1 Anatomy and Embryology of the Colon, Rectum, and Anus

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Key Concepts

- The dentate line represents a true division between embryonic endoderm and ectoderm.
- The location of the anterior peritoneal reflection is highly variable and can be significantly altered by disease such as rectal prolapse.
- The right and left ischioanal space communicate posteriorly through the deep postanal space between the levator ani muscle and anococcygeal ligament.
- The junction between the midgut (superior mesenteric artery) and the hindgut (inferior mesenteric artery) leads to a potential watershed area in the area of the splenic flexure.
- There is a normal, three-stage process by which the intestinal tract rotates during development beginning with herniation of the midgut followed by return of the midgut to the abdominal cavity and ending with its fixation.

Anatomy of the Anal Canal and Pelvic Floor

Textbooks of anatomy would define the "anatomic" anal canal as beginning at the dentate (pectinate) line and extending to the anal verge. This definition is one defined truly by the embryology and mucosal histology. However, the "surgical" anal canal, as first defined by Milligan and Morgan [1], extends from the anorectal ring to the anal verge. The surgical definition of the anal canal takes into account the surrounding musculature that is critical to consider during the conduct of operations from low anterior resection to anal fistulotomy. The surgical anal canal is formed by the internal anal sphincter, external anal sphincter, and puborectalis (Fig. 1.1) and is easily identified on digital examination, ultrasound imaging [2], and magnetic resonance imaging (MRI) [3].

On average, the surgical anal canal is estimated to be longer in males than in females. Intraoperative measurements of the posterior anal canal have estimated the surgical anal canal to be 4.4 cm in men compared with 4.0 cm in women [4]. In addition, the anal canal was shown to be a unique muscular unit in that its length did not change with age. However, when using MRI, the anatomy of the anal canal has been characterized differently. MR imaging did not show a difference in the length of the posterior anal canal in men and women but did show that the anterior and posterior external anal sphincter length (not including puborectalis) was significantly shorter in women [5].

The anal canal forms proximally where the rectum passes through the pelvic hiatus and joins with the puborectalis muscle. Starting at this location, the muscular anal canal can be thought of as a "tube within a tube." The inner tube is the visceral smooth muscle of the internal anal sphincter and longitudinal layer that is innervated by the autonomic nervous system. The outer muscular tube consists of somatic muscles including the components of the puborectalis and external anal sphincter [6]. It is the outer muscular tube that provides conscious control over continence and is strengthened during Kegel exercises. The external anal sphincter extends distal to the internal anal sphincter, and the anal canal terminates at the anal verge where the superficial and subcutaneous portions of the external anal sphincter join the dermis.

Anal Canal Epithelium

The proximal anal canal has a pink appearance and is lined by the columnar epithelium of the rectal mucosa. Six to 12 mm proximal to the dentate line, the anal transition zone (ATZ) begins. The ATZ appears purple in color and represents an area of gradual transition of columnar epithelium to squamous epithelium. In a parallel to cervical anatomy, the

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S. R. Steele et al. (eds.), *The ASCRS Textbook of Colon and Rectal Surgery*, https://doi.org/10.1007/978-3-030-66049-9_1

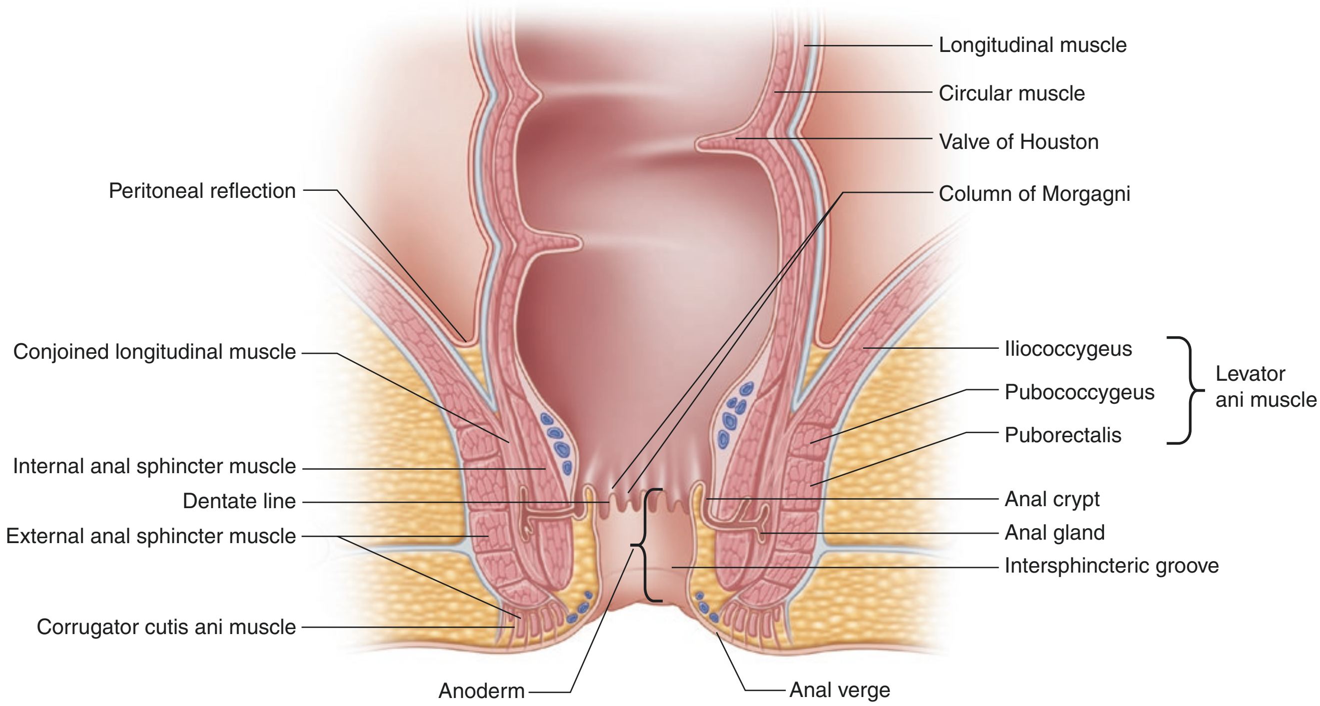


Fig. 1.1 Anal canal

ATZ is the area in which the majority of human papillomavirus-related dysplastic lesions are found in the anal canal [7]. The columns of Morgagni are noted in this area where redundant columns of tissue are noted with anal crypts at their base. This forms the rippled dentate line (or pectinate line) which can be most easily identified by locating the anal crypts at the base of the anal columns (columns of Morgagni). Anal crypts are connected to underlying anal glands which are the presumed source of sepsis in the majority of anorectal abscesses and fistula. On average, there are six anal glands surrounding the anal canal (range, 3–12) [6, 8, 9], and they tend to be more concentrated in the posterior quadrants. More than one gland may open into the same crypt, and some crypts may not be connected to anal glands. The anal gland ducts proceed inferior and lateral from the anal canal and enter the submucosa where two-thirds enter the internal anal sphincter and half terminate in the intersphincteric plane [8]. It is theorized that obstruction of these ducts leads to anal abscess and fistula [6]. Knowledge of the anatomy also explains why the internal opening of a "cryptoglandular" anal fistula should typically be at the dentate line.

Distal to the dentate line, the anoderm begins and extends for approximately 1.5 cm. According to Milligan and Morgan, anoderm or "anal canal skin… has the structure of skin, but there are no hairs and sweat glands and it consists of modified squamous transitional epithelium" [10]. In other words, anoderm has squamous histology and is devoid of hair, sebaceous glands, and sweat glands. At the anal verge, the anal canal lining becomes thickened and pigmented and contains hair follicles – this represents normal skin.

The dentate line represents a true division between embryonic endoderm and ectoderm. Proximal to the dentate line, the innervation is via the sympathetic and parasympathetic systems, with venous, arterial, and lymphatic drainage associated with the hypogastric vessels. Distal to the dentate line, the innervation is via somatic nerves with blood supply and drainage from the inferior hemorrhoidal system.

Internal Anal Sphincter

The internal anal sphincter (IAS) is the downward continuation of the circular smooth muscle of the rectum and terminates with a rounded edge approximately 1 cm proximal to the distal aspect of the external anal sphincter. 3D imaging studies of this muscle demonstrate the overall volume does not vary according to gender, but the distribution is different with women tending to have a thicker medial/distal internal anal sphincter [11]. Overall, the IAS was found to be approximately 2 mm in thickness and 35 mm in length. The authors note that on any study, it is difficult to identify the proximal portion of the IAS as it is a continuation of the wall of the lower rectum.

Conjoined Longitudinal Muscle

The anatomy and function of the perianal connective tissue is often overlooked but plays a significant role in normal anorectal function. Measuring approximately 0.5 mm to 2.0 mm in thickness, the conjoined longitudinal muscle (or conjoined longitudinal coat) lies in between the internal and external anal sphincters. It begins at the anorectal ring as an extension of the longitudinal rectal muscle fibers and descends caudally joined by fibers of the puborectalis muscle [12]. At its most caudal aspect, some of the conjoined longitudinal muscle fibers (referred to as *corrugator cutis ani muscle*) traverse the distal external anal sphincter and insert into the perianal skin, and some enter the fat of the ischiorectal fossa. Fibers of the conjoined longitudinal muscle also pass obliquely and caudally through the internal anal sphincter to interlace in a network within the subepithelial space. These subepithelial smooth muscle fibers were originally described by Treitz in 1853 [13] and have been referred to as Treitz's muscle. They have also been referred to as *corrugator cutis ani*, *musculus submucosae ani*, *mucosal suspensory ligament*, and *musculus canalis ani* [14]. It has been hypothesized by Thomson that disruption of Treitz's muscles results in anal cushion prolapse, vascular outflow obstruction, and hemorrhoidal bleeding and thrombosis [15]. Haas and Fox have hypothesized that the conjoined longitudinal muscle, and the network of connective tissue that it supports, plays a role in minimizing anal incontinence after sphincterotomy [12].

External Anal Sphincter

The external anal sphincter (EAS) is composed of striated muscle that forms an elliptical tube around the internal anal sphincter and conjoined longitudinal muscle. As it extends beyond the distal most aspect of the internal anal sphincter, the intersphincteric groove is formed. At its distal most aspect, *corrugator cutis ani muscle* fibers from the conjoined longitudinal muscle traverse the external anal sphincter and insert into the perianal skin. Milligan and Morgan described the external anal sphincter as having three distinct divisions from proximal to distal that were termed sphincter ani externus profundus, superficialis, and subcutaneous [1]. With time, this theory of three distinct divisions was proven invalid by Goligher who demonstrated that the external anal sphincter was truly a continuous sheet of skeletal muscle extending up to the puborectalis and levator ani muscles [16]. While the external anal sphincter does not have three distinct anatomic layers, it is common to see the proximal portion of the EAS referred to as deep EAS, the midportion as the superficial EAS, and the most distal aspect as the subcutaneous EAS. The mid EAS has posterior attachment to the coccyx via the anococcygeal ligament, and the proximal EAS becomes continuous with the puborectalis muscle. Anteriorly, the proximal EAS forms a portion of the perineal body with the transverse perineal muscle. There are clear differences in the morphology of the anterior external anal sphincter that have been demonstrated on both MRI and three-dimensional endoanal ultrasound studies in normal male and female volunteers [17, 18]. The normal female external anal sphincter has a variable natural defect occurring along its proximal anterior length below the level of the puborectalis sling that was demonstrated in 75 percent of nulliparous volunteers. This defect correlated with findings on anal manometry, and the authors noted that it can make interpretation of an isolated endoanal ultrasound difficult resulting in overreporting of obstetric sphincter defects [17]. This natural defect of the anterior anal sphincter provides some justification as to why anterior anal sphincterotomy is not routinely recommended in women.

The external anal sphincter is innervated on each side by the inferior rectal branch of the pudendal nerve (S2 and S3) and by the perineal branch of S4. There is substantial overlap in the pudendal innervation of the external anal sphincter muscle on the two sides which enables reinnervation to be partially accomplished from the contralateral side following nerve injury [19].

Hemorrhoids

Hemorrhoids are a normal feature of human anatomy and have been identified as present in the embryonic stage of development [20]. While many perceive hemorrhoids as a pathologic phenomenon, they are present in all humans and function to improve anal continence. The pathogenesis and treatment of hemorrhoids will be discussed elsewhere in this book, but here we will review the features of non-pathologic hemorrhoids.

Hemorrhoids are blood-filled cushions that line the anal canal. Hemorrhoids are located above and below the dentate line and have three important components: (1) the lining (mucosa or anoderm), (2) the stroma (blood vessels surrounded by connective tissue), and (3) anchoring connective tissue that secures the hemorrhoid to the internal sphincter and conjoined longitudinal muscle [20]. Hemorrhoids receive their blood supply from terminal branches of the superior hemorrhoidal artery [21]. While it has been previously stated that the terminal branches of the superior hemorrhoidal artery end in the right anterior, right posterior, and left lateral positions of the anal canal [20], this has been disputed [21]. At the level of the hemorrhoidal cushion, arteriovenous anastomosis (A-V shunts) exists in a complex vascular network termed the "corpus cavernosum recti" by Steltzner [22]. This vascular network with an arterial blood supply is why pulsatile bleeding can be seen at the time of hemorrhoidectomy.

