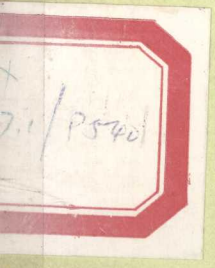
Alberto Peña

Atlas of Surgical Management of Anorectal Malformations



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Atlas of Surgical Management of Anorectal Malformations

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To all the unfortunate children of the
world who suffer from congenital defects.

Preface

The first half of this century was characterized in pediatric surgery by the advent of new surgical techniques designed to treat congenital defects. This, of course, required a detailed description of the basic anatomy of each one of the most common defects including esophageal atresia, intestinal atresia, pyloric stenosis, diaphragmatic hernia, etc. Thus, with relatively small effort, pioneers, giants of this century, made remarkable contributions in the field of pediatric surgery, which allowed the repair of those defects, integrating to normal life many previously unfortunate children.

After around 1950, there were few remaining congenital malformations that were not susceptible to surgical treatment in the field of general pediatric surgery. Therefore, the main advances in pediatric surgery after that time have been mainly related to the medical care of those patients subjected to surgery. These include parenteral nutrition, adequate management of antibiotics, progress in the knowledge of metabolic concerns of the surgical patient, monitoring, anesthesia, ventilatory support, and miniaturization of monitoring devices. All these advances work to the surgeon's advantage, allowing performance of surgical procedures in a calm, well-illuminated field, changing many of the previous urgent procedures into elective operations.

The conjunction of improved surgical techniques with progress in the medical care of the surgical patients has brought us to the present era in which we almost take for granted that children with different types of atresias, congenital defects of the heart, lungs, and G.I. tract now survive, when properly treated in most American and European hospitals.

However, we are now learning that many times, after we have repaired a congenital defect, the baby still may continue to show signs or symptoms of organ malfunction. Thus, we are now facing problems that cannot be treated surgically, such as pulmonary hypertension and persistent pulmonary vascular fetal pattern in babies with diaphragmatic hernias; peristaltic esophageal problems and tracheomalacia in cases of esophageal atresia; repeated bouts of jaundice (cholangitis) in

children with biliary atresia after a portoenterostomy; not to mention our inability to treat babies who have had massive bowel resection, severe dysplastic kidneys, and many other defects. Facing these great challenges now requires exploring the very basic foundations of the problems to find adequate solutions. This requires a much greater effort for diminishing reward and progress. Thus, we are now privileged to live in the era of transplants, artificial organs, fetal surgery, and manipulation of the genetic material.

One of the few areas of pediatric surgery in which the knowledge of basic anatomy was still very limited, even in the 1980s, was anorectal malformations. Since 1980 the approach of these defects through a wide posterior sagittal incision, with the use of an electrical stimulator, has allowed us to explore the basic anatomy of these defects in a detailed manner and also to establish important correlations between the external anatomy, the internal anatomy, surgical techniques, and clinical results.

Exploration in the above-mentioned way has exposed us to an anatomic area that previously was a matter of speculation. Prior to this approach, the only access to these defects was either through the perineum, the abdomen, or a combination of the two. Surgical procedures frequently involved blind maneuvers with the consequent risk of injuring important structures. The rectum was pulled down through a path that was assumed to be the right one. These assumptions were based on a few anatomical postmortem dissections, which were hardly representative of the entire spectrum of defects. A single common procedure would be used for different defects, ignoring anatomic details that now are considered important. The most obvious of these is the fact that many times there is a marked discrepancy between the size of the rectum and the available space through which it must be pulled. This could only be recognized by directly exposing the anatomy of the area. The experience obtained in 400 cases operated on by a single surgeon is documented here in a series of illustrations aimed at helping the pediatric surgeon treat these defects in a technically adequate way.

In the 1980s, even in the face of the fantastic progress of medicine, sound surgical techniques are still crucial if adequate function is to be achieved through the correction of basic human defects. We cannot, therefore, underestimate the value of technically and anatomically sound surgery. This atlas is meant to be a tribute to the art of surgery.

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ALBERTO PEÑA

Contents

1
Important Basic Considerations

Incidence

This defect occurs with a frequency of approximately 1 to 4 or 5,000 newborns [1–3]. It seems to be more frequent in males than in females.

