of patients). They often occur during the night and can of course wake the patient up; they can also appear after certain situations such as defecation or sexual intercourse or spontaneously. Proctalgia fugax is common (10–20% of people are said to suffer from it) but may be underreported by patients who feel ashamed to talk about it.

Pain may be associated with classic neurovegetative manifestations of pain such as pallor, sweating, nausea, and faintness. Often attributed to spasms of the anal muscles, the physiopathology of this pain is not known. Its acute and brief cramp-like nature is reminiscent of calf muscle cramps.

Other causes of ano-rectal pain (e.g., hemorrhoids, fissure, abscess, etc.) must be excluded, but in most cases, with typical episodic pain, investigation other than digital rectal examination is rarely necessary.

The treatment is not well codified. Postural techniques have been identified spontaneously by patients, such as sitting with flexion of the lower limbs but sometimes also gas or stool emission or digital rectal introduction. Medications (aiming to relax the anal sphincter) are rarely effective for this brief pain (because of their delayed onset of action); topical or sublingual nitroglycerin or inhaled salbutamol may possibly help. Baths in warm water have been suggested, as well as relaxation techniques (self-hypnosis, autogenic training, etc.). Patients are often worried about this sharp pain and may be ashamed to talk about it; reassurance is crucial for this frequent and typically benign condition.

7.8.2 Levator Ani Syndrome

Levator ani syndrome is a painful syndrome in the upper part of the anal canal, with a sensation of heaviness or sometimes of an intrarectal foreign body. The pain may extend to the genital or urinary sphere, being sometimes accompanied by dysuria; it may also radiate posteriorly in the gluteal fold. The pain is moderate, prolonged

(more than 20 min; in contrast with the brief pain of proctalgia fugax), with a tendency to worsen over the course of the day. Like proctalgia fugax, the pain may be promoted by certain events such as sexual intercourse or after defecation.

This pain is attributed to chronic or prolonged tension or spasm of the levator ani muscle, particularly of the pubococcygeus bundle. On examination, digital palpation and posterior traction of the levator ani muscle are often painful. Manometry, which is not essential, may reveal hypertonia of the anal canal. Pelvic examinations (CT scan, MRI, sigmoidoscopy, etc.) may be required to eliminate visceral or other lesions.

Treatment remains empirical for this rare condition. Digital massages of the levator ani muscles have been reported to be effective, as well as medical treatments with muscle relaxants (cyclobenzaprine, diazepam) or relaxation techniques (hot baths, etc.).

7.8.3 Anorectal Dyssynergia/Anismus

Anorectal dyssynergia is a paradoxical contraction (or lack of relaxation) of the external anal sphincter during defecation. Fecal evacuation must therefore take place through expulsion thrusts fighting against an obstacle made by an abnormally high sphincter resistance. This asynchrony in the defecation maneuver can lead to dyschezia (difficulty in evacuating stool, with exaggerated defecation efforts, use of digital facilitation, etc.), sensation of incomplete rectal emptying after defecation, or constipation.

There are two main causes of anorectal dyssynergia: neurological causes are rare but can involve spinal or root damages (cauda equina syndrome, multiple sclerosis, pudendal nerve trauma, etc.). In the vast majority of cases, a behavioral cause is identified. A high frequency of sexual abuse is found in anismus (which is a sensitive but nonspecific marker of sexual abuse). The term anismus should be reserved for behavioral causes of anorectal dyssynergia.

Clinically, anorectal dyssynergia is suspected constipation, especially if accompanied by difficulties in stool evacuation. During physical examination, at the time of the digital rectal examination, the examiner may ask the patient to push in order to simulate defecation; if during this test there is no paradoxical contraction perceived with the finger, this eliminates, with good sensitivity, the possibility of having anismus. Anorectal manometry is the ultimate test to reveal paradoxical contraction of the external sphincter during defecation; inability to expel a water-filled balloon is usually present as well.

First-line treatment is typically that for constipation. If unsuccessful, reeducating the defecation maneuver with the help of physiotherapy or biofeedback may be used.

7.8.4 Hirschsprung's Disease

Hirschsprung's disease is due to a defect in the migration of neural crest cells (precursors of intestinal ganglion cells) during embryonic gut development as discussed in ▶ Sect. 7.3.3. The absence of ganglionic myenteric plexus most often affects the rectosigmoid region (80% cases) but can extend to the entire colon and sometimes even to the small intestine (5% cases). Aganglionosis causes permanent contracture without relaxation of the affected areas and results in distension of the normal intestine upstream of this functional obstacle (hence the name congenital megacolon).

The condition is usually suspected in a newborn with intestinal occlusion and without passage of meconium and/or stool. The diagnosis can sometimes be suspected later in a constipated child or even (rarely) in adulthood.

The diagnosis is suspected by a barium enema X-ray that shows colonic occlusion on a contracted distal (usually rectal and lower sigmoid) segment. Anorectal manometry demonstrates the absence of the recto-anal inhibitory reflex (reflex relaxation of the internal anal sphincter during rectal distension by a balloon). Rectal biopsy confirms the absence of neurological ganglionic cells.