Perineal Body

The perineal body represents the intersection of the external anal sphincter, superficial transverse perinei, deep transverse perinei, and bulbospongiosus (also referred to as bulbocavernosus) muscles (Fig. 1.2). Recent research, based on advanced magnetic resonance imaging and ultrasound, has suggested that the transverse perinei (TP) and bulbospongiosus (BS) muscles contribute significantly to anal incontinence [23]. It has been proposed that the EAS, TP, and BS muscles be collectively referred to as the "EAS complex muscles." In this theory, the EAS complex morphology is "purse string" shaped rather than the typical "donut" shape previously considered. When these muscles are considered as a functional unit, it lends further support to the idea that it is critical to attempt to repair the perineal body during overlapping sphincter reconstructions.

Pelvic Floor Muscles

In addition to the anal sphincter and perineal body, the levator ani (LA) muscles contribute to pelvic organ support. For example, injury to the LA is seen in 55% of women with pelvic organ prolapse but in only 16% without prolapse [24]. The LA has three subdivisions including the pubococcygeus (aka pubovisceral), puborectalis, and iliococcygeus. Some authors had previously suggested that the puborectalis was part of the deep portion of the EAS [25] or that the LA did not actually have three definable divisions [26]; however, a significant amount of evidence has been presented to the contrary. In vivo MRI measurements in women have shown distinct, visible muscle fascicle directions for each of the three LA component muscles [27]. Embryology studies have also demonstrated that the puborectalis muscle is a portion of the LA muscle and shares a common primordium with the iliococcygeus and pubococcygeus muscles [28].

Innervation of the levator ani muscles has been described in detailed cadaveric studies [29]. The contemporary cadaveric studies suggest that the LA muscles are innervated by the pudendal nerve branches: perineal nerve and inferior rectal nerve as well as direct sacral nerves S3 and/or S4 (aka levator ani nerve) [30]. The pubococcygeus muscle and puborectalis muscle are primarily innervated by the pudendal nerve branches, while the iliococcygeus muscle is primarily innervated by the direct sacral nerves S3 and/or S4 (Fig. 1.3).

Puborectalis Muscle

The puborectalis muscle (PRM) fibers arise from the lower part of the symphysis pubis and from the superior fascia of the urogenital diaphragm and run alongside the anorectal junction. Posterior to the rectum, the fibers join forming a sling. The "anorectal ring" is composed of the upper borders of the internal anal sphincter and puborectalis muscle [1]. Contraction of the PRM sling causes a horizontal force [27] that closes the pelvic diaphragm and decreases the anorectal angle during squeeze. This is widely considered the most important contributing factor to gross fecal continence.

Iliococcygeus Muscle

Iliococcygeus muscle (ICM) fibers arise from the ischial spines and posterior obturator fascia, pass inferior/posterior and medially, and insert into the distal sacrum, coccyx, and anococcygeal raphe. The ICM, along with the pubococcygeus muscle, contributes to "lifting" of the pelvic floor [27].

Pubococcygeus Muscle

The pubococcygeus (PCM) muscle lies medial to the PRM. PCM fibers arise from the anterior half of the obturator fascia and the high posterior pubis. The PCM fibers are directed posterior/inferior and medially, where they intersect with fibers from the opposite side and form the anococcygeal raphe (or anococcygeal ligament). PCM muscle fibers insert in the distal sacrum and tip of the coccyx. Portions of the PCM contribute to the conjoined longitudinal muscle. The PCM forms the "levator hiatus" as it ellipses the lower rectum, urethra, and either the vagina in women or the dorsal vein of the penis in men. The levator hiatus is connected to the intrahiatal organs by a fascial condensation called the "hiatal ligament" (Fig. 1.4). The hiatal ligament arises circumferentially around the hiatal margin as a continuation of the fascia on the pelvic surface of the levator muscle [31]. Enlargement of the levator hiatus has been implicated as a cause of female pelvic organ prolapse [32]. The PCM is the portion of the levator ani that is typically injured during traumatic vaginal delivery [33].

Anatomy of the Rectum

The rectum is arbitrarily considered to have three distinct parts: the upper, middle, and lower rectum. Although not anatomically distinct, the upper, mid, and lower rectal divisions are important when considering surgical treatment of rectal cancer. From the anal verge, the lower rectum is 0–7 cm; middle rectum, 7–12 cm; and upper rectum, 12–15 cm [34]. However, the rectum is actually variable in length and may