Types of Defects (Classification)

Anorectal malformations have been classified in different ways [4–5]. Here, these defects are grouped mainly on the basis of their specific potential to attain continence which in turn depends on their anatomic characteristics. The following list includes the most common defects:

Male Defects

Low Defects. Cutaneous fistula, anal stenosis, anal membrane and "bucket handle" malformation
Rectourethral bulbar fistula
Rectourethral prostatic fistula
Rectovesical (bladder neck) fistula
Imperforate anus without fistula
Rectal atresia and stenosis

Female Defects

Cutaneous (perineal) fistula
Vestibular fistula
Vaginal fistula
Imperforate anus with no fistula
Rectal atresia and stenosis
Persistent cloacas

Complex Defects

Group of unusual heterogeneous defects.

Basic Anatomy

Normal Individual

Voluntary Muscles

In a normal individual, the voluntary striated muscle structures responsible for fecal control are represented by a funnel-like muscle structure that inserts in the pubic bone, the lowest part of the sacrum, and the middle portion of the pelvis. From there, this diaphragm-like muscle structure extends medially surrounding the rectum all the way down to the perianal skin (Figure 1-1). The upper portion of this funnel-like structure is known as the "levator muscle" and the lowermost part as the "external sphincter." Different slings and subdivisions of these structures have been described, including the "ischiococcygeus," "ileococcygeus," "pubococcygeus," "puborectalis," "deep external sphincter," and "superficial external sphincter."

On the basis of the experience obtained through the posterior sagittal surgical exploration of normal individuals, for the purpose of removing pelvic tumors or repairing sequellae from trauma, the author has been unable to differentiate the different portions or slings already mentioned. Rather, there seems to be a continuum of striated muscle that runs around the rectum and parallel to it, down to the skin (Figure 1-1).

The external sphincter seems to be represented by a group of parasagittal muscle fibers that meet anterior and posterior to the anus (Figures 1-1, 1-2, and 1-3). The muscle fibers that come from the upper insertion of the levator muscle run parallel to the rectum and therefore, perpendicular to these parasagittal fibers (Figures 1-4 and 1-5). The upper portion of the funnel is known as the levator muscle, and the lowest part is known as an external sphincter. We called the intermediate portion (vertical fibers) "muscle complex" [6–8]. In reality, there is no separation between one group of fibers and another. Vertical fibers' "muscle complex" and parasagittal fibers create two corners at the point where they meet, which represent the anterior and posterior limit of the anus. The electrical stimulation of the upper portion of the levator reveals a contraction which pulls the rectum forward. The same type of stimuli, when used at the level of the muscle complex, elicits an elevation of the anus, and the same stimuli, when applied to the parasagittal fibers, will provoke a contraction following the direction of those fibers that tend to close the anus, giving the false impression that these are circular fibers (Figure 1-4). Figure 1-2 shows the effect of the muscle contraction.

The striated muscle urethral sphincter is part of this funnel-type muscle structure and represents an anterior prolongation of it. In the female, this muscle extends along both sides of the entire urethra, as demonstrated by electrical stimulation in cases of persistent cloaca.

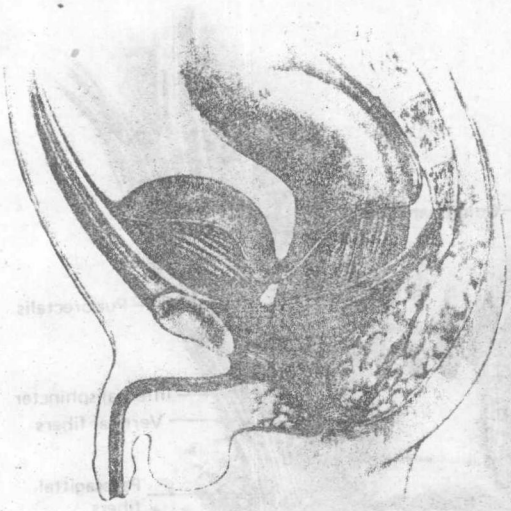

FIGURE 1-1. Normal male anatomy, sagittal view (relaxation).