Surgical treatment is required, in early life, to resect the aganglionic intestinal segment and bring the normal intestine to the anal sphincter, to allow a future life without colostomy and as normal as possible.

7.8.5 Fecal Incontinence

Anal continence requires four conditions: (1) solid stool consistency (liquids stools are more difficult to contain), (2) good rectal compliance (to act as a reservoir for temporary holding of waste material present in the rectum), (3) strong sphincter muscles (to effectively close the exit aperture), and (4) effective regulatory nerves (to control muscle activity).

Fecal incontinence is discussed extensively in ▶ Chap. 18.

7.9 Miscellaneous

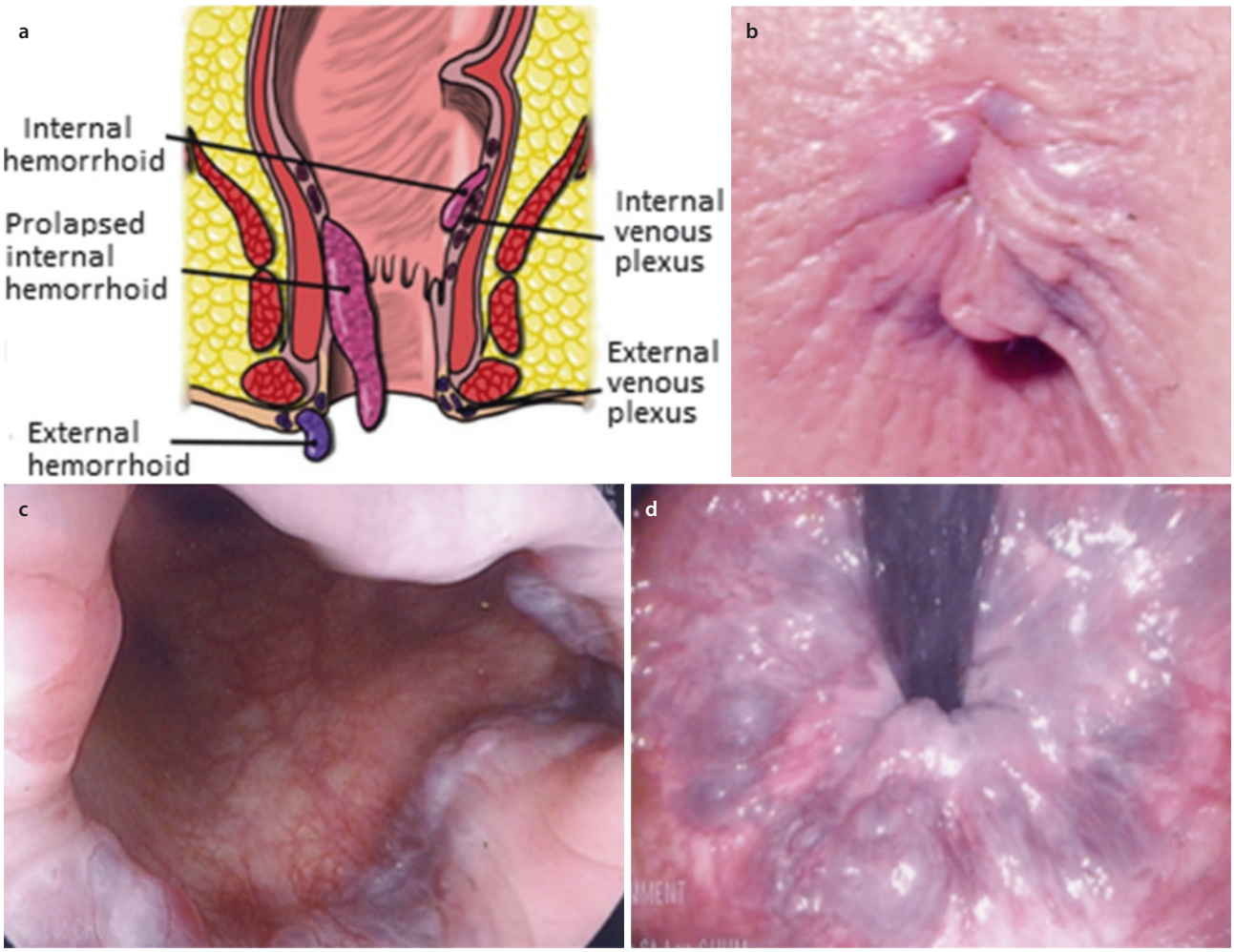
7.9.1 Vascular Diseases: Hemorrhoids/Varices

(a) Hemorrhoids The prevalence of hemorrhoids is not well-known and varies from 4.4% to 86%, depending on the studied population. Hemorrhoidal pathology (i.e., hemorrhoids with symptoms) typically appears in the third decade and increases with age, peaking between the

ages of 45 and 65. No ethnic or gender differences are seen. Factors contributing to hemorrhoidal disease include family history, constipation, and, in women, pregnancy and delivery; hormonal factors may be involved (especially since estrogen receptors are present in hemorrhoidal tissues), but mechanical factors are obvious (e.g., pressure of the fetus on pelvic veins limiting venous drainage of hemorrhoids during pregnancy, or baby delivery which is accompanied, one out of three times, by hemorrhoidal thrombosis).