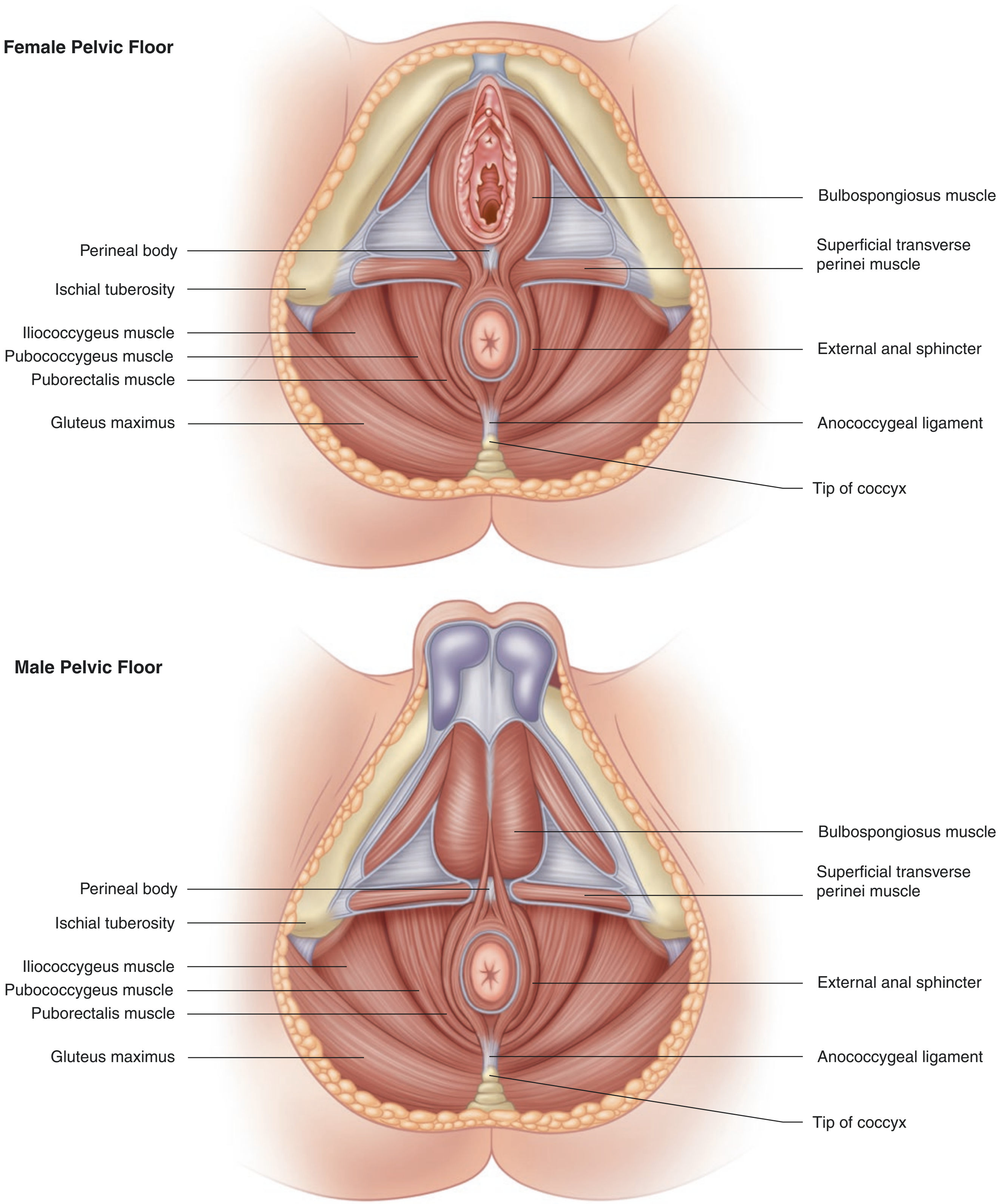


Fig. 1.2 Pelvic floor muscles

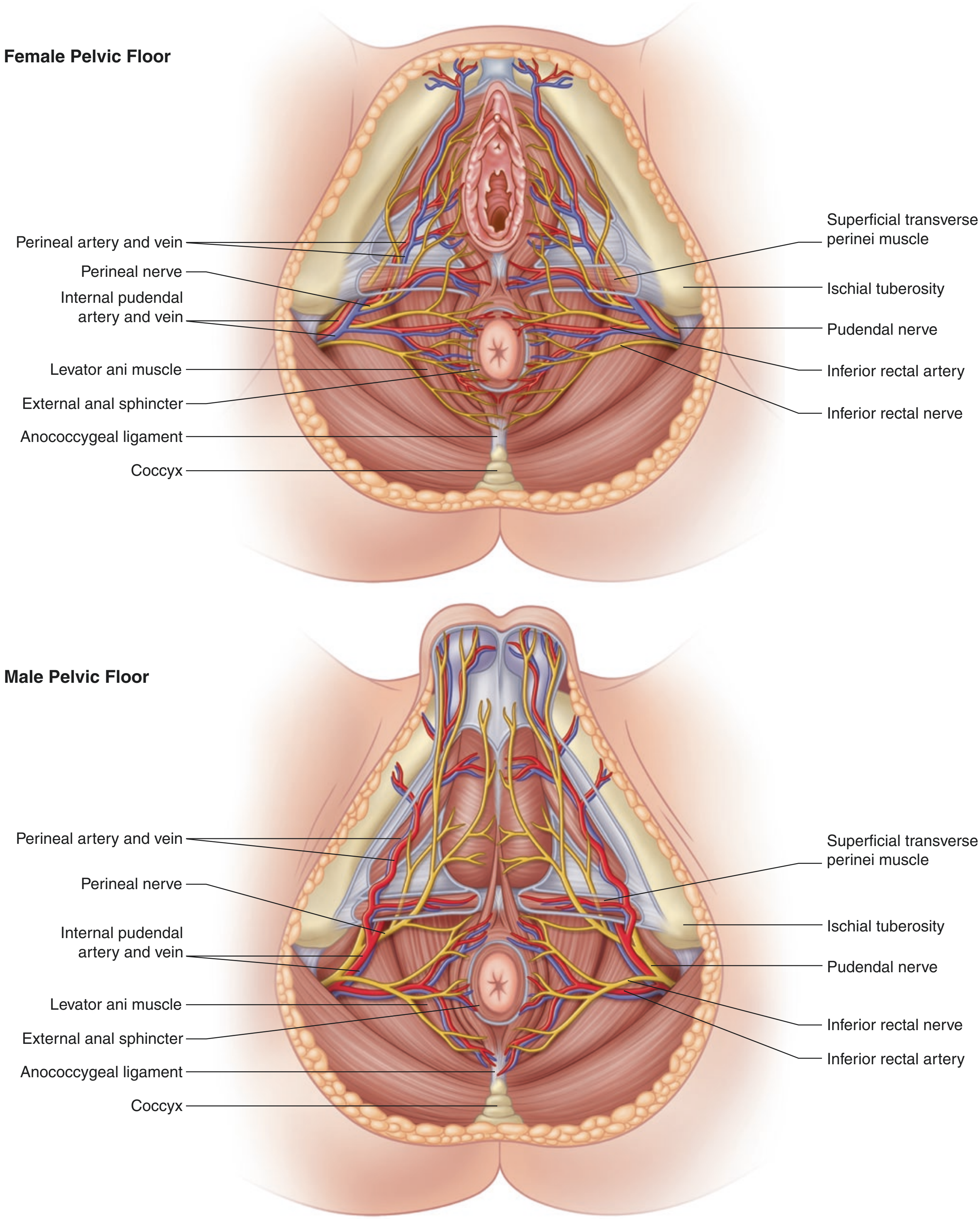


Fig. 1.3 Pelvic floor nerves and blood supply

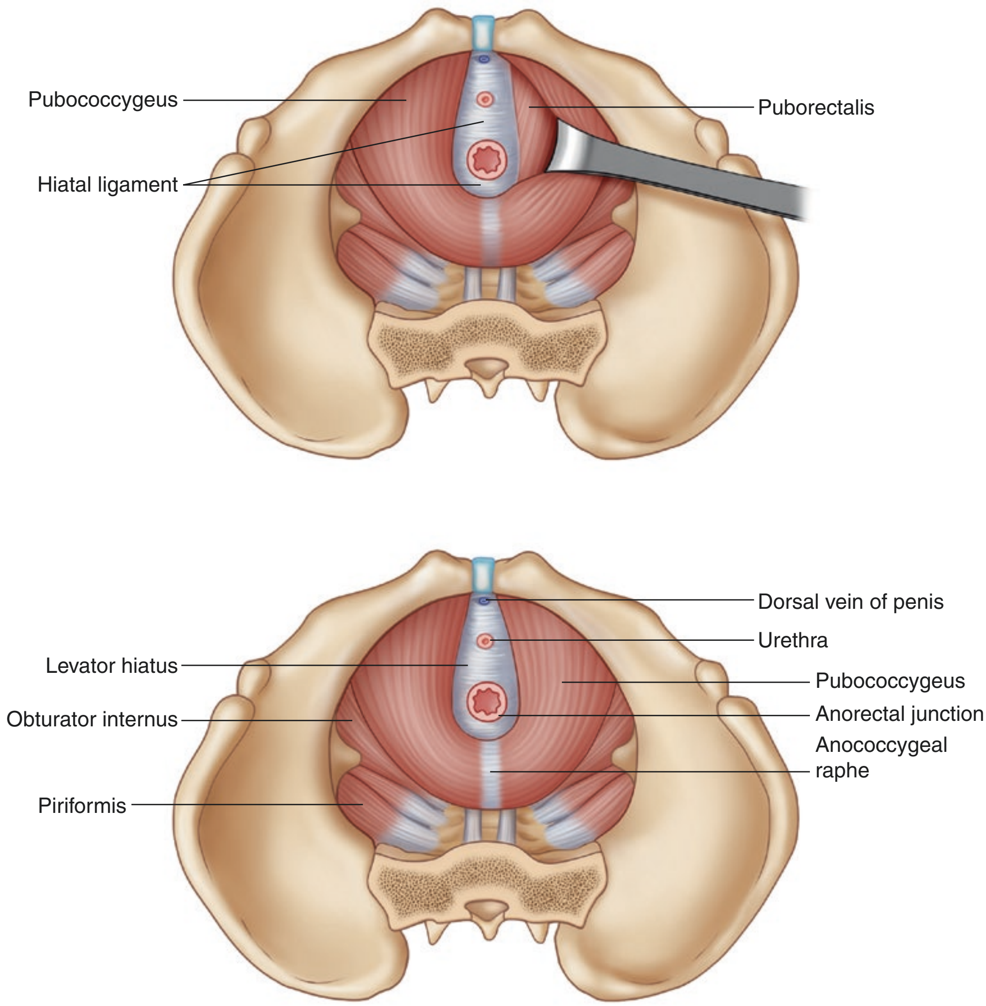


Fig. 1.4 Pelvic floor anatomy, abdominal view

extend beyond 15 cm from the anal verge. The upper rectum can be distinguished from the sigmoid colon by the absence of taenia coli and epiploic appendages.

The majority of the rectum lies outside of the peritoneal cavity, although anteriorly and laterally the upper rectum is covered by a layer of visceral peritoneum down to the peritoneal reflection. The location of the anterior peritoneal reflection is highly variable and can be significantly altered by disease such as rectal prolapse. Given the importance of the location of the peritoneal reflection with respect to transanal excision of rectal tumors, one study sought to identify the location of the anterior peritoneal reflection in 50 patients who were undergoing laparotomy [35]. It was found that the anterior peritoneal reflection was located on average 9 cm from the anal verge in females and 9.7 cm from the anal verge in males – there was no statistically significant difference based on gender.

Mesorectum

The origin of the word "mesorectum" is difficult to identify and may be attributed to Maunsell in 1892 [36] but was certainly later popularized by Heald [37]. Unfortunately, the term mesorectum is a misnomer that is not generally acknowledged in classic texts of anatomy such as the *Nomina Anatomica* [38]. In anatomic terms, the prefix "meso" refers to two layers of peritoneum that suspend an organ, and the suffix applied indicates the target organ (e.g., mesocolon). The term "meso" cannot be assigned to the rectum, as it implies a mobile, suspended rectum, which may only be the case in patients with rectal prolapse.

The mesorectum is a term employed by surgeons to describe the fascial envelope of the rectum that is excised during surgical treatment of rectal cancer. Indeed, failure to completely excise this envelope intact has been associated with an increased

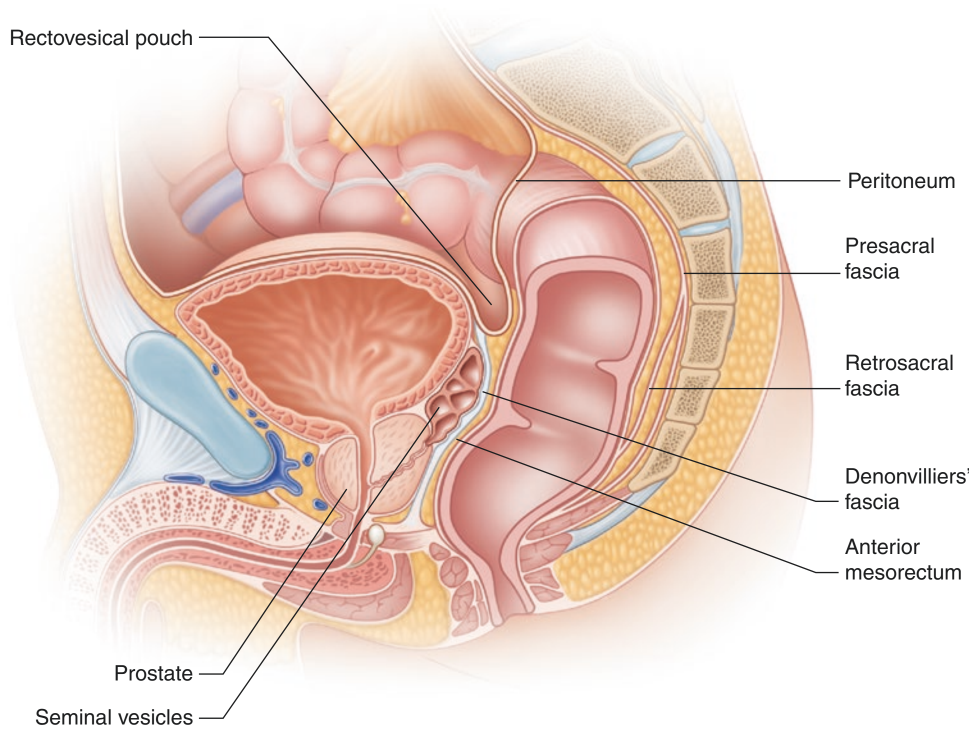


Fig. 1.5 Fascial relationships of the rectum

incidence of local recurrence of rectal cancer [39]. The mesorectum is contained within the fascia propria. The fascia propria is an upward projection of the parietal endopelvic fascia that lines the walls and floor of the pelvis. The fascia propria encloses the perirectal fat, lymphatics, blood vessels, and nerves and is not considered a barrier strong enough to prevent the spread of infection or malignancy [40].

Presacral Fascia

The presacral fascia is a thickened portion of the parietal endopelvic fascia overlying the sacrum that covers the presacral veins and hypogastric nerves (Fig. 1.5). It extends laterally to cover the piriformis and upper coccyx. As the presacral fascia extends laterally, it becomes continuous with the fascia propria and contributes to the lateral ligaments of the rectum. Caudally, this fascia extends to the anorectal junction covering the anococcygeal ligament. During total mesorectal excision, the fascia propria is elevated sharply off the presacral fascia. Leaving the presacral fascia intact eliminates the possibility of causing presacral bleeding.

Retrosacral Fascia

The retrosacral fascia originates at the third and fourth portion [41] of the sacrum and extends anteriorly to the posterior layer of the fascia propria 3–5 cm proximal to the anorectal junction [42]. This tough fascia layer is surgically relevant as it must be sharply incised during total mesorectal excision [40]. The space posterior to the retrosacral fascia is referred to as the supralevator or retrorectal space.

Waldeyer's Fascia

There is significant confusion about what Waldeyer's fascia represents as the eponym has been used to describe the presacral fascia, the retrosacral fascia, or all fascia posterior to the rectum. In Waldeyer's original description of pelvic fascia, there was no particular emphasis on the presacral component [40, 42]. While the debate continues regarding "Waldeyer's fascia," it is important to simply understand that the phrase can have the potential to mean presacral fascia, retrorectal fascia, or both [43].

Denonvilliers' Fascia

Denonvilliers' fascia arises from the fusion of the two walls of the embryological peritoneal cul-de-sac and extends from the deepest point of the rectovesical pouch to the pelvic floor [44]. Originally described by Denonvilliers in 1836 as a "prostatoperitoneal" membranous layer between the rectum and seminal vesicles, Denonvilliers fascia is also present in females as part of the rectovaginal septum and is sometimes referred to as rectovaginal fascia. It is found immediately beneath the vaginal mucosa and is clearly what most would consider as part of the vaginal wall. It merges superiorly with the cardinal/uterosacral complex in females or the rectovesical pouch in males. It merges laterally with the endopelvic fascia overlying the levator muscle and distally with the perineal body. It contains collagen, some strands of smooth muscle, and heavy elastin fibers. Rectoceles represent a defect in this layer that allows the rectum to bulge anteriorly [45].

Microscopically, the Denonvilliers' fascia has two layers; however, it is not possible to discern two layers during pelvic dissection [44]. In the anterior rectal plane, the mesorectum is contained by the fascia propria which lies dorsal to Denonvilliers' fascia. The cavernous nerves run in neurovascular bundles at the anterolateral border of Denonvilliers' fascia.

Lateral Ligaments

While frequently referred to by surgeons, there are two controversial points regarding the lateral ligaments of the rectum. First, do the lateral ligaments exist? Second, what do they contain? Miles refers to division of the lateral ligaments of the rectum in his seminal description of abdominoperineal resection in 1908. Specifically, he notes "In these structures the middle haemorrhoidal arteries are found but seldom require a ligature" [46]. It is interesting to note that at least one modern cadaveric dissection study identified the presence of a middle rectal artery in only 22% of specimens [41] which could be a contributing factor as to why Miles saw no significant bleeding in this area.

Total mesorectal excision, as popularized and described by Heald, involves sharp dissection along the fascia propria circumferentially to the pelvic floor. While acknowledging that the middle rectal vessels are "divided as far from the carcinoma as possible," Heald does not mention "lateral ligaments" of the rectum at all [47].

In an extensive review of the anatomy of the lateral ligament, Church notes that it is a common misconception that the lateral ligaments contain the middle rectal artery at all. It appears that the lateral ligaments comprise "primarily nerves and connective tissue" and their division without bleeding attests to the absence of a "significant accessory rectal artery in this location in the majority of patients" [40].

In a separate cadaveric study, the lateral ligaments of the rectum were identified as trapezoid structures originating from mesorectum and anchored to the endopelvic fascia at the level of the midrectum. It was recommended that, as lateral extensions of the mesorectum, the ligaments must be cut and included in the total mesorectal excision (TME) specimen. It was further noted that the lateral ligaments did not contain middle rectal arteries or nerve structures of importance. The urogenital bundle runs just above the lateral ligament at its point of insertion on the endopelvic fascia, the middle rectal artery (if present) runs posterior to the lateral ligament, and the nervi recti fibers (which originate from the inferior hypogastric plexus) course transversely under the lateral ligament to the rectal wall [48]. Other modern cadaveric investigations note the rarity of middle rectal arteries and the absence of clinically relevant neurovascular structures in the lateral ligaments [49].

Rectal Valves: The Spiral Valves of Houston and Kohlrausch's Valve

The first anatomic description of rectal valves is credited to Giovanni Morgagni [50]; however, it was John Houston, an Irish anatomist and surgeon, who presented the first seminal work on the structures [51, 52]. Houston described an average of three oblique valves with an upward orientation and concave surface that were located successively on opposite sides of the rectum that formed "a sort of spiral tract down its cavity." Houston theorized that these valves might aid in continence by supporting "the weight of fecal matter"; however, this has not been substantiated elsewhere.

Modern anatomy texts usually also describe three rectal valves (Fig. 1.1) with the superior and inferior valves located on the left side of the rectum and the more prominent middle rectal valve on the right; however, this is not uniformly the case [53]. Only 45.5% of patients will have the classic three valve rectal anatomy; 32.5% will have only two valves; and 10.25% may have four valves.

After Houston's definitive description of rectal valves in 1830, Otto Kohlrausch, a physician and scientist in Germany, described a single mid-rectal valve in 1854 [54]. When there are three valves, current anatomists identify Kohlrausch's valve as the middle one [51]. This valve is usually the largest, located on the right and approximately 9–11 cm from the anal verge, and some authors have suggested this valve could serve as an intraluminal marker for the area of the anterior peritoneal reflection [55].

Anorectal Spaces

It is important to acknowledge and understand the anorectal spaces created by the various myofascial relationships in the pelvis as these spaces help us understand how anorectal sepsis can spread throughout the pelvis.