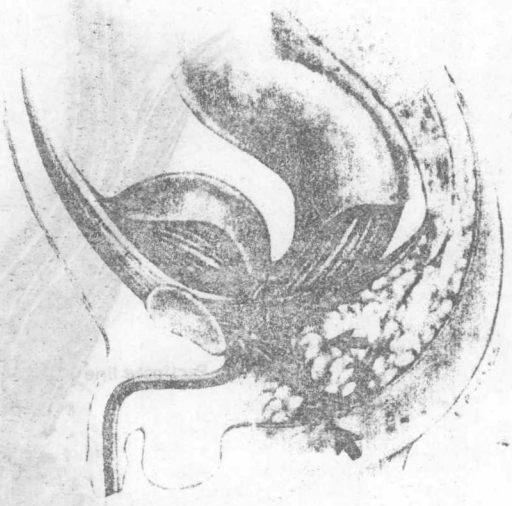

FIGURE 1-2. Male anatomy (contraction).

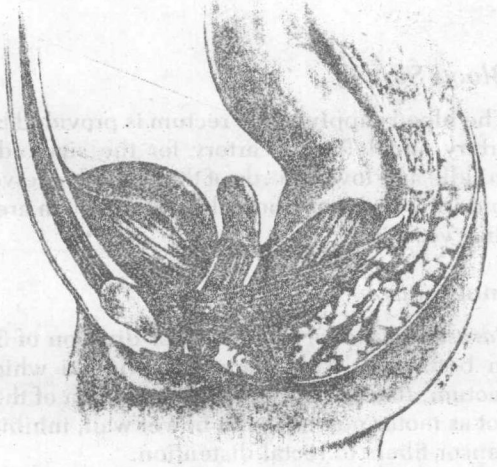

FIGURE 1-3. Female anatomy, sagittal view.

Smooth Muscle (Internal Sphincter)

The internal sphincter in a normal individual is represented by a thickening of the circular layer of the smooth involuntary muscle of the bowel, located in the anorectal area. The precise limits of this structure at different ages has not been accurately determined. (Figures 1-4 and 1-5).

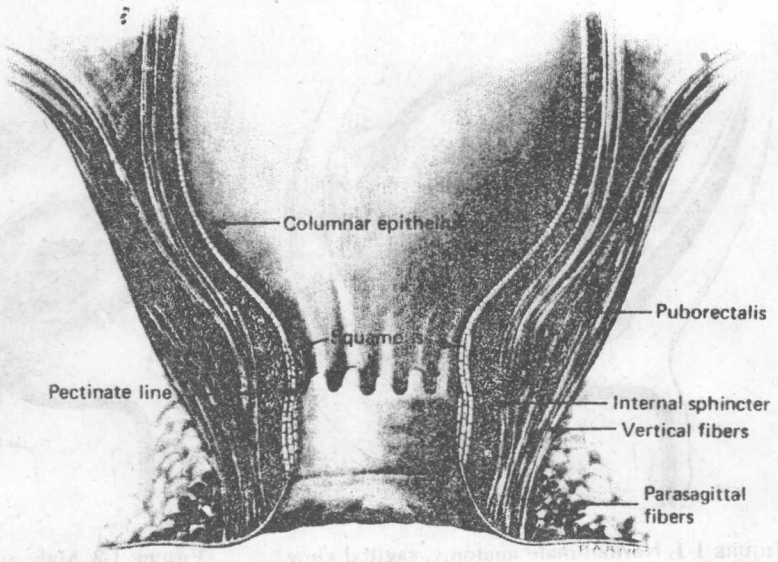

FIGURE 1-4. Detailed normal anatomy, coronal view.

Blood Supply

The blood supply of the rectum is provided by the inferior mesenteric artery and left colic artery for the sigmoid and upper rectum; the middle and lower thirds of the rectum receive blood from the superior and inferior hemorrhoidal vessels, which are branches of the internal iliac vessels.

Innervation

Parasympathetic. The anterior division of 3rd and 4th sacral nerves in both sides form the nervi erigenti which give branches to the rectum; there they relay in the ganglia of the Auerbach plexus. They act as motor nerves to the bowel wall, inhibitors to the sphincter, and sensor fibers to rectal distention.