Hemorrhoids are normal vascular plexuses located in the anus and present from birth (although poorly developed before the age of 10). Internal and external hemorrhoids are identified (see ◘ Fig. 7.17). External hemorrhoids are located below the pectinate line, at the epidermal anal margin, and their vascularity depends on the internal pudendal artery (a branch from the internal iliac artery). Internal hemorrhoids are located in the submucosal space of the anal canal above the pectinate line. They are classically arranged in three bundles emerged from branches of the superior rectal artery. The venous blood return from hemorrhoids is provided by the upper, middle, and lower rectal veins to abdominal portal circulation and to systemic inferior vena cava circulation. Hemorrhoidal plexuses are histologically made up of collagens, sinusoid vessels, and arteriovenous shunts sometimes including a corpus cavernosum structure. Internal hemorrhoids are normally fixed inside the anal canal against the internal sphincter. Sensitive receptors are present on hemorrhoids, including mechanoreceptors and thermoreceptors, which are essential for discrimination of rectal contents (gas, liquid, solid).

Physiopathology The physiopathology of hemorrhoidal disease is explained by two theories, one vascular and one mechanical. The vascular theory is based on the existing arteriovenous capillary-type shunts that could undergo



◘ **Fig. 7.17** Hemorrhoids: **a** schematic illustration; **b** external hemorrhoids on visual examination of the anus; **c** internal hemorrhoids on endoscopic examination (direct vision), **d** seen under retroflexion (black endoscope in the anus and retroflexed in the rectum). (Photos by P. Poitras)

significant variations in arterial flow under the influence of variable factors such as pregnancy or childbirth. The mechanical theory considers a degradation (with age) of supporting connective tissues as the essential cause for the appearance of rectal bleeding and prolapse of the hemorrhoidal tissue over time.

Clinical The limit between physiological hemorrhoids and hemorrhoidal disease is often thin. Hemorrhoidal disease is characterized by symptoms related to hemorrhoids.

- Hemorrhoidal thrombosis (Fig. 7.18) is the most important manifestation of *external hemorrhoids* with sudden onset of anal pain associated with swelling and a blood clot visible under the skin. The pain is continuous, non-pulsating and not related to defecation (in opposition to anal fissure pain). Without treatment, the pain will typically progress, in 1–5 days, toward the disappearance of the pain and of hemorrhoid swelling. During spontaneous healing, external hemorrhoid thrombosis can leave a fold of skin called skin tag which, most often, is not a medical problem but may be responsible for aesthetic discomfort or pruritus due to difficulty in cleaning. If treated early, external hemorrhoidal thrombosis can benefit from excision of the skin to remove the clot, resulting in immediate relief.
- Rectal bleeding is the most common manifestation of *internal hemorrhoids*, made of bright red blood occurring with defecation. Blood may be on the stool, on paper when wiping, or sometimes dripping after defecation. The rectal bleeding is painless and stops spontaneously, although, in rare cases, chronic bleeding may be responsible for anemia.
- Hemorrhoidal prolapse is a situation where externalization of internal hemorrhoids outside the anal canal occurs. It can happen with a single hemorrhoid or with several hemorrhoids. Prolapse most often occurs during defecation and is more common in chronic constipation with straining efforts. The prolapse may be spontaneously retractable, require manual reinsertion after a bowel movement, or be permanent. It may be associated with other symptoms such as oozing, bleeding, discomfort, and pruritus. It can sometimes resemble a rectal prolapse (having a circular groove, while the hemorrhoidal prolapse has radiating grooves).

  Hemorrhoids are classified into four stages: stage I, simple hemorrhoids with rectal bleeding and without prolapse; stage II, hemorrhoids prolapsing at defecation with spontaneous reintegration; stage III, hemorrhoids prolapsing during bowel movements and requiring manual reintroduction; and stage IV, hemorrhoids with permanent prolapse.
- Pain other than thrombosis: Hemorrhoids are rarely painful but may cause discomfort, sensation of anal heaviness, which is particularly exacerbated during the premenstrual period in women. There is no particular treatment for this symptom.
- Skin tags (Fig. 7.19) are sequelae of hemorrhoidal thrombosis, as are hypertrophic papillae that may be associated with it and may persist after hemorrhoidal

