

The reader may be surprised for not finding in this chapter many of the traditional terms historically used to refer to the different portions of the sphincter mechanism of the human being. We use a different terminology that we believe is realistic, useful, and with important practical and technical implications for the practicing surgeon. This is a result of our observations of the different anatomic variants, found in more than 2,032 surgical explorations of the pelvis and the anorectum of patients suffering from anorectal malformations, as well as many others operated to resect tumors and to repair pelvic organs (urethra, vagina, and rectum).

In the early times, when we performed the first posterior sagittal approaches to repair anorectal malformations, influenced by the traditional concepts expressed in the available textbooks on the subject [1, 2], we were looking for the “puborectalis sling,” the “pubococcygeal muscle,” the “pubourethralis muscle,” the “iliococcygeal muscle,” the “superficial portion of the external sphincter,” the “deep portion of the external sphincter,” and the “internal sphincter,” and we were rather frustrated for not seeing what was described in the textbooks. Or at least, what we were seeing was very different to what was described.

In addition, through the years we found that what we observed as part of the sphincter mechanism in one patient was never the same as the one that we saw in another one. After a large experience with the surgical treatment of anorectal malformations, we are now certain to believe that we are dealing with a spectrum of anatomic variations.

We are aware of the fact that human beings (and surgeons are not exceptions) prefer to deal with artificial, man-made, classifications to refer and to discuss biological phenomenon. Yet, we like to say that Mother Nature does not like our classifications and continues producing humans, animals, and biological specimens following a spectrum type of pattern. We recognize that it is not easy from the clinical point of view to talk about spectrums, but we could not ignore reality.

Figure 2.1 shows what we think is the best photograph ever taken of the pelvic anatomy of a male cadaver. The authors managed to show a perfect sagittal section. We feel admiration and respect for such achievement. This magnificent photograph was reproduced with permission from the excellent “Colon Atlas of Human Anatomy” by R.M.H. McMinn Emeritus Professor of Anatomy, Royal College of Surgeons of England and University of London and R.T. Hutchings, photographer, formerly Chief Medical Laboratory Scientific Officer, Royal College of Surgeons of England. Year Book Medical Publishers, Inc., Chicago 1977, page 248.

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Fig. 2.1 Photograph of a sagittal section of a human cadaver. 1 Rectus abdominis; 2 Extraperitoneal fat; 3 Sigmoid colon; 4 Promontory of sacrum; 5 Rectum; 6 Coccyx; 7 Anococcygeal body; 8 External anal sphincter; 9 Anal canal with anal columns of mucous membrane; 10 Perineal body; 11 Ductus deferens; 12 Epididymis; 13 Testicle; 14 Spongy part of urethra and corpus spongiosum; 15 Corpus cavernosum; 16 Bulbospongiosus; 17 Perineal membrane; 18 Sphincter urethrae; 19 Membranous part of urethra; 20 Pubic symphysis; 21 Prostate gland; 22 Prostatic part of urethra; 23 Seminal colliculus; 24 Bristle in ejaculatory duct; 25 Internal urethral orifice; 26 Bladder; 27 Bristle passing up into right ureteral orifice; 28 Rectovesical pouch

Figure 2.2 shows an MR image of the sagittal section of the pelvis of a normal child. It is really remarkable what modern imaging technology has achieved. We invite the readers to look in a very detailed way these two pictures (Figs. 2.1 and 2.2). The same muscle structures shown in the cadaver are present in the MRI picture. A non-biased, objective view of these two figures does not allow us to identify separately the puborectalis, ischiococcygeal, pubourethralis, and iliococcygeal muscles and deep external sphincter, superficial external sphincter, and internal sphincter.

Rather, one can see a muscle structure that comes from the anterior aspect of the lowest