Perianal Space

The perianal space contains external hemorrhoid cushions, the subcutaneous external anal sphincter and the distal internal anal sphincter. The perianal space is in communication

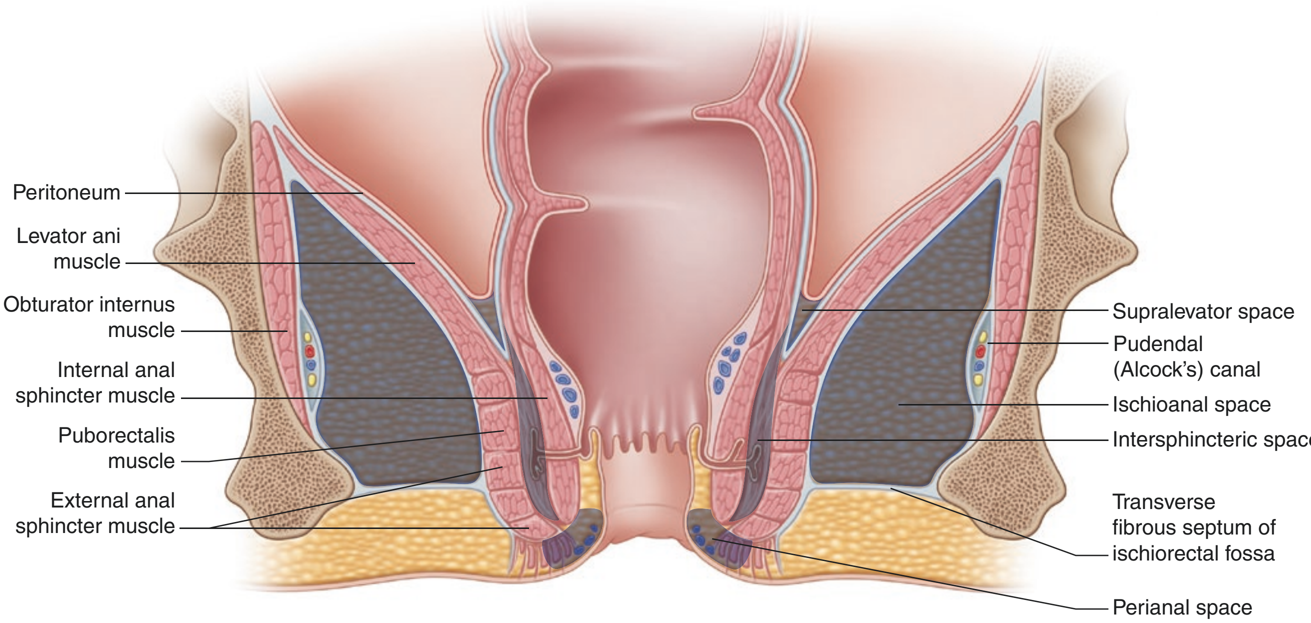


Fig. 1.6 Perianal and perirectal spaces, coronal view

with the intersphincteric space (Fig. 1.6). The perianal space has its cephalad boundary at the dentate line and laterally to the subcutaneous fat of the buttocks or is contained by fibers extending from the conjoined longitudinal muscle often referred to as *corrugator cutis ani* muscle fibers. Otherwise, the perianal space is contained by anoderm.

Intersphincteric Space

The intersphincteric space is the potential space that lies between the internal and external anal sphincter and is continuous with the perianal space. It is of clinical importance as cryptoglandular infections tend to begin in this area and expand elsewhere to create anal fistula [6].

Submucous Space

This space lies between the medial boarder of the internal anal sphincter and the anal mucosa proximal to the dentate line. It is continuous with the submucosa of the rectum. This area contains internal hemorrhoid vascular cushions.

Ischioanal/Ischiorectal Space

The ischioanal (also referred to as ischiorectal) space is the largest anorectal space. It has been described as a pyramid shape with its apex at the levator muscle insertion into the obturator fascia. The medial boarder is thus the levator ani muscle and external anal sphincter. The obturator internus muscle and obturator fascia make up the lateral boarder of the ischioanal space. The posterior boundary is formed by the lower border of the gluteus maximus muscle and the sacrotuberous ligament. The space has an anterior boundary formed by the superficial and deep transverse perineal muscles. The caudal boundary is skin of the perineum. The ischioanal fossa contains adipose tissue, pudendal nerve branches, and superficial branches of the internal pudendal vessels. The right and left ischioanal space communicate posteriorly through the deep postanal space between the levator ani muscle and anococcygeal ligament (Fig. 1.7) [56]. When the ischioanal and perianal spaces are regarded as a single space, it is referred to as the ischioanal fossa [43].

Supralevator Space

The upper boundary of the supralevator space is the peritoneum, the lateral boundary is the pelvic wall, the medial boundary is the rectum, and the inferior boarder is the levator ani muscle (Fig. 1.8).

Superficial and Deep Postanal Spaces

These spaces are located posterior to the anus and inferior to the levator muscle. The superficial postanal space is more caudal and is located between the anococcygeal ligament and the skin. The superficial postanal space allows communication of perianal space sepsis.

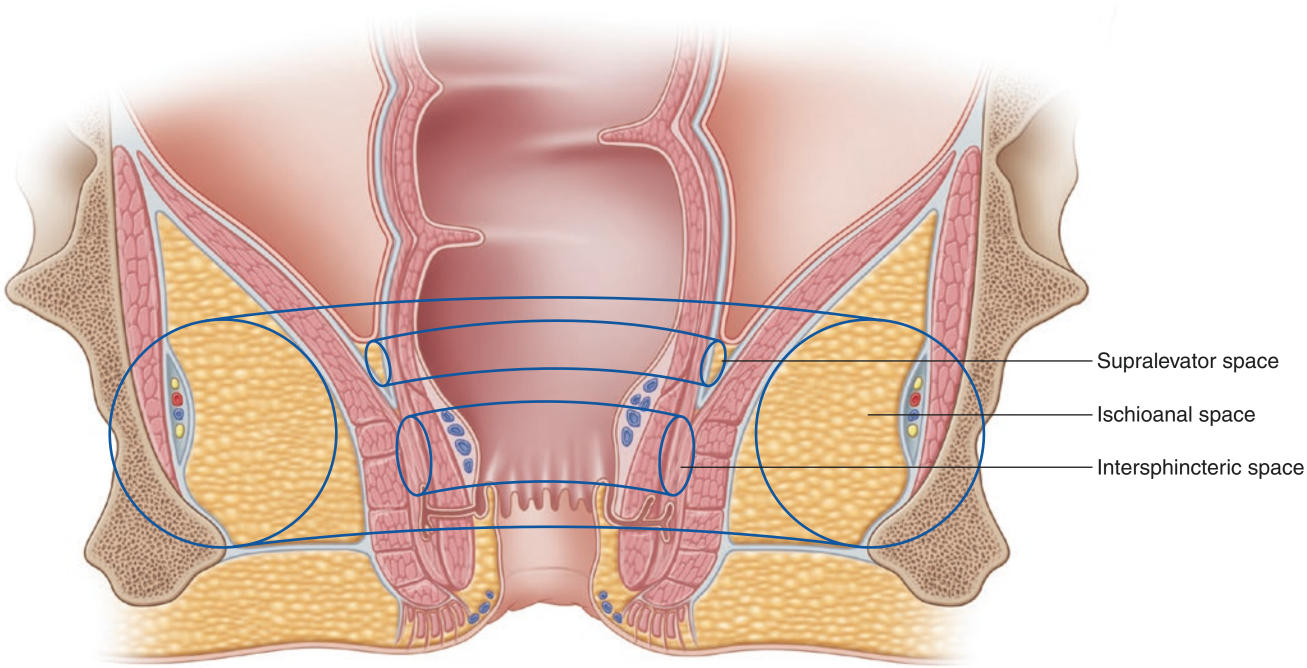
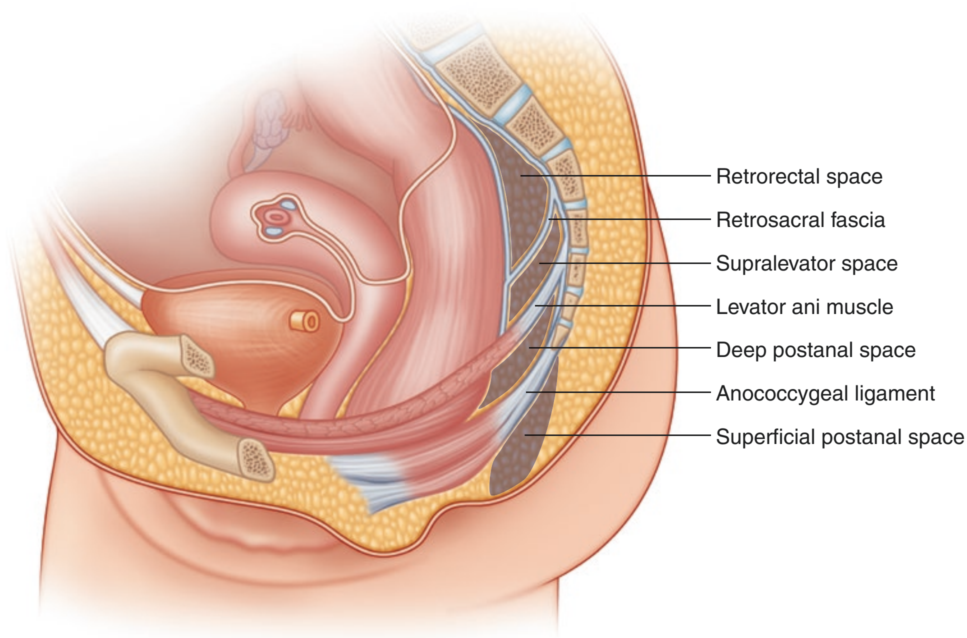


Fig. 1.7 Communication of the anorectal spaces

Fig. 1.8 Perianal and perirectal spaces, lateral view



The deep postanal space (retrosphincteric space of Courtney) [57] is located between the levator ani muscle and the anococcygeal raphe. This space allows ischioanal sepsis to track from one side to the other resulting in the so-called "horseshoe" abscess.

Retrorectal Space

The retrorectal space is found between the presacral fascia and fascia propria. It contains no major blood vessels or nerves. It is limited laterally by the lateral ligaments of the piriformis fascia and inferiorly by the retrosacral fascia. The fascia propria and presacral fascia come together at the apex of this space [40].

Rectal Blood Supply

The rectum is supplied by the superior, middle, and inferior rectal (hemorrhoidal) arteries (Fig. 1.9). Both the middle and inferior hemorrhoidal vessels are paired arteries, and the superior rectal artery is not.

Superior Rectal Artery

The superior rectal artery (SRA) is the continuation of the inferior mesenteric artery and is so named after the inferior mesenteric artery crosses the left iliac vessels. The SRA gives off a rectosigmoid branch, an upper rectal branch, and then bifurcates into right and left terminal branches in 80% [58] of cases as it descends caudally in the mesorectum. On average, eight terminal branches of the SRA have been identified in the distal rectal wall [21].

Middle Rectal Artery

The middle rectal artery (MRA) has been variably noted in many studies. It may be found on one or both sides of the rectum and has been noted to be present 12–28% of the time [49, 59]. At least one study reported the presence of the middle rectal artery in at least 91% of cadaveric specimens [48]. The MRA originates from the anterior division of the internal iliac or pudendal arteries. Please see the “Lateral Ligament” discussion above for more review on the anatomic course of the middle rectal artery.

Inferior Rectal Artery

The inferior rectal arteries (IRA) are paired vessels that originate as branches of the internal pudendal artery which receives its blood supply from the internal iliac artery. The artery originates in the pudendal canal and is entirely extrapelvic (caudal to the levator ani) in its distribution. The IRA traverses the obturator fascia and the ischiorectal fossa and pierces the wall of the anal canal in the region of the external anal sphincter [40].

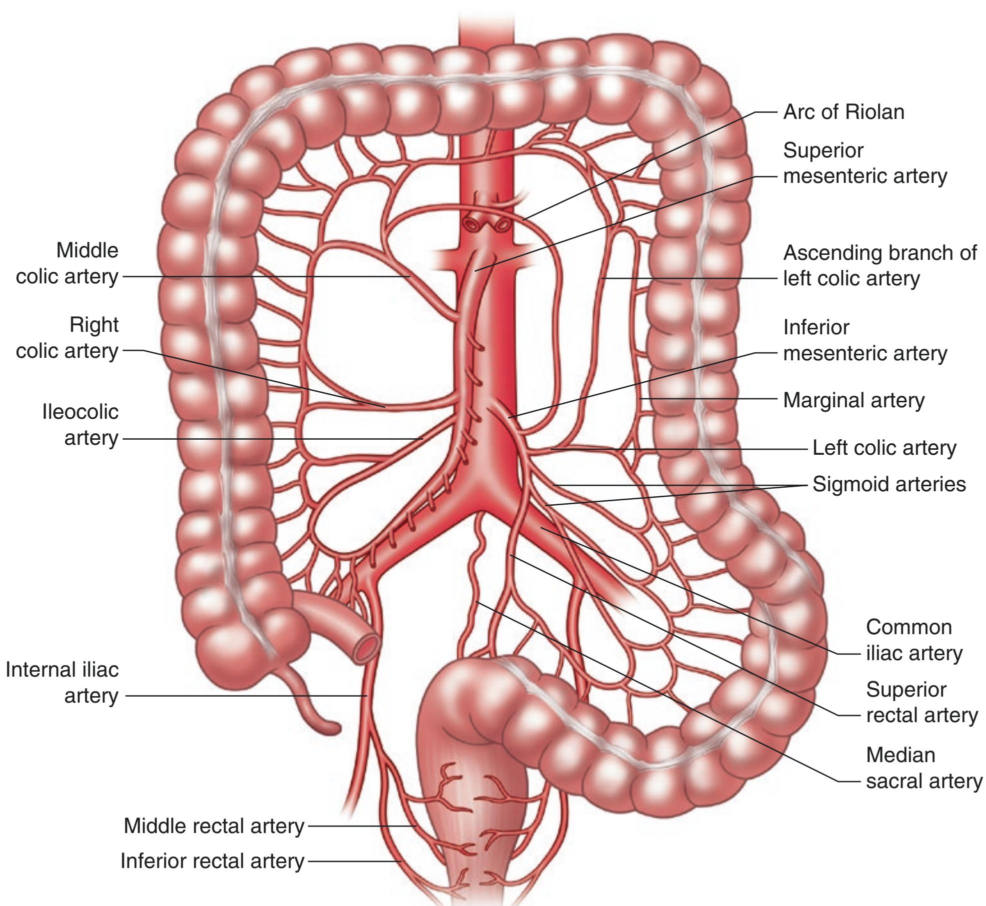


Fig. 1.9 Arterial anatomy of the colon and rectum

Venous and Lymphatic Drainage of the Rectum and Anus

Venous drainage from the rectum and anus occurs via both the portal and systemic systems. Middle and inferior rectal veins drain to the systemic systems via the internal iliac vein, while the superior rectal vein drains the rectum and upper anal canal into the portal system via the inferior mesenteric vein (Fig. 1.10). The two systems of drainage in the rectum, thus, explain the potential development of rectal varices in patients with portal hypertension.