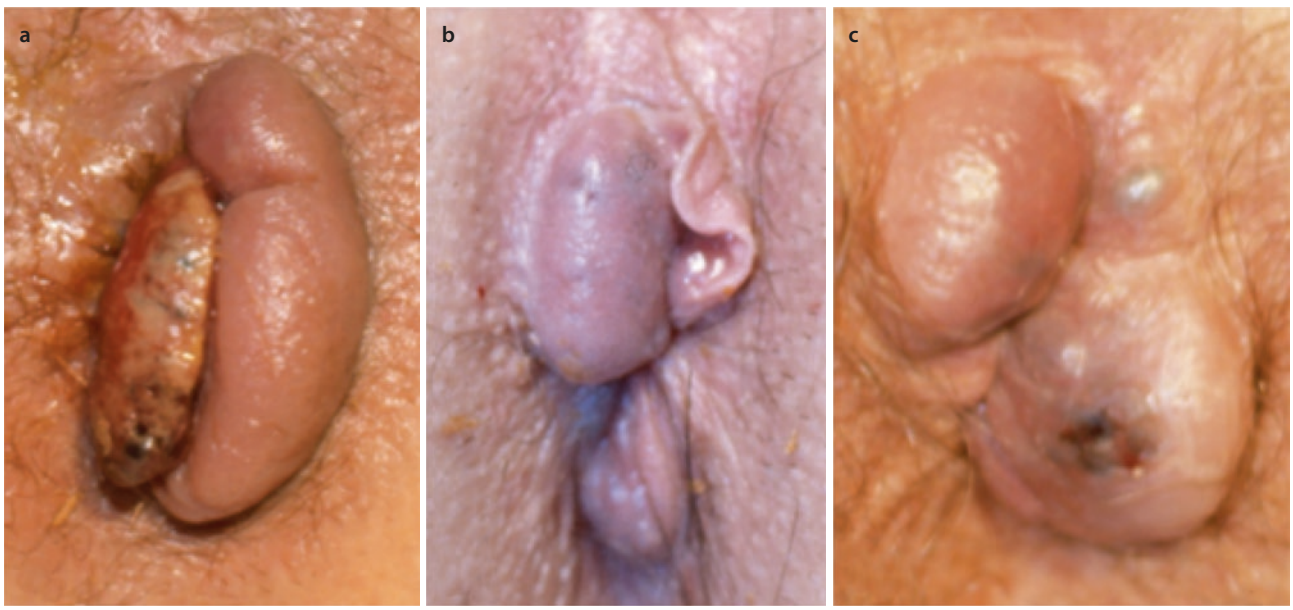


Fig. 7.18 Thrombosed hemorrhoids: **a** necrotic internal hemorrhoid (ischemic yellowish coloration; left side of anus) and edematous external hemorrhoid (right side of anus). **b** Thrombosed external hemorrhoid **c**, thrombosed hemorrhoid with spontaneous rupture that has caused partial evacuation of the clot. (Photos from D. Bernard)

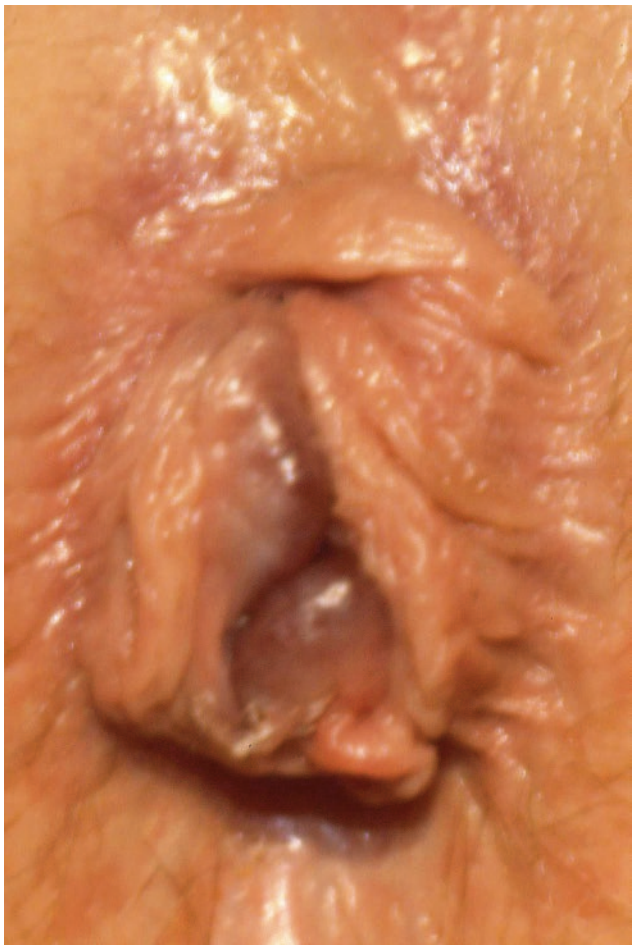


Fig. 7.19 Skin tag: a skin flap at the outer part of the anus (sequelae of an old hemorrhoidal thrombosis) surrounding hemorrhoids. (Photo from D. Bernard)

thrombosis. These may cause local discomfort, either aesthetic or due to difficulty in maintaining proper hygiene. They can be surgically resected, but any surgery in this area should be considered with caution since it can be very uncomfortable and can be complicated by infection, sometimes very serious.

- Anal fissure is not uncommon with hemorrhoids. The fissure syndrome is often clinically predominant and is not a complication of hemorrhoids.