Sympathetic. These nerves originate in the 2nd, 3rd, and 4th lumbar ganglia and the preaortic plexus. They form the hypogastric plexus at the level of the 5th lumbar vertebra; from there, they descend through the posterolateral pelvic walls as the presacral nerves, which join the pelvic ganglion on either side of the pelvis. They act as inhibitors of the bowel wall and motor nerves to the internal sphincter.

Somatic Innervation to the Voluntary Muscles. Most of the levator muscle, particularly its upper portion (ischiococcygeus) and the most anterior part (including the vertical fibers that we call muscle complex), usually known as pubococcygeus portions, receives innervation from the anterior roots of the 3rd and 4th sacral nerves.

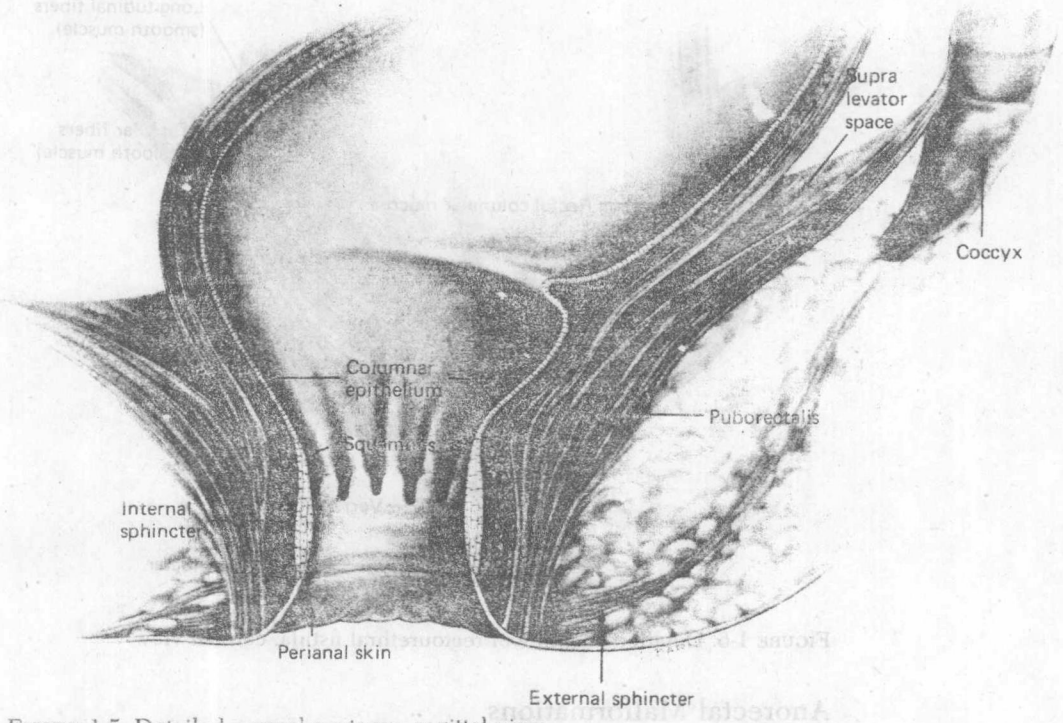

FIGURE 1-5. Detailed normal anatomy, sagittal view.

These roots unite to form a trunk that runs on the upper surface of the levator muscle. The pudendal nerve, which arises from the 2nd, 3rd, and 4th sacral nerves, also innervates these muscles. The lower portion of the levator muscle, known as puborectalis, as well as the external sphincter, form an inseparable structure that receives its innervation by the perineal branch of the 4th sacral nerve and also from the inferior hemorrhoidal and perineal branches of the pudendal nerves.

Sensory Innervation. The anal canal, including a zone about 1 cm above the pectinate line and extending down to the skin, is exquisitely sensitive, as demonstrated by Duthie and Gairns [9]; they described nerve endings capable of detecting pain (free intraepithelial), touch (Meissner's corpuscles), cold (Krause's end-bulbs), pressure or tension (corpuscles of Pacini and Golgi-Mazzoni), and friction (genital corpuscles).