Lymphatics from the upper two-thirds of the rectum drain to the inferior mesenteric lymph nodes and then to the para-aortic lymph nodes. Lymphatic drainage from the lower third of the rectum occurs along the superior rectal artery and laterally along the middle rectal artery to the internal iliac lymph nodes. In the anal canal, lymphatics above the dentate drain to the inferior mesenteric and internal iliac lymph nodes. Below the dentate line, lymphatics drain along the inferior rectal lymphatics to the superficial inguinal nodes.

Innervation of the Rectum and Anus

Sympathetic fibers arise from L1, L2, and L3 and pass through the sympathetic chains and join the preaortic plexus (Fig. 1.11). From there, they run adjacent and dorsal to the inferior mesenteric artery as the mesenteric plexus and innervate the upper rectum. The lower rectum is innervated by the presacral nerves from the hypogastric plexus. Two main hypogastric nerves, on either side of the rectum, carry sympathetic information from the hypogastric plexus to the pelvic plexus. The pelvic plexus lies on the lateral side of the pelvis at the level of the lower third of the rectum adjacent to the lateral stalks (please see discussion of lateral stalks above).

Parasympathetic fibers to the rectum and anal canal originate from S2, S3, and S4 to penetrate through the sacral fora-

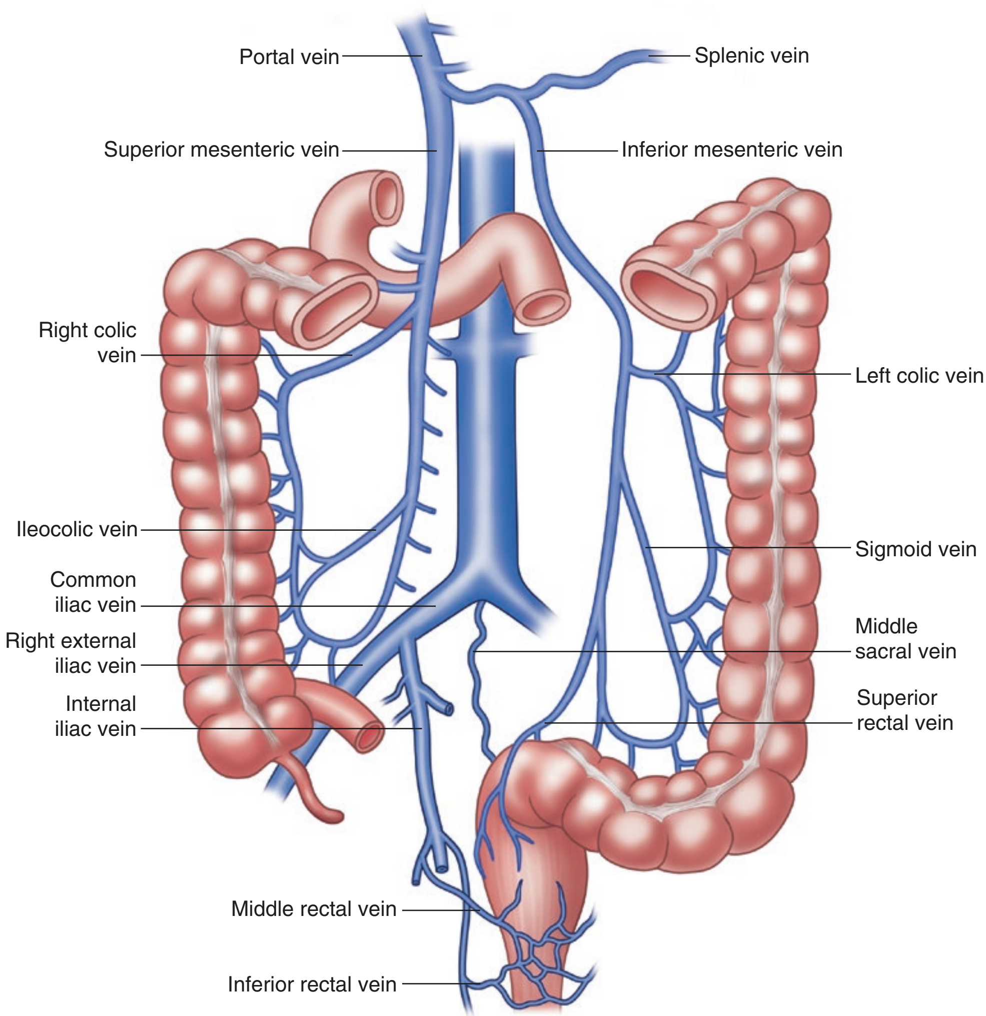
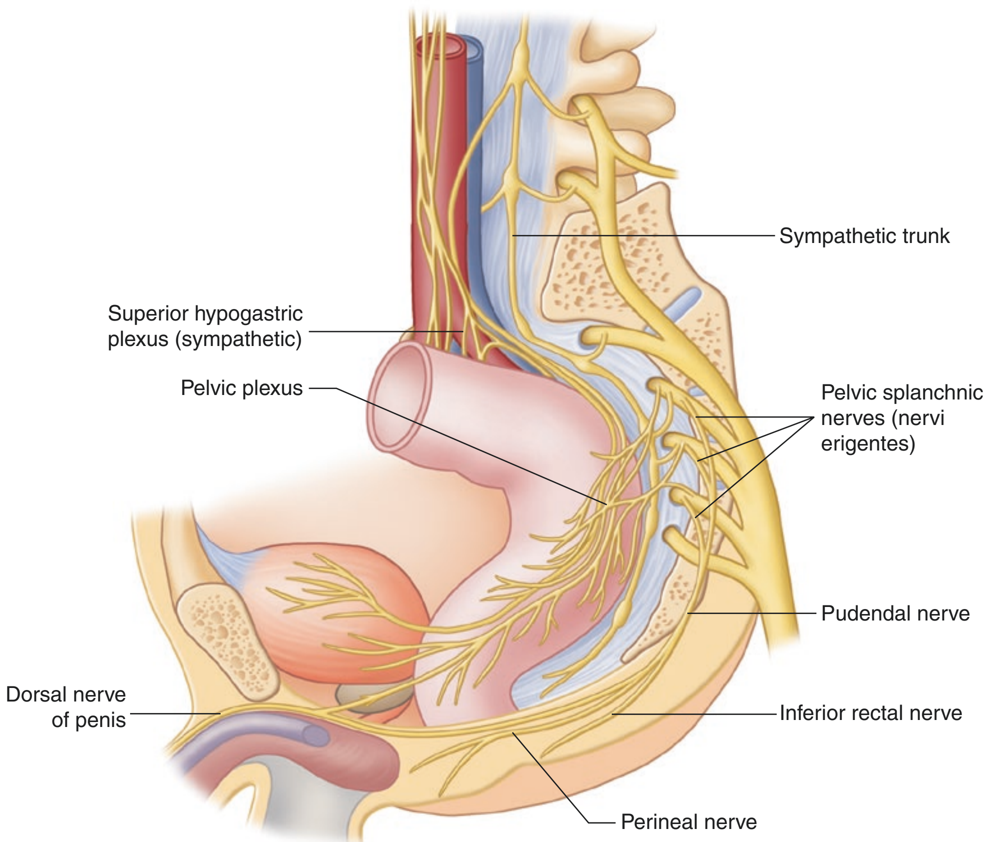


Fig. 1.10 Venous anatomy of the colon and rectum

Fig. 1.11 Nerves of the rectum



men and are called the nervi erigentes. These nerves course laterally and anteriorly to join the sympathetic hypogastric nerves and form the pelvic plexus on the pelvic sidewall. From here, postganglionic mixed parasympathetic and sympathetic nerve fibers supply the rectum, genital organs, and anal canal. The periprostatic plexus is considered a subdivision of the pelvic plexus and supplies the prostate, seminal vesicles, corpora cavernosa, vas deferens, urethra, ejaculatory ducts, and bulbourethral glands.

The internal anal sphincter is innervated by sympathetic (L5) and parasympathetic (S2, S3, and S4) nerves following the same route as the nerves to the rectum as noted above. The external anal sphincter is innervated on each side by the inferior rectal branch of the internal pudendal nerve (S2 and S3) and by the perineal branch of S4. Anal sensation is mediated by the inferior rectal branch of the pudendal nerve.

Anatomy of the Colon

The colon is a long tubular organ consisting of muscle and connective tissue with an inner mucosal layer. The diameter of the colon differs depending upon which segment is evaluated and generally decreases from proximal to distal (cecum about 7 cm and sigmoid colon about 2.5 cm in diameter). The overall length is variable with an average length approximating 150 cm. The right and left sides of the colon are fused to the posterior retroperitoneum (secondarily retroperitonealized), while the transverse colon and sigmoid colon are relatively free within the peritoneum. The transverse colon is held in position via its attachments to the right/left colon at the flexures (hepatic and splenic, respectively) and is further fused to the omentum. Generally, the colon is located peripherally within the abdomen with the small bowel located centrally.

There are three classic anatomic points of differentiation between the colon and the small intestine: the appendices epiploicae, the taeniae coli, and the haustra. The appendices epiploicae are non-mesenteric fat protruding from the serosal surface of the colon. They are likely residual from the antimesenteric fat of the embryologic intestine which dissipates (unlike the omentum on the stomach). The taenia coli are three thickened bands of outer, longitudinal muscle of the colon. This outer layer of muscle is indeed circumferentially complete [60] but is considerably thicker in three areas represented by the taenia. The three taeniae have been given separate names by some: *taenia libera* to represent the anterior band, *taenia mesocolica* for the posteromedial band, and *taenia omentalis* for posterolateral band. The bands are continuous from their origin at the base of the appendix until the rectosigmoid junction where they converge (marking an anatomically identifiable differentiation between the sigmoid colon and rectum). Though they run along the full length of the colon, they are not as long as the bowel wall. This difference in length results in outpouchings of the bowel wall between the taenia referred to as haustra. The haustra are further septated by the plicae semilunares.

Cecum

The proximal most portion of the colon is termed the cecum, a saclike segment of the colon below (proximal to) the ileocecal valve. The cecum is variable in size but generally is about 8 cm in length and 7 cm in diameter. At its base is the appendix. Terminating in the posteromedial area of the cecum is the terminal ileum (ileocecal valve). The cecum is generally covered by visceral peritoneum, with more variability near the transition to the ascending colon (upper or distal cecum). The ileocecal valve is a circular muscular sphincter which appears as a slit-like ("fish-mouth") opening noted on an endoscopic evaluation of the cecum. The valve is not competent in all patients, but when present, its competence leads to the urgency of a colon obstruction as it develops into a closed-loop obstruction. Regulation of ileal emptying into the colon appears to be the prime task in ileocecal valve function [61].

The Appendix

The appendix is an elongated, true diverticulum arising from the base of the cecum. The appendiceal orifice is generally about 3–4 cm from the ileocecal valve. The appendix itself is of variable length (2–20 cm) and is about 5 mm in diameter in the non-inflamed state. Blood is supplied to the appendix via the appendiceal vessels contained within the mesoappendix. This results in the most common location of the appendix being medially on the cecum toward the ileum, but the appendix does have great variability in its location including pelvic, retrocecal, preileal, retroileal, and subcecal. Though traditionally thought to be an unnecessary vestige, modern research points to the appendix which is rarely absent and rarely altered actually playing an important role in immune function and/or the colonic microbiome [62].

Ascending Colon

From its beginning at the ileocecal valve to its terminus at the hepatic flexure where it turns sharply medially to become the transverse colon, the ascending colon measures on average, about 15–18 cm. Its anterior surface is covered in visceral peritoneum, while its posterior surface is fused with the retroperitoneum. The lateral peritoneal reflection can be seen as a thickened line termed the white line of Toldt, which can serve as a surgeon's guide for mobilization of the ascending colon off of its attachments to the retroperitoneum, most notably the right kidney (Gerota's fascia) and the loop of the duodenum located posterior and superior to the ileocolic vessels. The right ureter and the right gonadal vessels pass posteriorly to the ascending mesocolon within the retroperitoneum.