Treatment Hemorrhoid treatment is only indicated if the hemorrhoids are symptomatic:

- Topical ointments or creams, although very popular, have not proven to be effective in the treatment of hemorrhoidal disease. Oral diosmin (from *Citrus sinensis* fruit; with antiinflammatory and venotonic properties), popular in Europe, is now available in Canada.

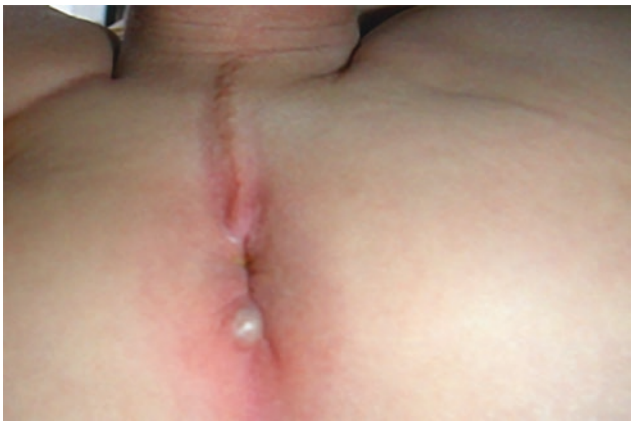


Fig. 7.20 5-year-old boy with an anal venous dilatation due to constipation. (Photos from P. Jantchou)

- Hemorrhoidal thrombosis is treated with analgesics and anti-inflammatory drugs during the painful period. During the first 48 h, incising the hemorrhoid to release the clot is a simple procedure, performed under local anesthesia, which provides immediate pain relief.
- Hemorrhagic hemorrhoids can be treated with a variety of instruments. The most commonly used and effective is the rubber-band ligation; this is a simple procedure performed on an outpatient basis and does not require sedation or preparation. Infrared photocoagulation or sclerosing injections are less popular.
- Surgical treatment by hemorrhoidectomy or hemorrhoidopexy (10% of symptomatic hemorrhoids) is used when other therapeutic treatments have failed, or with stage IV or III hemorrhoids.

In pediatrics, hemorrhoidal pathology is almost nonexistent in children under 10 years of age due to the poor development of the hemorrhoidal plexus before this age. It occurs only in situations of portal hypertension, a context in which rectal varicose veins can be observed (see below). Venous dilatations related to excessive abdominal straining in chronic constipation can sometimes be mistaken for hemorrhoid (Fig. 7.20).

(b) Rectal varices Rectal varicose veins are dilatations of the rectal veins. Diagnosis is made by rectal endoscopy, which easily sees non-pulsatile venous cords (Fig. 7.21). They are usually diagnosed in cirrhotic patients with portal hypertension. Complications, although rare, are dominated by hemorrhage, the treatment of which is not clearly codified but may include treatment of portal hypertension by TIPS or other means (see ▶ Chap. 8).

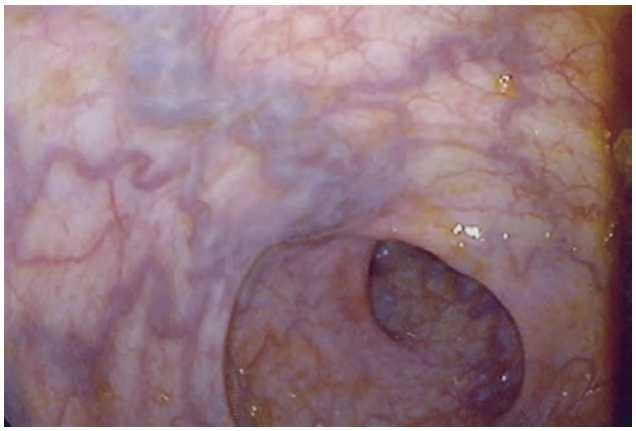


Fig. 7.21 Rectal varices (dilated varicose veins) seen during endoscopy. (Photo by P. Poitras)

7.9.2 Anal fissure

The anal fissure is a longitudinal tear in the mucosa of the anal canal, most often on its posterior side (Fig. 7.22).

Pathophysiology The pathophysiology of anal fissure is not fully elucidated, but it is probably multifactorial.

A "mechanical" factor is usually the initial triggering factor. It is often a trauma related to a hard or bulky stool, which results in tearing the anoderm, often at the posterior commissure of the anus, an area of weak anatomical resistance and relatively poor vascularization.

The "sphincter" factor is related to high-pressure contraction of the internal sphincter, which could be a primary factor in fissure creation, or could be secondary to the pain induced by the fissure, as well as to the stress or anxiety of new stools generating this pain.

The "vascular" factor is related to the fact that the posterior commissure of the anus is an area with limited blood supply and that hypertonia of the internal sphincter can limit blood flow further, all of which lead to delayed or incomplete healing of the fissure. One treatment option, lowering sphincter pressure by surgical sphincterotomy, results in an improvement in sphincter blood flow, thus promoting healing of the fissure.