The rectum (above anal canal) is not sensitive to the stimuli mentioned above; there is, however, some sensation to rectal distension which is thought to be provided by the parasympathetic innervation of the smooth muscle and by proprioceptive receptors located in the voluntary muscle mechanism surrounding the rectum.

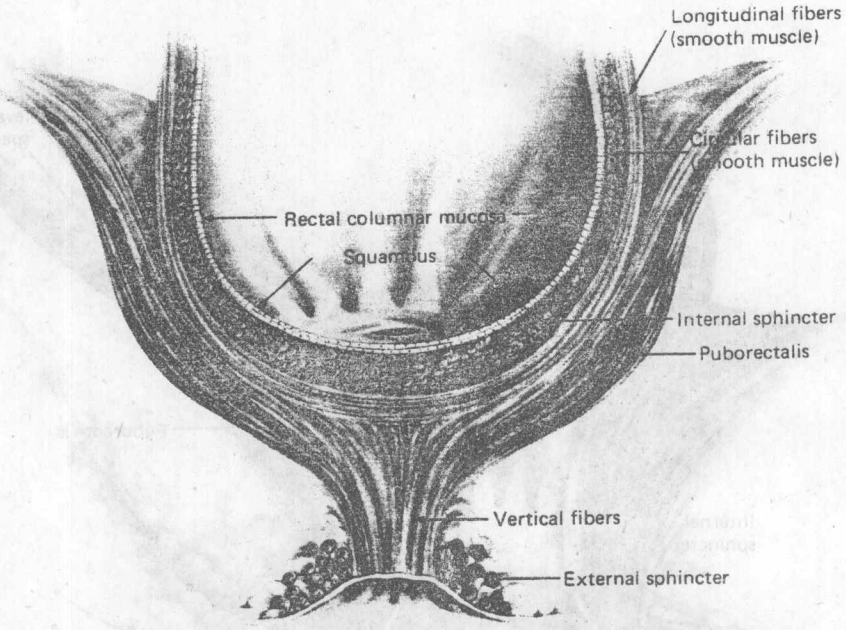

FIGURE 1-6. Detailed anatomy of rectourethral fistula, coronal view.

Anorectal Malformations

We are dealing with a spectrum of defects, therefore, each malformation has different anatomical characteristics. In the corresponding chapters, the reader will be able to see a diagram of the anatomy of each defect. However, there are certain general characteristics in these defects that will be discussed here briefly.

The normal descent of the rectum has been halted, therefore, the parasagittal fibers get closer together in the midline (Figure 1-6). Below the lowest part of the rectum, all the striated muscle fibers meet together forming a solid mass of muscle which is very thin in its lateral aspect. The length of the solid mass of muscle depends upon the height of the defect. The lower the defect, the shorter that solid mass and the wider the funnel-like muscle structure. Figures 1-6 and 1-7 show a view of a detailed anatomy of a male with a rectourethral fistula. Consistent with the idea of a spectrum of defects, the degree of muscle development varies from case to case. In one extreme of the spectrum, low defects can be seen which are associated with almost normal muscles. At the opposite extreme of the spectrum, there are very high defects with severe degrees of muscle underdevelopment. Very high defects are more frequently associated with abnormal sacrums and poor muscle development (Figures 3-1 and 3-2).

There is some evidence [10] that the internal sphincter is present in patients with anorectal malformations regardless of the height of the defect. That involuntary sphincter seems to be located in the most distal part of the rectum (Figures 1-6 and 1-7).