part of the sacrum and coccyx and runs all the way down to the skin attached to the posterior rectal wall. It actually runs in continuum and one cannot see any hint of separation of different structures. In our observations of normal human beings (not only in cases of anorectal malformations) operated posterior sagittally for other reasons (tumors, trauma), and using an electrical stimulator, we have always been able to identify this strong funnel-like muscle structure. If one touches that muscle structure in the upper portion, one can elicit a contraction that pushes the rectum forward as described for the levator muscle. If one touches the lowest part of that muscle, one can see an elevation of the anus, and perhaps that is why originally this structure was called levator muscle. That portion of this sphincter mechanism made mainly of vertical fibers running parallel to the rectum we call it “muscle complex,” to differentiate it from the upper portion (levator) that is made out of horizontal fibers that compress the rectum from behind. However, these different portions of the sphincter mechanism do not contract separately; in real life, they contract in a massive unified way (Animations 2.1, 2.2, 2.3, 2.4, and 2.5). The muscle fibers running longitudinally (parallel to the rectum) elevate the anus when they contract, whereas those fibers that surround the rectum are the ones that produce the compression of the rectum posteriorly. Perhaps the point of maximal contraction of this funnel-like muscle structure is what has been considered the puborectalis sling, but we must emphasize that there is no real anatomic separation of these structures. Near the skin, the funnel-like muscle joins other types of muscle that run superficial under the skin and divide into two portions, one on each side of the anus, and that is why we call those “parasagittal fibers.” The contraction of these muscles may produce the impression of closing the anus in a circular fashion; yet, actually these are parasagittal fibers that join one side with the other, posterior and anterior to the anal opening. The parasagittal fibers run actually perpendicular to the muscle complex. We arbitrarily use the term “levator” to refer to the upper portion of the funnel-like muscle mecha-



Fig. 2.2 MRI of a sagittal section of the normal pelvis. (a) Relaxed sphincter mechanism. (b) Contracted sphincter mechanism

nism and use the term “muscle complex” to its lower portion. The point where the muscle complex and parasagittal fibers cross represents the limits of the sphincter.

When the anatomy of the anorectal sphincters is presented in this rather simplistic way, it is easy to teach and to learn.

Traditional anatomy concepts have been repeated from one generation to another, full of details or concepts without clinical relevance. The medical students are frequently obligated to learn some of those concepts.

We have been impressed by publications related with the anatomy of the sphincters; the authors frequently show real photographs of the anatomy, yet they over-impose arrows to show inexistent, imaginary structures [2–5]. In other words, photographs show the real anatomy, whereas the diagrams or the arrows show what the authors wanted to see.

More recently, advances in the technology of imaging show images (Fig. 2.2) of the real anatomy; however, again the authors fabricate other-

wise unnecessary diagrams showing their preconceptions and biases [6–13].

It is even more impressive to see how rather bizarre anatomic concepts, like the idea of a “triple loop” without any evidence to support its existence, are accepted, published, and repeated between surgeons [14, 15].

In a medical community where everybody seem to be able to see clearly structures such as the puborectalis muscle and all the other portions of the sphincter mechanism, very few dare to express skepticism and disagree [16, 17]. During our literature review, we found an excellent honest paper written by Dr. Arthur F. Dalley II, PhD, [17]. After a very thoughtful discussion, he concludes saying: “I recommend that the three-part external anal sphincter be removed from gross anatomy texts, dissectors and atlases and be relegated to the junkyard of anatomic trivia where it may languish for the sake of the historical anatomist or the rare individual who spends time carving out the most meticulous of dissections.” This is a paper that all colorectal surgeons must read.

2.1 Internal Sphincter

This elusive structure has been described as a thickening of the circular layer of smooth muscle of the bowel in the area of the anorectum. The literature related to this structure, in general, has the following characteristics:

- There are no good quality photographs showing the sphincter.
- There is no precise description of its size and limits at different ages.
- Most papers discuss the “internal sphincter” based on manometric findings.
- Some authors believe that the “internal sphincter” is very important for bowel control [18, 19].
- Others believe that its contribution for bowel control is not significant [20].
- Many authors believe that the lack of relaxation of this structure is responsible for many patients suffering from constipation and megarectosigmoid (see Chap. 25).
- Our direct observations of the entire posterior anorectal wall in normal individuals did not allow us to see the thickening of the smooth muscle that presents the “internal sphincter.”
- As a consequence of all the importance that many doctors gave to the “internal sphincter,” pediatric surgeons have been debating about the possible existence of an “internal sphincter” in cases of anorectal malformations, located at the most distal part of the bowel, the portion attached to the urogenital tract (fistula). Some surgeons believe that it is very important to preserve the most distal portion of the bowel in order to guarantee bowel control [21–23], whereas others believe that it must be resected to avoid constipation [24]. Our results in terms of bowel control show that fecal continence and constipation are unrelated to the preservation or resection of that portion of the bowel (see Chap. 15).