Clinical Three elements of the diagnosis are linked in a variable manner:

- The fissure is an epithelial ulceration that sits in the anus, usually in the shape of a paper cut or of a tennis racket (with the widest part at the exterior margin and the handle tapering upward in the anal canal). It is located posteriorly in the majority of cases.
- Sphincter contracture results in a hypertonic anal canal and makes digital rectal examination difficult.

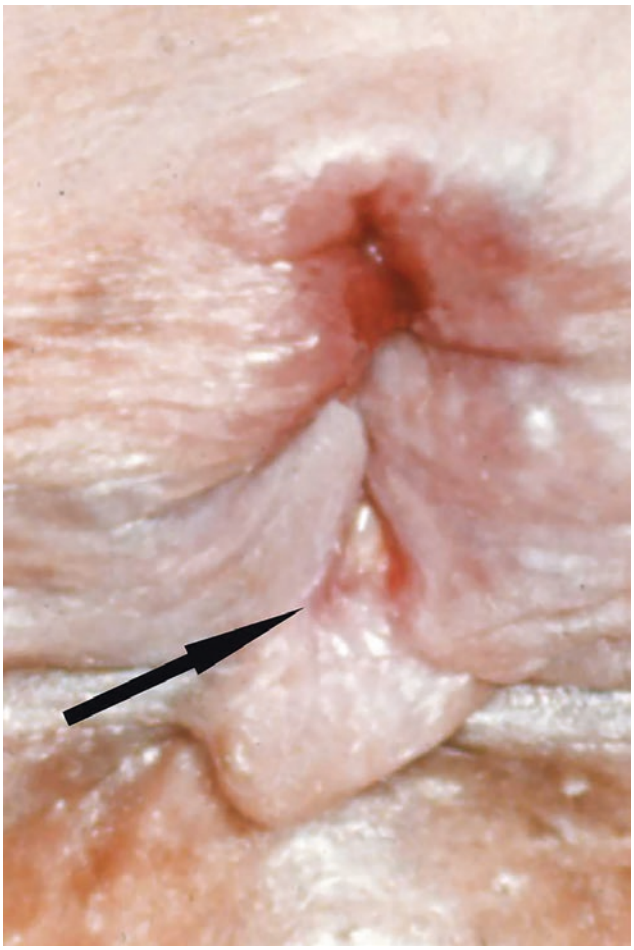


Fig. 7.22 Anal fissure (indicated by the arrow) is a longitudinal, most often posterior, tear in the anal mucosa. (Photo from D. Bernard)

- The pain syndrome typically evolves in three stages: (1) pain is triggered at the time of defecation by the passage of stools, (2) and then there is a decrease or even disappearance of this pain for a few minutes, and (3) finally, there is a secondary reappearance of the pain, often prolonged.

With the passage of the stools, bright red rectal bleeding (in the toilet or on toilet paper) is frequent.

Diagnosis of anal fissure It is most often detected by visual examination of the anus (by spreading the buttocks apart and asking the patient an abdominal thrust to push the anal sphincter out).

Anal fissure must be differentiated from tumor lesions (epidermoid or adenocarcinoma), inflammation (Crohn's, etc.), infection (syphilis, herpes, chlamydia, etc.), or skin lesions (eczema, etc.).

The fissure can be *acute*. The ulceration is superficial, the crater floor is pinkish, and the edges are thin and barely raised; on examination circular fibers of the inter-

nal sphincter may be seen. The pain is usually intense, with its characteristic three-step pattern. The contracture of the internal sphincter is palpable, and rectal examination is almost impossible. This type of fissure can heal spontaneously. The risk of recurrence is significant, and it can also evolve into a chronic (nonhealing) anal fissure.

In the case of *chronic* fissure, the ulceration is deep, with elements of fibrosis around the fissure (raised and sclerotic edges, hypertrophic papilla close to the pectinate line, skin tag on the cutaneous side). Spontaneous healing rate occurs in only 30% of cases.

Treatment of anal fissure Medical treatment is the first-line treatment since it is safe and effective in up to 85% of cases. However, recurrences are frequent (around 50%). Medical treatment consists of:

(a) Regularize stool transit and avoid constipation or large, hard stools that cause anal trauma with tearing. "Soft" (non-stimulant) laxatives are often helpful (fibers (psyllium), emollients (mineral oil), osmotic agents (PEG)) to generate stools of soft consistency.
(b) Decrease pain with oral pain relievers (acetaminophen or NSAIDs, but avoid narcotics that will aggravate constipation). Suppositories or ointments for analgesic purposes, particularly those with a xylocaine base, can be used for a short period of time.
(c) Relax the anal sphincter (to promote tissue oxygenation and healing) through:
   - Sitz baths with lukewarm water (three to four times/day).
   - Nitro derivatives (nitroglycerin 0.2–0.4% in ointment qid) induce the release of nitric oxide (NO), a neuro-mediator relaxing the internal anal sphincter. Side effects of nitrate derivatives, including headache, lead to discontinuation of treatment in more than 30% of cases.
   - Calcium channels blockers (in ointment or gel, such as nifedipine 0.2% bid or diltiazem 2% bid) for 8 weeks are better tolerated than nitrates and give good results in 75–90% of cases.
   - Botulinum toxin (Botox) is a neurotoxin that induces muscle relaxation (presynaptic blocking of acetylcholine at the neuromuscular junction) and can be injected into the anal sphincter. Its long-term efficacy is controversial.