Transverse Colon

The transverse colon traverses the upper abdomen from the hepatic flexure on the right to the splenic flexure on the left. It is generally the longest section of colon (averaging 45–50 cm) and swoops inferiorly as it crosses the abdomen. The entire transverse colon is covered by visceral peritoneum, but the greater omentum is fused to the anterosuperior surface of the transverse colon. Superior to the transverse mesocolon, inferior to the stomach, and posterior to the omentum is the pocket of the peritoneal cavity termed the lesser sac, with the pancreas forming the posterior most aspect. The splenic flexure is the sharp turn from the transversely oriented transverse colon to the longitudinally oriented descending colon. It can be adherent to the spleen and to the diaphragm via the phrenicocolic ligament.

Descending Colon

The descending colon travels inferiorly from the splenic flexure for the course of about 25 cm. It is fused to the retroperitoneum (similarly to the ascending colon) and overlies the left kidney as well as the back/retroperitoneal musculature. Its anterior and lateral surfaces are covered with visceral peritoneum, and the lateral peritoneal reflection (white line of Toldt) is again present.

Sigmoid Colon

The sigmoid colon is the most variable of the colon segments. It is generally 35–45 cm in length. It is covered by visceral peritoneum, thereby making it mobile. Its shape is considered "omega-shaped," but its configuration and attachments are variable. Its mesentery is of variable length but is fused to the pelvic walls in an inverted-V shape creating a recess termed the intersigmoid fossa. Through this recess travel the left ureter, gonadal vessels, and often the left colic vessels.

Rectosigmoid Junction

The end of the sigmoid colon and the beginning of the rectum is termed the rectosigmoid junction. It is noted by the confluence of the taeniae coli and the end of epiploicae appendices. While some surgeons have historically considered the rectosigmoid junction to be a general area (comprising about 5 cm of distal sigmoid and about 5 cm of proximal rectum), others have described a distinct and clearly defined segment. It is the narrowest portion of the large intestine, measuring 2–2.5 cm in diameter. Endoscopically, it is noted as a narrow and often sharply angulated area above the relatively capacious rectum and above the three rectal valves.

In the early nineteenth century, it was proposed that the sigmoid acts as a reservoir for stool, thus aiding in continence [63]. Subsequently, an area of thickened circular muscle within the wall of the rectosigmoid was described and felt to function as a sphincter of sorts. Historically, it has been variably named the *sphincter ani tertius*, *rectosigmoid sphincter*, and *pylorus sigmoidorectalis* [64–68]. A more recent evaluation of the rectosigmoid junction utilizing anatomic and histologic studies as well as radiographic evaluation concluded that there was an anatomic sphincter at the rectosigmoid junction [69]. Microscopic evaluation of the area does reveal thickening of the circular muscle layer as it progresses toward the rectum. Though not identifiable externally, radiologic evaluation can identify the area as a narrow, contractile segment [69].

Blood Supply

The colon receives blood supply from two main sources, branches of the superior mesenteric artery (SMA) (cecum, ascending, and transverse colon) and branches of the inferior mesenteric artery (IMA) (descending and sigmoid colon) (Fig. 1.9). There is a watershed area between these two main sources located just proximal to the splenic flexure where branches of the left branch of the middle colic artery anastomose with those of the left colic artery. This area represents the border of the embryologic midgut and hindgut. Though the blood supply to the colon is somewhat variable, there are some general common arteries. The cecum and right colon are supplied by the terminus of the SMA, the ileocolic artery. The right colic artery is less consistent and, when present, can arise directly from the SMA, from the ileocolic, or from other sources. The transverse colon is supplied via the middle colic artery, which branches early to form right and left branches. The middle colic artery originates directly from the SMA. The left colon and sigmoid colon are supplied by branches of the IMA, namely, the left colic and a variable number of sigmoid branches. After the final branches to the sigmoid colon, the IMA continues inferiorly as the superior hemorrhoidal (rectal) artery.

Superior Mesenteric Artery

The superior mesenteric artery (SMA) is the second, unpaired anterior branch off of the aorta (the first being the celiac trunk). It arises posterior to the upper edge of the pancreas (near the L1 vertebrae), courses posterior to the pancreas, and then crosses over the third portion of the duodenum to continue within the base of the mesentery. From its left side, the SMA gives rise to up to 20 small intestinal branches, while the colic branches originate from its right side. The most constant of the colic branches is the ileocolic vessel which courses through the ascending mesocolon where it divides into a superior (ascending) branch and an inferior (descending) branch [70]. A true right colic artery is absent up to 20% of the time and, when present, typically arises from the SMA. Alternatively, the right colic artery can arise from the ileocolic vessels or from the middle colic vessels [58, 70, 71]. The middle colic artery arises from the SMA near the inferior border of the pancreas. It branches early to give off right and left branches. The right branch supplies the hepatic flexure and right half of the transverse colon. The left branch supplies the left half of the transverse colon to the splenic flexure. In up to 33% of patients, the left branch of the middle colic artery can be the sole supplier of the splenic flexure [70, 72]. Recent reports describe an accessory middle colic artery (AMCA). One single-center study demonstrates that more than one-third of patients had an AMCA (36.4%) supplying the splenic flexure with about 85% originating off of the SMA and coursing along the inferior border of the pancreas toward the splenic flexure [73].

Inferior Mesenteric Artery

The inferior mesenteric artery (IMA) is the third unpaired, anterior branch off of the aorta, originating 3–4 cm above the aortic bifurcation at the level of the L2 to L3 vertebrae. As the IMA travels inferiorly and to the left, it gives off the left colic artery and several sigmoidal branches. After these branches, the IMA becomes the superior hemorrhoidal (rectal) artery as it crosses over the left common iliac artery. The left colic artery divides into an ascending branch (splenic flexure) and a descending branch (the descending colon). The sigmoidal branches form a fairly rich arcade within the sigmoid mesocolon (similar to that seen within the small bowel mesentery). The superior hemorrhoidal artery carries into the mesorectum and into the rectum. The superior hemorrhoidal artery bifurcates in about 80% of patients.

The Marginal Artery and Other Mesenteric Collaterals

The major arteries noted above account for the main source of blood within the mesentery. However, the anatomy of the mesenteric circulation and the collaterals within the mesentery remain less clear. Haller first described a central artery anastomosing all mesenteric branches in 1786 [74]. When Drummond demonstrated its surgical significance in the early twentieth century, it became known as the marginal artery of Drummond [75, 76]. The marginal artery has been shown to be discontinuous or even absent in some patients, most notably at the splenic flexure (Griffiths' critical point), where it may be absent in up to 50% of patients [77]. This area of potential ischemia is the embryologic connection between the midgut and hindgut. Inadequacy of the marginal artery likely accounts for this area being most severely affected in cases of colonic ischemia. Another potential (though controversial) site of ischemia is at a discontinuous area of marginal artery located at the rectosigmoid junction termed Sudeck's critical point. Surgical experience would question whether this potential area of ischemia exists; a recent fluorescence study indicates that it does [78], though its clinical importance remains in doubt.

Venous Drainage

Venous drainage of the colon largely follows the arterial supply with superior and inferior mesenteric veins draining both the right and left halves of the colon (Fig. 1.10). They ultimately meet at the portal vein to reach the intrahepatic system. The superior mesenteric vein (SMV) travels parallel and to the right of the artery. The inferior mesenteric vein (IMV) does not travel with the artery but rather takes a longer path superiorly to join the splenic vein. It separates from the artery within the left colon mesentery and runs along the base of the mesentery where it can be found just lateral to the ligament of Treitz and the duodenum before joining the splenic vein on the opposite (superior) side of the transverse mesocolon. Dissecting posterior to the IMV can allow for separation of the mesenteric structures from the retroperitoneal structures during a medial-to-lateral dissection.

Lymphatic Drainage

The colon wall has a dense network of lymphatic plexuses. These lymphatics drain into extramural lymphatic channels which follow the vascular supply of the colon. Lymph nodes are plentiful and are typically divided into four main groups. The *epiploic* group lies adjacent to the bowel wall just below the peritoneum and in the epiploicae. The *paracolic* nodes are along the marginal artery and the vascular arcades. They are most filtering of the nodes. The *intermediate* nodes are situated on the primary colic vessels. The *main* or *principal* nodes are on the superior and inferior mesenteric vessels. Once the lymph leaves the main nodes, it drains into the cisterna chili via the para-aortic chain.

Nervous Innervation

The colon is innervated by the sympathetic and parasympathetic nervous systems and closely follows the arterial blood supply. The sympathetic innervation of the right half of the colon originates from the lower six thoracic splanchnic nerves which synapse within the celiac, preaortic, and superior mesenteric ganglia. The postganglionic fibers then follow the SMA to the right colon. The sympathetic innervation for the left half originates from L1, L2, and L3. Parasympathetic fibers to the right colon come from the posterior (right) branch of the vagus nerve and celiac plexus. They travel along the SMA to synapse with the nerves within the intrinsic autonomic plexuses of the bowel wall. On the left side, the parasympathetic innervation comes from S2, S3, and S4 via splanchnic nerves.

Embryology

Though the embryologic development of the GI system is complex, a working knowledge of the development of the small bowel, colon, and anorectum is critical for a colorectal surgeon as it can aid in understanding pathophysiology and is essential for recognizing surgical planes.

Anus and Rectum

The colon distal to the splenic flexure, including the rectum and the anal canal (proximal to the dentate line), is derived from the hindgut and therefore has vascular supply from the inferior mesenteric vessels (Fig. 1.9). The dentate line (Fig. 1.1) is the fusion plane between the endodermal and ectodermal tubes. The cloacal portion of the anal canal has both endodermal and ectodermal components which develop into the anal transitional zone [79]. The terminal portion of the hindgut or cloaca fuses with the proctodeum (an ingrowth from the anal pit).

The cloaca originates at the portion of the rectum below the pubococcygeal line, while the hindgut originates above it. Before the fifth week of development, the intestinal and urogenital tracts are joined at the level of the cloaca. By the eighth week, the urorectal septum migrates caudally to divide the cloacal closing plate into an anterior urogenital plate and a posterior anal plate. Anorectal rings result from a posterior displacement in the septum and the resultant smaller anal opening. By the tenth week, the anal tubercles fuse into a horseshoe-shaped structure dorsally and into the perineal body anteriorly. The external anal sphincter forms from the posterior aspects of the cloacal sphincter earlier than the development of the internal sphincter. The internal sphincter develops from enlarging fibers of the circular muscle layer of the rectum [80]. The sphincters migrate during their development with the internal sphincter moving caudally, while the external sphincter enlarges cephalad. Meanwhile, the longitudinal muscle descends into the intersphincteric plane [9]. In females, the female genital organs form from the Müllerian ducts and join the urogenital sinus by the 16th week of development. In contrast, in males, the urogenital membrane obliterates with fusion of the genital folds, while the sinus develops into the urethra.

Colon and Small Intestine

The endodermal roof of the yolk sac develops into the primitive gut tube. This initially straight tube is suspended upon a common mesentery. By week 3 of development, it has three discernible segments: namely, the foregut, midgut, and hindgut. The midgut starts below the pancreatic papilla to form the small intestine and the first half of the colon (all supplied by the superior mesenteric artery). The distal colon and rectum, as well as the anal canal, develop from the hindgut and are therefore supplied by the inferior mesenteric artery.