All these descriptions of our literature findings related with the “internal sphincter,” plus our concepts discussed in Chap. 25, explain our skepticism related with the existence, function, and relevance of this structure.

2.2 General Anatomic Principles in Anorectal Malformations

Figures 2.3, 2.4, 2.5, and 2.6 show the anatomic variations that form part of the spectrum seen in cases of anorectal malformations. Animation 2.1 shows the sagittal view of the anatomy of the pel-



Fig. 2.3 Diagram showing the most common anatomic sphincter pattern seen in patients with perineal fistula



Fig. 2.4 Diagram showing the most common anatomic sphincter pattern seen in patients with bulbar fistula



Fig. 2.5 Diagram showing the most common anatomic sphincter pattern seen in patients with prostatic fistula

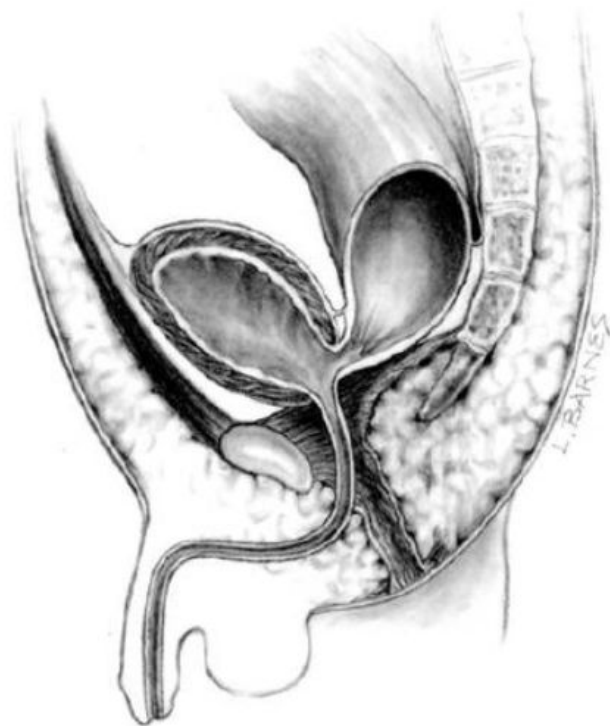


Fig. 2.6 Diagram showing the most common anatomic sphincter pattern seen in patients with bladder neck fistula

vis of a normal human being. Figure 2.3 shows the most benign of all defects which is the perineal fistula. The sphincter mechanism in these types of cases is almost normal. The rectum, on the other hand, deviates in its lowest portion to

open in the perineal body, anterior to the center of the sphincter. The rectum is dilated.

Figure 2.4 shows the anatomy of a patient born with a rectourethral bulbar fistula. Most of these patients have a sphincter mechanism reasonably good, perhaps not as strong and good as the sphincter of a normal person or a patient with a perineal fistula. The rectum connects to the lowest portion of the posterior urethra which we call bulbar urethra.

Figure 2.5 shows the anatomy on a patient with a rectoprostatic fistula. One can see that the sphincter mechanism is much more primitive and weak. In addition, the distance between the sacrum and the pubis is significantly shorter. The available space for a pull-through is getting smaller, the sphincter mechanism weaker, and obviously the prognosis is not as good as in the previous defects.

Figure 2.6 shows the anatomy of a patient with a recto-bladder neck fistula. The rectum opens at the bladder neck, the sphincter mechanism is very tenuous, sometimes almost nonexistent, and the distance between the sacrum and the pubis is very short. Sometimes it makes it almost impossible to pull the rectum down. As one can see in this series of diagrams, in the case of the rectourethral bulbar fistula, once we separate the rectum from the urethra, it is conceivable that the rectum can be placed within the limits of the sphincter with minimal mobilization and will be completely covered by the sphincter mechanism. Whereas in Fig. 2.5, sometimes we find a rectum that does not fit into the tenuous, delicate sphincter mechanism, and one has the feeling that the reconstruction was not ideal. And finally, in some of the recto-bladder neck fistulas, it becomes very obvious that the sphincter is very weak and that the patient most likely will have fecal incontinence. In fact, as we will discuss in our results, patients with recto-perineal fistulas have a 100 % chance of bowel control provided they have a good operation. Patients with bulbar fistula have 85 % chances, prostatic fistula 60 % chances, and bladder neck fistula only 20 % chances. Similarly, characteristically, the sacrum usually is more and more primitive, as we go into higher and higher