Surgical treatment is indicated in cases of fissures that are resistant to medical treatment or are recurrent after medical treatment. Lateral leiomyotomy (or sphincterotomy) is still the reference technique. It consists in a partial section of the internal sphincter to reduce sphincter tension and thus promote vascularization and oxygenation of anal tissues to heal the fissure. Results are 90% successful. However, its inconvenience is the potential appearance of incontinence, particularly gas incontinence (up to 15–40% of cases).

7.9.3 Neurological Diseases with Anorectal Impact

Neurological diseases can have repercussions on anorectal function.

After a *stroke*, fecal incontinence symptoms are present in 20% of patients after 6 months. Many cofactors can contribute to fecal incontinence, such as age, diabetes, pre-stroke neurological status, injured brain territory, etc.

In *multiple sclerosis*, fecal continence, intestinal transit disorders, and bladder and bowel dysfunction are present in almost half of cases, and these disorders may be presenting symptoms of the disease in 1/3 of cases.

In *Parkinson's disease*, constipation is frequent. Continence disorders are more rare.

In *spinal cord injury*, the neurological alteration can be complex since the lesion may be complete or incomplete, affecting sensory or motor pathways, autonomic or somatic innervation. In complete spinal cord lesions, where supraspinal control of the sensory-motor functions is absent, sphincter dyskinesia affecting both urinary and anal organs may impair evacuation function. In high spinal cord lesions, constipation, due to reduced colonic motor activity, is often the main symptom. Lower spinal cord injuries, as in cauda equina syndrome, may reduce rectal perception of need to defecate and result in constipation and/or encopresis.

7.9.4 Pelvic Floor Disorders

The pelvic organs lie on a muscular floor or diaphragm made of four main muscles: the pubococcygeus, the iliococcygeus, the puborectalis (all three forming the levator ani muscle), and the coccygeus which attach to the pubis in the front, the sacrum at the back, and the inner surface of the pelvic skeleton forming a thin hammock to support them. The pelvic floor may be altered by childbirth, defecation, age, menopause, or certain surgical procedures. Pelvic floor disorders are clinically characterized either by problems with rectal evacuation (constipation), fecal incontinence, or pain or abnormal sensations in the lesser pelvis.

(a) Rectocele A rectocele is a herniation of the front wall of the rectum into the back wall of the vagina (see

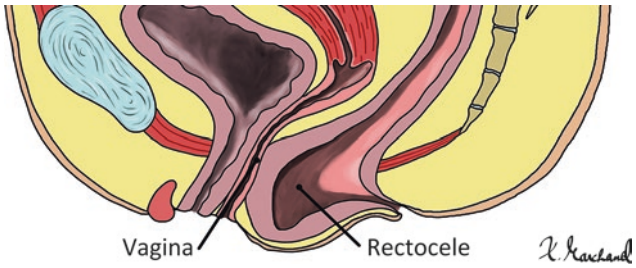


Fig. 7.23 Rectocele is a saccular deformation and bulging of the rectum toward the vaginal cavity and where stools can be trapped and difficult to evacuate

Fig. 7.23). It can be promoted by various factors including age, pregnancy, hysterectomy, or chronic constipation. The rectocele is manifested by dyschesia (difficulty in passing stools); perception of need to defecate is not affected, and the frequency of bowel movements remains normal. Signs associated with dyschezia are as follows:

- Excessive straining to evacuate stools
- Need to use digital pressure in the vagina or to apply pressure on the pubic (or perineal) area to facilitate emptying of the rectocele and stool evacuation from the rectum
- Incomplete stool evacuation after defecation, forcing the patient to return several times to the bathroom to finish emptying, or with staining of undergarments after defecation due to persistent stools in the rectocele

Diagnosis of rectocele It is based on the symptomatology and perineal examination (in supine or gynecological position, during a rectal examination with the finger forward, when the patient is asked to push, the finger gets into the rectocele and may protrude into the vagina).

Investigation by defecography is useful when surgical treatment is considered. This X-ray examination films the defecation of a barium paste introduced into the rectum. A rectocele will be considered significant when its depth during straining exceeds 3 cm, and if, after defecation, there is still contrast material left in the rectocele.

Treatment of rectocele It should be done in a stepwise fashion and according to symptoms. Medical treatment begins with a precise explanation of the anatomical abnormalities to the patient. Manual maneuvers, if they are not disturbing, can be continued by the patient and the best anatomical knowledge will allow them to be maximized. Rectal emptying of hard stools maybe difficult and osmotic laxatives in combination with fiber are often helpful; rectal suppositories, e.g., glycerin-based suppositories, can be used to promote complete evacuation.

Most rectocele surgeries are performed by gynecologic surgeons and aim to strengthen the posterior vaginal wall.