Midgut Rotation

There is a normal process by which the intestinal tract rotates (Fig. 1.12) [81]. The first stage is the physiologic herniation of the midgut, the second stage is its return to the abdomen, and the third stage is the fixation of the midgut. Abnormalities in this

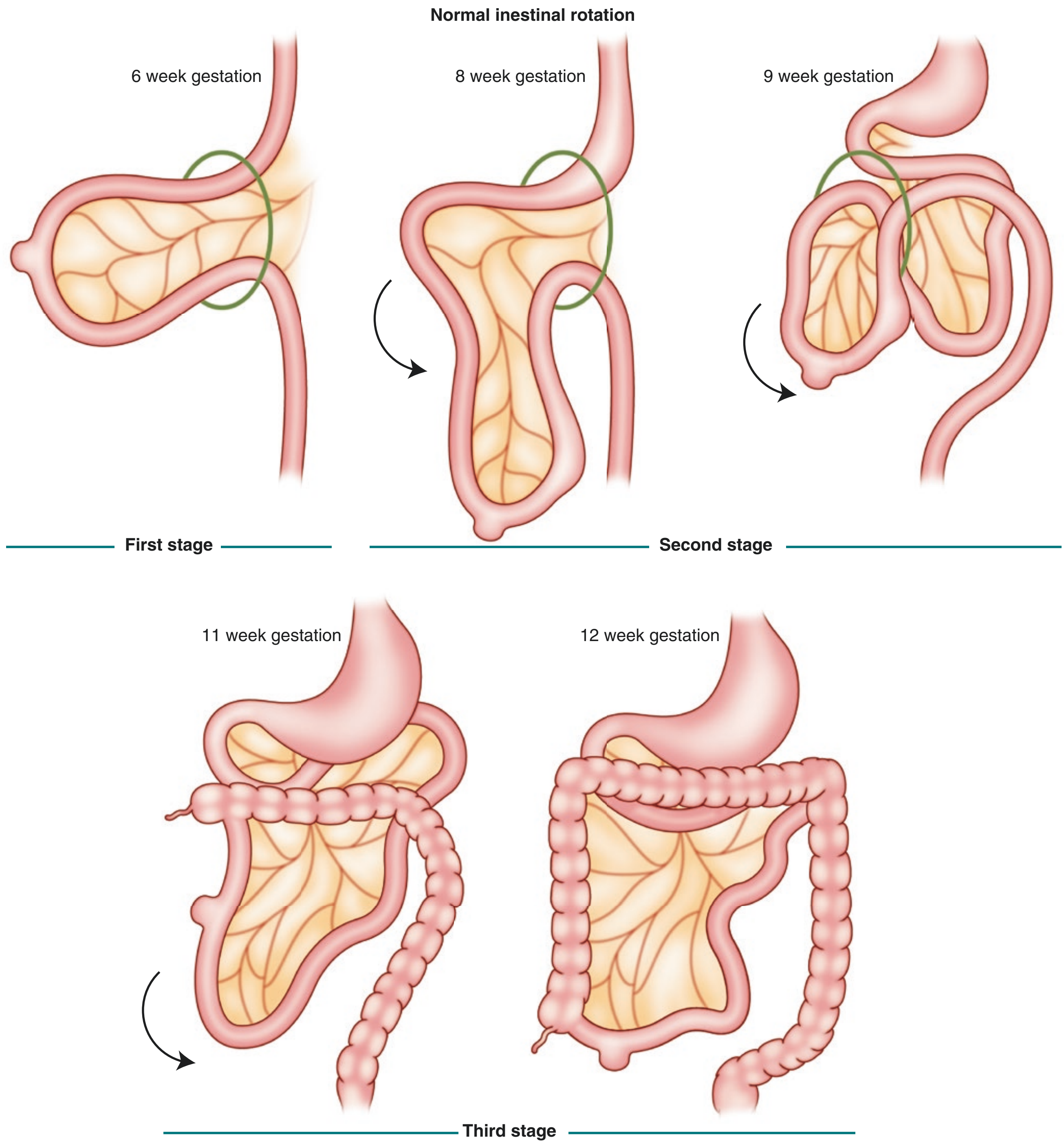


Fig. 1.12 Summary of normal intestinal rotation during development

normal process lead to various malformations (see below). The physiologic herniation (first stage) occurs between weeks 6 and 8 of development. The primitive gut tube elongates over the superior mesenteric artery and bulges out through the umbilical cord (Fig. 1.13). During the eighth week, these contents move in a counterclockwise fashion, turning 90° from the sagittal to the horizontal plane (Fig. 1.14). Anomalies at this stage are rare but include situs inversus, duodenal inversion, and extroversion of the cloaca. During the second stage (tenth week of gestation), the midgut loops return to the peritoneal cavity and simultaneously rotate an additional 180° in the counterclockwise direction (Fig. 1.15). The pre-arterial portion of the duodenum returns to the abdomen first, followed by the counterclockwise rotation around the superior mesenteric vessels, resulting in the duodenum lying behind them. The colon returns after the rotation, resulting in their anterior location. Anomalies in this stage are more common and result in non-rotation, malrotation, reversed rotation, internal hernia, and omphalocele. The third stage (fixation of the midgut) begins once the intestines have returned to the peritoneal cavity and ends at birth. The cecum migrates to the right lower quadrant from its initial position in the upper abdomen (Fig. 1.16). After the completion of this 270° counterclockwise rotation, fusion begins, typically at weeks 12–13. This results in fusion of the duodenum as well as the ascending and descending colons (Fig. 1.17).

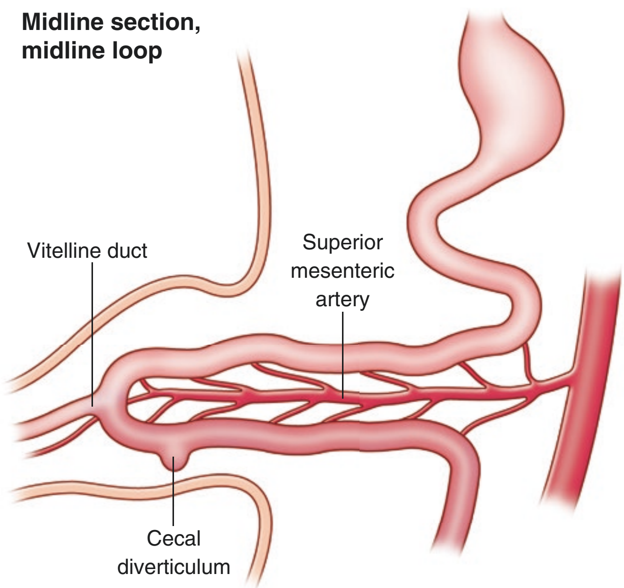


Fig. 1.13 Elongation of the midgut loop

Major Anomalies of Rotation

Non-rotation

The midgut returns to the peritoneum without any of the normal rotation. This results in the small intestine being on the right side of the abdomen and the colon on the left side (Fig. 1.18). This condition can remain asymptomatic (a finding noted at laparoscopy or laparotomy) or result in volvulus affecting the entirety of the small intestine. The twist generally occurs at the duodenojejunal junction as well as the midtransverse colon.

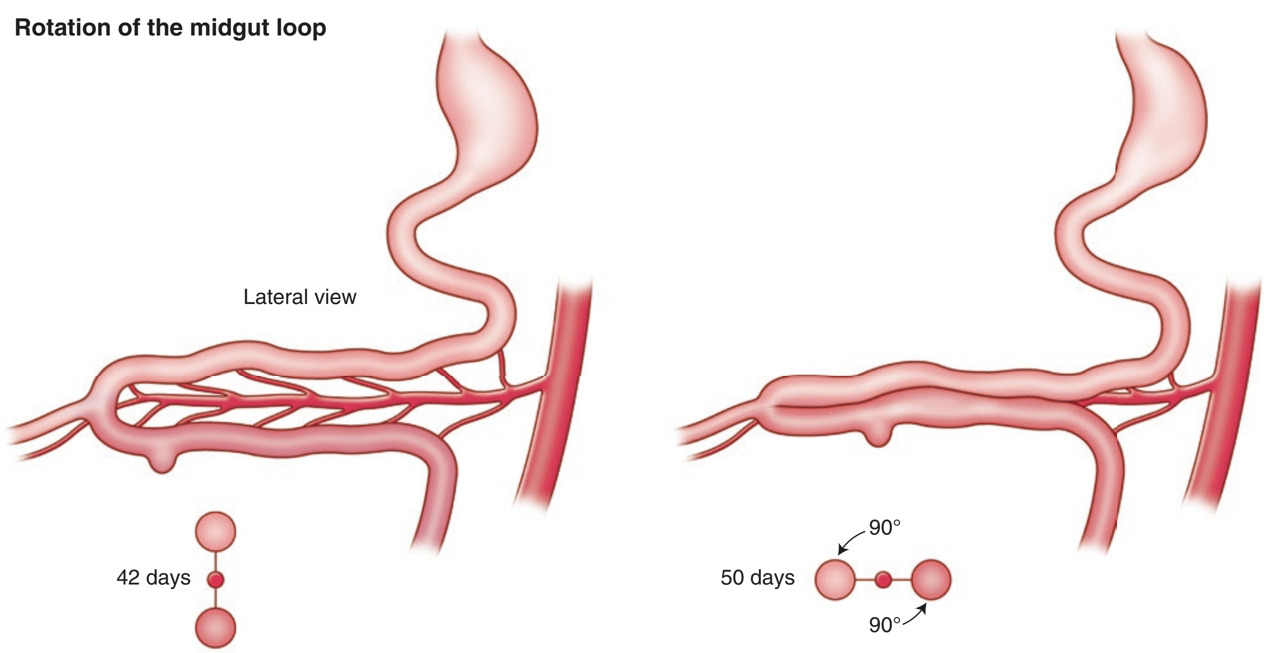


Fig. 1.14 Rotation of the midgut loop

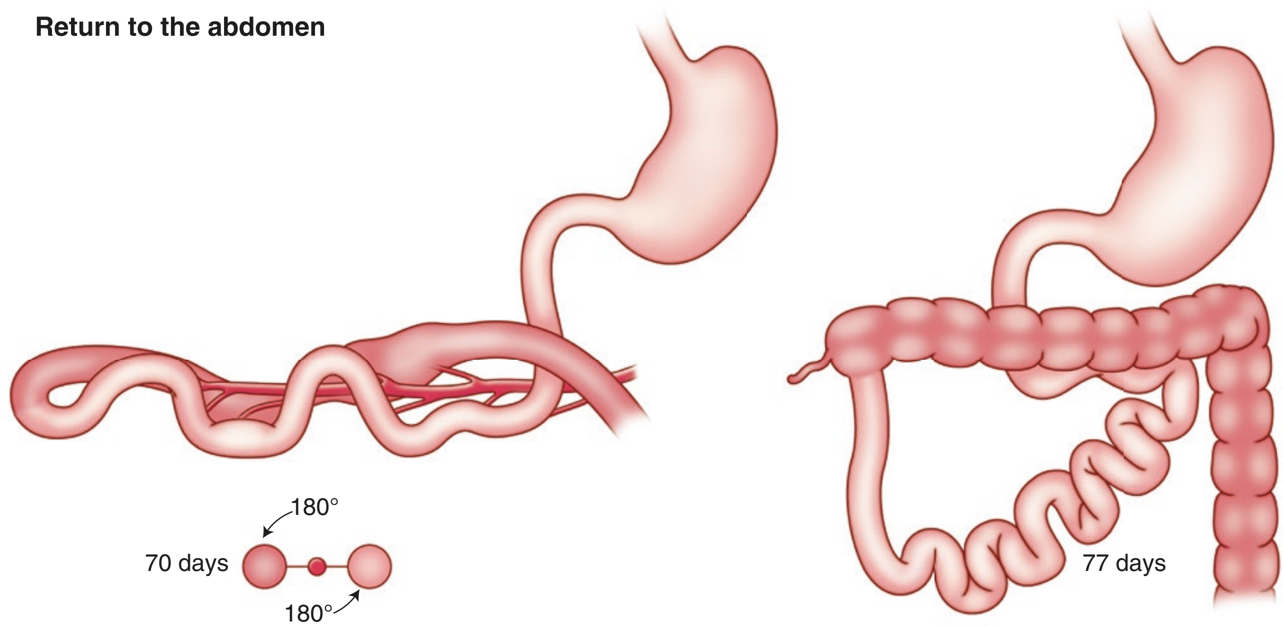


Fig. 1.15 Return of the intestinal loop to the abdomen

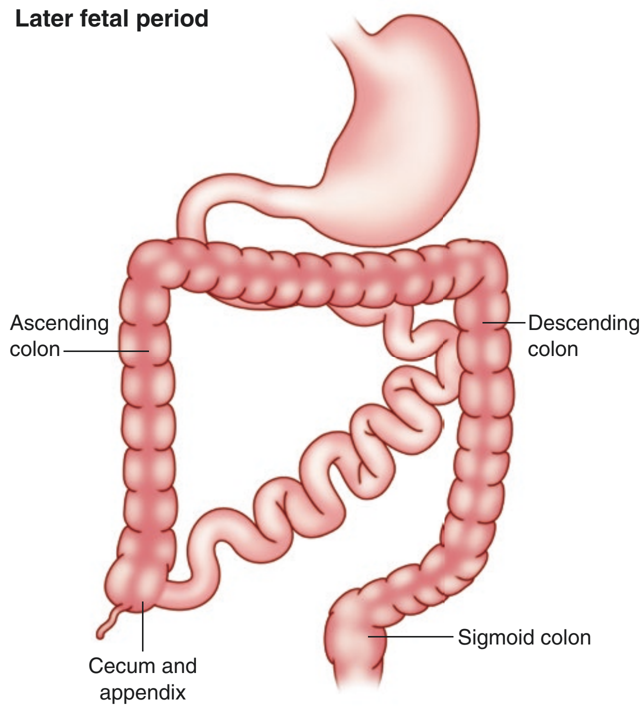


Fig. 1.16 Later fetal development

Malrotation

There is normal initial rotation, but the cecum fails to complete the normal 270° rotation around the mesentery. This results in the cecum being located in the mid-upper abdomen with lateral bands (Ladd's bands) fixating it to the right abdominal wall (Fig. 1.19). These bands can result in extrinsic compression of the duodenum.

Reversed Rotation

Clockwise (rather than counterclockwise) rotation of the midgut results in the transverse colon being posterior to the superior mesenteric artery while the duodenum lies anterior to it.

Omphalocele

An omphalocele is, basically, the retention of the midgut within the umbilical sac and its failure to return to the peritoneal cavity. The bowel remains enclosed in a membrane as it herniates through a defect larger than 4 cm [82].