locations of the rectum, as well as the characteristics of the sphincter and the space available between the sacrum and pubis. The chance of suffering from tethered cord also increases in higher malformations. The exception is represented by some patients with perineal fistulas. This particular group of patients has more tendencies to suffer from presacral masses and tethered cord. The anatomic differences shown in Figs. 2.3, 2.4, 2.5, and 2.6 and Animations 2.2, 2.3, 2.4, 2.5, and 2.6 are seen in real life. However, as it is well known by all surgeons, in medicine and surgery, there are no “nevers” and there are no “always.” In other words, it is possible to see, although very unusual, a perineal fistula with very poor sphincter mechanism and also to see a recto-bladder neck fistula with a rather good-looking sphincter mechanism, but those are exceptions. Most of the time, as the rectum is located higher connecting to the urogenital tract, the sacrum tends to be shorter, the distance between the sacrum and pubis decreases, and the characteristics of the sphincter become more and more rudimentary and weak.

2.3 Nerves

We surgeons frequently refer to the “lack of nerves” when we deal with patients with anorectal malformations with bad functional prognoses. Yet, we actually never see the nerves when operating on these patients. There are no precise scientific studies that give evidence of the presence or absence, as well as characteristics and precise location, of the pelvic nerves in cases of anorectal malformations. We work, always assuming, that the higher the location of the rectum and its connection to the urogenital tract, as well as the more deficient sacrum is, the more deficient the nerves are. In addition, the fact that we have to mobilize the rectum from higher distances most likely means that we have to sacrifice more nerves. We also assume that in a patient with absent sacrum, all the nerves that normally come out of the segments of the sacrum and innervate the pelvic organs are absent, or deficient, and that may explain why patients with absent sacrum have

zero possibility of having bowel and urinary control. Patients with anorectal malformations are represented by a spectrum in terms of sacrum deficiency that goes from patients with normal sacrum to patients with completely absent sacrum.

We know that under normal circumstances, the nerves that innervate the bladder neck urinary tract and corpora come from the orifices of the sacrum and run lateral to the rectum in order to reach the corpora, the bladder neck, and the rectum itself. Therefore, from the early times, Dr. Douglas Stephens [25] recommended (and is still valid) to try to remain exactly in the midline as much as possible during our surgical explorations. Once the surgeon reaches the rectum, all the dissection of the rectum must be performed, staying as close as possible to the rectal wall to avoid the damage of nerves that supposedly run lateral to the rectum. In other words, the further away from the rectum, the more chances to injure nerves.

Concerning the anatomy of the pelvic autonomic nerves, the reader is referred to anatomy books and an excellent paper [26]. From the reading of that material, we reinforce our belief that it is essential to remain in the midline while approaching the pelvis and to dissect the rectum remaining as close as possible to the bowel wall, in order to minimize the possibilities to injure autonomic nerves and avoid neurogenic bladder and impotence.

We have evidence from patients that had previous failed attempted repairs and that were born with a “good” malformation and yet they suffered from fecal and sometimes urinary incontinence. Reading the operative reports of those patients, one finds that the surgeon actually got lost and certainly went out of the midline, which may explain the nerve damage.

2.4 Blood Supply

Unfortunately, there are no scientific detailed anatomic studies of the blood supply of the pelvic organs of patients with anorectal malformations. Again, what we have learned from our surgical

explorations is that it seems like the rectum has an excellent intramural blood supply, as evidenced by the fact that in every case of anorectal malformation that we operate on, we separate the rectum from the urogenital tract, and then we have to mobilize the rectum enough to reach the perineum. In order to do that, we performed a circumferential dissection, dividing all the extrinsic vessels and bands that hold the rectum up in the pelvis. In other words, we are basically devascularizing the rectum. We sacrifice its entire extrinsic blood supply. Yet, provided the wall of the rectum remains intact and the inferior mesenteric vessels are preserved, the intramural blood supply of the proximal part provides enough to maintain alive the distal part of the rectum. This finding should not be extrapolated to other parts of the colon. The blood supply of the colon in patients with anorectal malformations is basically the same as in normal individuals, except in patients with cloacal extrophies and patients with a malformation called "rectal pouch" in which the entire colon is represented by a single saccular piece of colon with a very abnormal bizarre type of blood supply (picture).