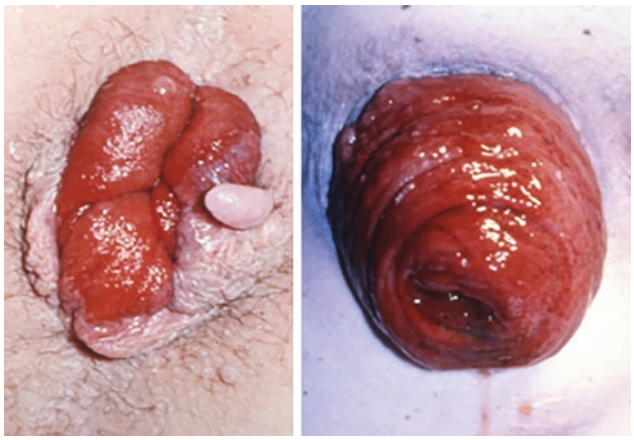


Fig. 7.24 Rectal prolapse can be partial, i.e., made of mucosal and submucosal layers only, giving rise to radiating folds (left image); it can also be complete, i.e., include all rectal wall layers, including the muscularis, and be recognizable by its circular folds (right image). (Photos from D. Bernard)

(b) Rectal prolapse Rectal prolapse is the exteriorization of the rectum through the anal opening.

Rectal prolapse is ten times more frequent in women than in men, with a peak prevalence between the ages of 60 and 70 years; it seems to be associated with chronic constipation with straining during defecation, previous obstetric damage (which may have occurred decades prior), previous perineal surgery, weakness of pelvic floor muscles, etc.

In children, a recurrent prolapse should be investigated for three conditions: chronic constipation, celiac disease, and cystic fibrosis.

Clinical diagnosis of rectal prolapse The patient usually consults for a lump or swelling in the anus (Fig. 7.24). This swelling is initially often intermittent, occurring at the time of defecation. Initially, it spontaneously reduces at the end of defecation but may over time need to be reduced manually by the patient. Prolapse can also become permanent or occur under various circumstances such as stress or walking.

The patient may also consult for complications related to this prolapse such as oozing, discharge, incontinence, obstructive constipation, or "false" diarrhea. Rectal prolapse is not accompanied by pain except in the case of strangulation with an irreducible prolapse, a complication leading to prolapse ischemia and requiring urgent surgical treatment.

The diagnosis of a prolapse is obvious when it is visible. It is sometimes difficult for the physician to identify it when it is intermittent, but it may be revealed during a defecation thrust from the patient.

Treatment of rectal prolapse Conservative medical treatment can be used if the prolapse is not very disabling,

easily reducible, and without significant medical or quality of life consequences. In the case of external prolapse with strangulation, manual reduction should be attempted as soon as possible. The best technique is to apply powdered sugar on the prolapse in order to reduce the edema by an osmotic effect; then, a manual reduction with gradual gentle pressure to retract the prolapse usually avoids emergency surgery.

Surgical treatment is the only curative treatment for an externalized or disabling rectal prolapse.

(c) Enterocele The enterocele is a hernia of the small bowel in the pouch of Douglas. It is characterized by a sensation of pelvic heaviness, pain in sitting position, urinary symptoms, or constipation. The main risk factor is hysterectomy.

The diagnosis can sometimes be made clinically by a bi-digital examination with a rectal and vaginal examination; during bearing down, an intestinal loop can be felt between the two fingers. Defecography most often confirms the diagnosis by showing small intestinal loops descending into the lesser pelvis.

(d) Excessive perineal descent (or descending perineum syndrome) will present clinically either by constipation or by incontinence. Its diagnosis is difficult on clinical examination and is essentially made by defecography (on lateral views) showing the anorectal angle in an abnormally low position. Excessive perineal descent is rarely isolated and is most often accompanied by other disorders of the pelvic floor, including pudendal neuropathy due to repeated tension on the nerve as the perineum stretches downward.

7.9.5 Fecaloma

A fecaloma is a hardened bulk of feces accumulated in the rectum (sometimes higher up in the sigmoid). It is due to excessive stagnation of stools in the rectal ampulla and is therefore usually related to prolonged constipation and to defecation needs not felt or relieved by the patient. Adult fecal impaction is most often found in an individual with a central or peripheral neurological condition that alters rectal perception and defecation. In children, it can be found in functional constipation with or without encopresis (incontinence or fecal leakage in underwear).

Fecal impaction can induce:

- Constipation.
- Intestinal obstruction (bloating, vomiting, cramps, etc.).
- Incontinence or paradoxical overflow diarrhea (fecaloma is always to be considered in a diarrheic old man or in an encopresis child).
- Pelvic or perineal pain in the form of pressure, heaviness.

Digital rectal examination reveals a rectal ampulla full of hardened stools.

Treatment of fecaloma consists of emptying the rectum, preferably with laxatives administered rectally (enema, etc.) or combined with oral laxatives (if no obstruction). Digital disimpaction is certainly effective but often painful for the patient (and "uncomfortable" for the doctor).

PS: for complementary readings on the anorectum, see ► Chaps. 18, 19, and 20.