Internal Hernias

Internal hernias, as well as congenital obstructive bands, can cause congenital bowel obstructions. These are considered failures of the process of fixation (the third stage

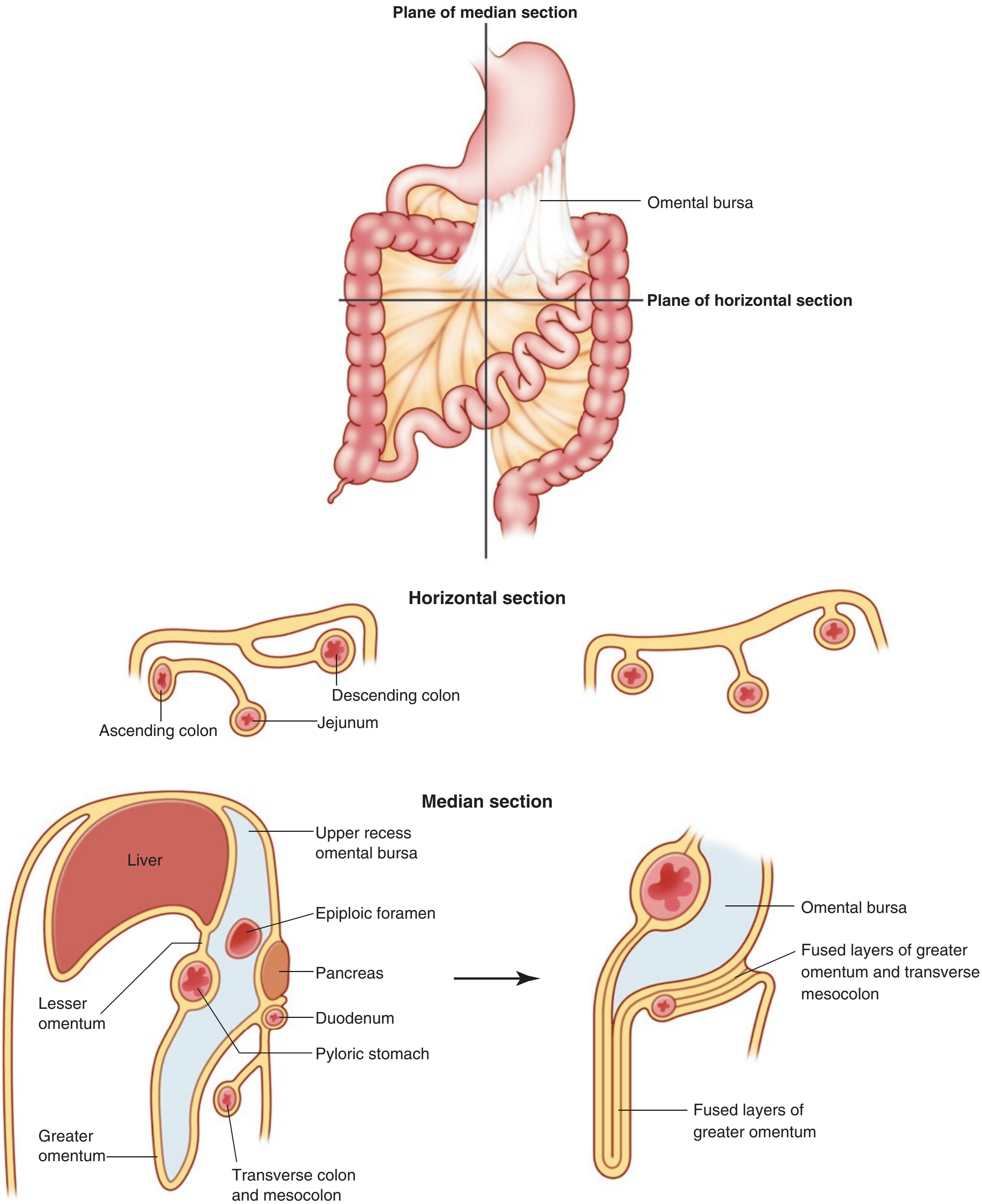


Fig. 1.17 Development of the mesentery and omental fusion

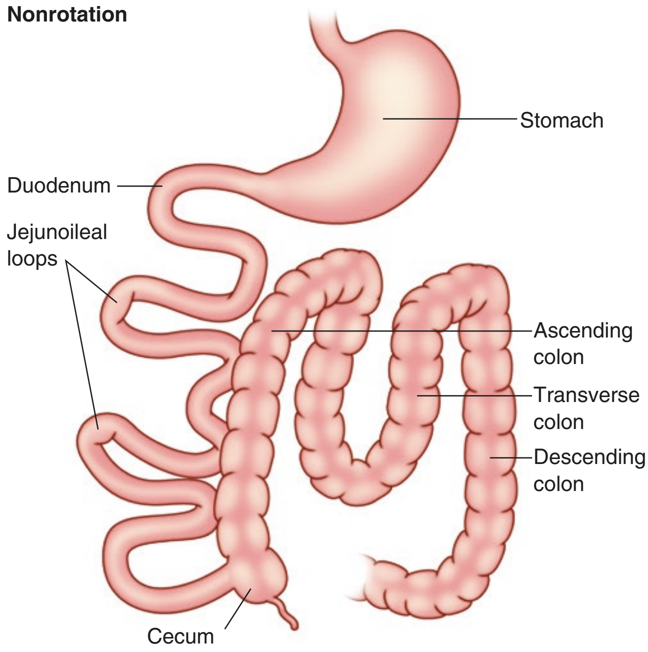


Fig. 1.18 Intestinal non-rotation

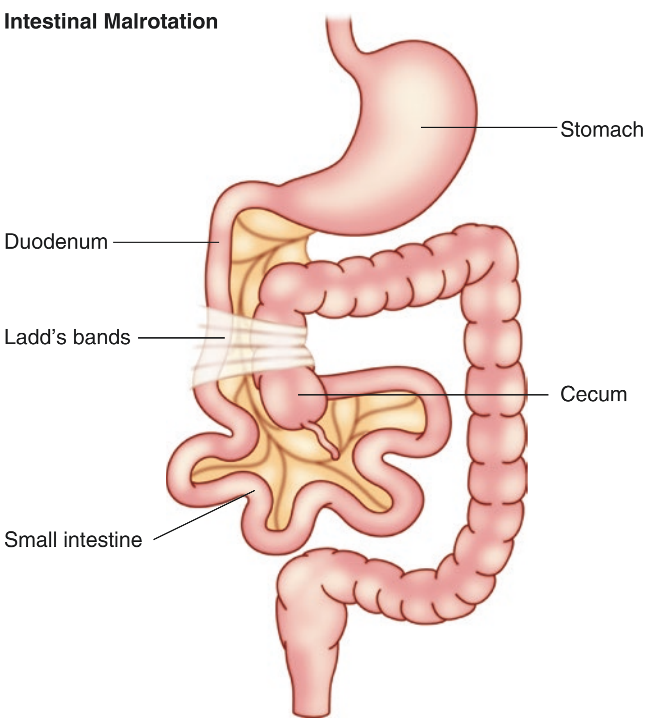


Fig. 1.19 Intestinal malrotation

of rotation). This can be the result of an incomplete fusion of the mesothelium or when structures are abnormally rotated. Retroperitoneal hernias can occur in various positions, most notably paraduodenal, paracecal, and intersigmoid.

Other Congenital Malformations of the Colon and Small Intestine

Proximal Colon Duplication

There are three general types of colonic duplication: mesenteric cysts, diverticula, and long colon duplication [83]. Mesenteric cysts are lined with intestinal epithelium and variable amounts of smooth muscle. They are found within the colonic mesentery or posterior to the rectum (within the mesorectum). They may be closely adherent to the bowel wall or separate from it. They generally present as a mass or with intestinal obstruction as they enlarge. Diverticula can be found on the mesenteric or antimesenteric sides of the colon and are outpouchings of the bowel wall. They often contain heterotopic gastric or pancreatic tissue. Long colonic duplications of the colon are the rarest form of duplication. They parallel the functional colon and often share a common wall throughout most of their length. They usually run the entire length of the colon and rectum, and there is an association with other genitourinary abnormalities.

Meckel's Diverticulum

A Meckel's diverticulum is the remnant of the vitelline or omphalomesenteric duct (Fig. 1.13). It arises from the antimesenteric aspect of the terminal ileum, most commonly within 50 cm of the ileocecal valve. They can be associated with a fibrous band connecting the diverticulum to the umbilicus (leading to obstruction), or it may contain ectopic gastric mucosa or pancreatic tissue (leading to bleeding or perforation) (Fig. 1.20). An indirect hernia containing a Meckel's diverticulum is termed a Littre's hernia. Meckel's diverticulum is generally asymptomatic and, per autopsy series, is found in up to 3% of the population [84]. Surgical complications, which are more common in children than adults, include hemorrhage, obstruction, diverticulitis, perforation, and umbilical discharge. Generally, there is no hard indication for excision of an incidentally discovered Meckel's diverticulum, though its removal is generally safe [85, 86].

Atresia of the Colon

Colonic atresia, representing only 5% of all gastrointestinal atresias, is a rare cause of congenital obstruction. They are likely the result of vascular compromise during development [87]. They vary in severity from a membranous diaphragm blocking the lumen to a fibrous cord-like remnant, on to a complete absence of a segment [88].

Hirschsprung's Disease

This nonlethal anomaly, which is more common in males, results from the absence of ganglion cells within the myenteric plexus of the colon. It is caused by interruption of the normal migration of the neuroenteric cells from the neural

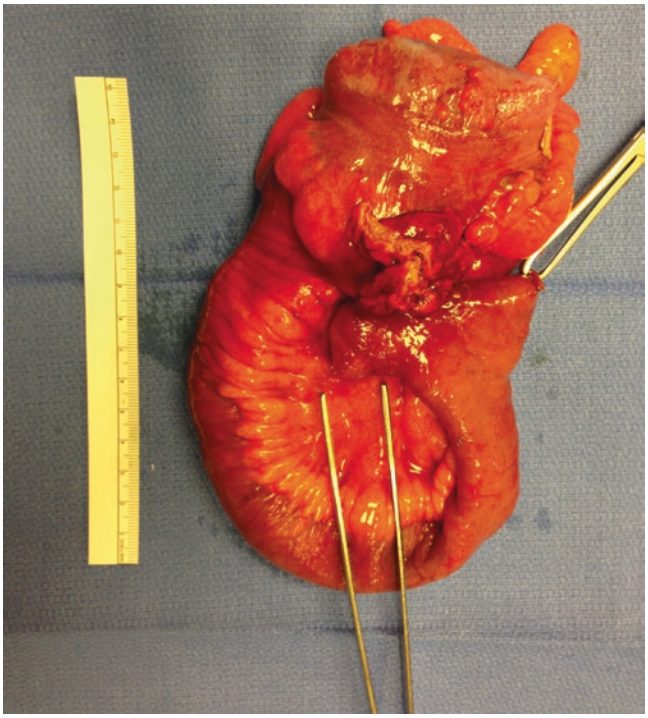


Fig. 1.20 Perforated Meckel's diverticulum with fistula to the ileum

crest before they reach the rectum. This results in dilation and hypertonicity of the proximal colon. The extent of the aganglionosis is variable, though the internal sphincter is always involved. Its severity is dependent upon the length of the involved segment. It will be discussed fully in a subsequent chapter.

Anorectal Malformations

Abnormalities in the normal development of the anorectum can be attributed to "developmental arrest" at various stages of normal development. These abnormalities are often noted in concert with spinal, sacral, and lower limb defects, as noted by Duhamel, and theorized to be related to a "syndrome of caudal regression" [89]. Indeed, skeletal and urinary anomalies are associated in up to 70% [90], while digestive tract anomalies (e.g., tracheoesophageal fistula or esophageal stenosis) and cardiac and abdominal wall abnormalities are also noted in patients with anorectal anomalies.

Anal Stenosis

While anal stenosis in a newborn is relatively common, noted in 25–39% of infants, symptomatic stenosis is only noted in 25% of these children [91]. The majority of these children undergo spontaneous dilation in the first 3–6 months of life.

Membranous Atresia

This very rare condition is characterized by the presence of a thin membrane of skin between the blind end of the anal canal and the surface. It is also termed the covered anus. It is more common in males.

Anal Agenesis

The rectum develops to below the puborectalis where either it ends in an ectopic opening (fistula) in the perineum, vulva, or urethra or it ends blindly (less commonly). The sphincter is present at its normal site.

Anorectal Agenesis

Anorectal agenesis is the most common type of "imperforate anus." More common in males, the rectum ends well caudal to the surface, and the anus is represented by a dimple with the anal sphincter usually being normal in location. In most cases, there is a fistula to the urethra or vagina. High fistulae (to the vagina or urethra) with anorectal agenesis develop as early as the sixth or seventh week of gestation, while the low fistulae (perineal) or anal ectopia develop later, in the eighth or ninth week of development.

Rectal Atresia or "High Atresia"

In rectal atresia, the rectum and the anal canal are separated from one another by an atretic portion. It is embryologically the distal most type of colon atresia but is still considered an anorectal disorder clinically.

Persistent Cloaca

This rare condition, which only occurs in female infants, is the result of total failure of descent of the urorectal septum. It occurs at a very early stage of development.

Acknowledgments This chapter builds on previous chapters written by José Marcio Neves Jorge and Angelita Habr-Gama in the first and second editions of this textbook and by Steven Mills and Joseph Carmichael in the third edition of this textbook.

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