2.5 Basic Physiology Principles of Bowel Control

We take care of many babies born with severe anatomic deficiencies; we can certainly repair their anatomy, but we cannot restore their normal function; however, we try to help them to have a normal social life, with the implementation of our "bowel management program." Unfortunately, there is another large group of patients who suffer from fecal incontinence as a consequence of a technically deficient operation. That is obviously something highly regrettable, but most important is the fact that it is preventable. For that, it is imperative for the surgeon to know a few basic but extremely important anatomic and physiologic principles.

In order to have bowel control, it is necessary to have three very important elements:

- A. Sensation
- B. Sphincter

C. Rectosigmoid motility and reservoir function

A. *Sensation* – this is the first indispensable element for bowel control. The anal canal is perhaps the most sensitive part of the human body. There, we are capable of discriminating gas, from liquid and from solid fecal matter [27]. The anal canal remains collapsed by the effect of the muscle tone of the sphincter mechanism that surrounds it. When the fecal matter (liquid, solid, or gas) reaches the anal canal due to active rectal peristalsis, we perceive it and, depending on the surrounding circumstances, decide to use our voluntary sphincter mechanism, to occlude the lumen of the anal canal and avoid a bowel movement, until the circumstances are appropriate to have a bowel movement.

Above the anal canal, in the rectum, we do not have the exquisite sensation described for the anal canal. However, a distention of the rectum with a balloon causes a vague sensation of fullness that is known as proprioception [28].

The implications of these facts for us surgeons are obvious. We must try to preserve intact the anal canal during our operations in patients with normal anal canal, such as patients suffering from Hirschsprung's disease, severe constipation, inflammatory bowel disease, and familial polyposis.

The overwhelming majority of patients with anorectal malformations are born without an anal canal, except for a rare malformation called rectal atresia. Patients with perineal fistula have a rather primitive anal canal. This means that patients with anorectal malformations, under the best circumstances, do not have "perfect" bowel control. Many patients born with a malformation with good functional prognosis behave like normal children, yet when they have a severe episode of diarrhea, it becomes evident that they are less than perfect in terms of bowel control.

B. *Sphincter* – the voluntary sphincter mechanism maintains a certain tone constantly. However, there is a common misconception consisting on believing that a sudden relaxation of the sphincter will produce escape of

feces. That is simply not true. In fact, a human being can only have a bowel movement when the rectosigmoid has a peristaltic wave that pushes the stool out. Human beings actually use the voluntary sphincter occasionally, to prevent the passing of gas or fecal matter in inadequate circumstances.

- C. *Rectosigmoid motility and reservoir function* – after many years of working with patients suffering from bowel control problems, we came to realize that rectosigmoid motility and its reservoir function are the most important element for bowel control. The rectosigmoid remains relaxed most of the time, acting as a reservoir of fecal matter. That is an extremely important function, since it allows human beings to have a social life and only use the toilet every 24–48 h.

The implications of this, for us surgeons, are also very obvious. The removal of the rectosigmoid and connection of a more proximal portion of the colon, to the anal canal, means elimination of the reservoir function which results in an almost constant attempt of the colon to empty. If the anal canal is maintained intact, the resection of the rectosigmoid will result in many bowel movements during the day and a constant effort to avoid them. We all have seen how a colostomy works. It passes stool almost constantly. An ascending colostomy will be passing more often liquid stool, and as we move distally, the stool becomes more solid and the peristalsis less active. That means that connecting a piece of colon directly to the anal canal (without reservoir) will produce very frequent bowel movements, and it will require a well-preserved anal canal to maintain continence.

We can easily imagine what happens when we remove the natural reservoir (rectosigmoid) and damage or resect the anal canal. The result will be permanent fecal incontinence.

Another scenario is the case of an absent anal canal and sphincter; we have seen that in cases of trauma. The motility of the rectosigmoid and its reservoir function is preserved, and therefore, it is possible that a particular patient behaves like if he/she was fecally continent, provided he/she

does not have episodes of diarrhea or multiple, irregular, unpredictable bowel movements.

Using our imagination, we conceive the possibility of manipulating the rectosigmoid motility, using pharmacologic agents, in order to paralyze the rectosigmoid when required and to provoke a peristaltic wave to empty the rectosigmoid when the surrounding circumstances are appropriate. In fact, we consider that kind of treatment more likely to be successful rather than trying to reconstruct the sphincters or use artificial sphincters, without taking into consideration the two more important elements that are sensation and motility.

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