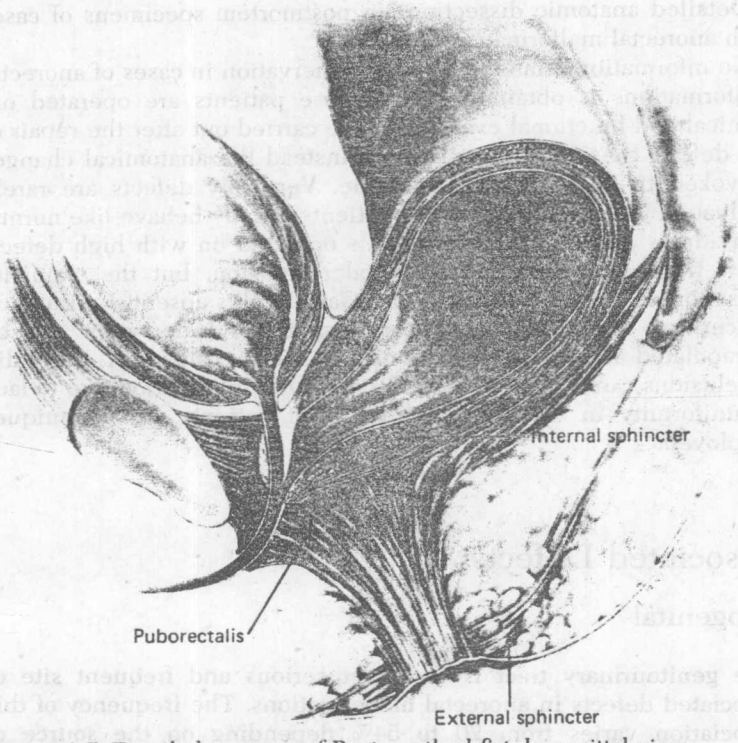

FIGURE 1-7. Detailed anatomy of Rectourethral fistula, sagittal view.

Very little is known about the blood supply of the rectum and pelvis in cases of anorectal malformations. Observations made during the surgical procedures confirm the impression that we are dealing with a spectrum of abnormalities; thus, very low types of defects are seen to have good muscles, blood supply, and innervation, indistinguishable from normal individuals. At the other end of the spectrum, extremely high defects (rectobladder neck or high vaginal fistulae) can be seen where hemorrhoidal vessels cannot be identified because the bowel is located very high above the pelvis.

Something similar must occur with the innervation of the rectum in cases of anorectal malformations, although this has not been documented. Bladder innervation as well as nervi erigenti in cases of high anorectal malformations [11] seem to run closer to the midline than normal, becoming more susceptible to surgical damage when violating the basic principle of staying in the midline.

There seems to be a direct correlation between the degree of sacral abnormality and the neurologic deficit of the pelvic organs, and the degree of muscle atrophy. Very high defects are more frequently associated with an abnormal sacrum and poor innervation.

Detailed anatomic dissections in postmortem specimens of cases with anorectal malformations are scanty.

No information related to sensory innervation in cases of anorectal malformations is obtained before these patients are operated on. Clinical and functional evaluations are carried out after the repair of the defect; thus, we are evaluating instead the anatomical changes provoked by the surgical technique. Very low defects are rarely evaluated, mainly because these patients usually behave like normal individuals postoperatively. Patients operated on with high defects show intrarectal sensation to rectal distension, but the exquisite sensation described by Duthie and Gairns [9] is absent. Information concerning sympathetic and parasympathetic innervation can be extrapolated from postop manometric studies; however, no valid conclusions can be drawn from the available information due to lack of uniformity in the selected groups of patients and techniques employed.

Associated Defects

Urogenital

The genitourinary tract is the most serious and frequent site of associated defects in anorectal malformations. The frequency of this association varies from 20 to 54% depending on the source of reference [12–17]. Those centers that follow a more strict protocol of urologic evaluation in patients with anorectal malformations usually detect a higher incidence of these defects. It is also true that the higher the malformation, the more frequently it is associated with a severe urologic problem. Forty-eight percent of our patients had associated genitourinary anomalies [18]. It is important to mention that our series is not representative of what happens in most places because we receive many referrals of the more complex malformations which have more chances of being associated with a urologic defect. Patients with a persistent cloaca or a rectovesical fistula demonstrated almost a 90% incidence of associated genitourinary abnormalities, whereas low defects had a frequency of association of less than 14% [18].

Sacrum and Spine

The sacrum is frequently abnormal. Sacral vertebrae may be deformed or reduced in number. It is well known that the absence of more than three sacral vertebrae is associated with a severe neurogenic deficit, including neurogenic bladder and lack of bowel control [19]. Other defects such as hemisacrum, may also lead to important neurogenic deficits. The upper spine frequently shows hemivertebra.