Anorectal Malformations

24

Tommaso Gargano and Mario Lima

24.1 Introduction

Anorectal malformations (ARMs) are rare birth defects of the digestive system affecting 2–6 per 10.000 births worldwide with an estimated prevalence rate of 3 per 10.000 births in Europe. They are more common among Asians and are somewhat more common in boys (60 %) than in girls. Male patients tend to have more severe malformations than female ones [1]. ARMs are the result of an abnormal development of the distal end of the digestive tract interesting the anus and/or rectum that occur early between the sixth and tenth week of embryonic development. They carry a malformation spectrum of severity depending on the level of disruption of the anorectal canal and of the associated caudal malformations (sacrum and spine). In most ARMs, the anus is not perforated, and the distal enteric component may end blindly (atresia) (Fig. 24.1) or as a fistula into the urinary tract, genital tract, or perineum (Figs. 24.2 and 24.3) [2].

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24.2 History

ARM or imperforate anus has been a well-known condition since antiquity. For many centuries, physicians created an orifice in the perineum of children with imperforate anus. Those that survived most likely suffered from a type of defect that would now be recognized as "low." Those with a "high" defect did not survive that treatment. Amussat, in 1835, was the first individual who sutured the rectal wall to the skin edges, which could be considered the first anoplasty. During the first 60 years of the twentieth century, surgeons performed a perineal operation without a colostomy for the so-called low malformations. High imperforate anus was usually treated with a colostomy performed in the newborn period, followed by an abdominoperineal pull-through some time later in life, but surgeons lacked objective anatomic guidelines. Unfortunately this left many patients incontinent and was not an appropriate solution to the spectrum of malformations. The surgical approach to repairing these defects changed dramatically in 1980 with the introduction of the posterior sagittal approach, which allowed surgeons to view the anatomy of these defects clearly, to repair them under direct vision, and to learn about the complex anatomic arrangement of the junction of the rectum and genitourinary tract. It has become the predominant surgical method for anorectal anomalies. In cases when the rectum or the vagina is very high and an

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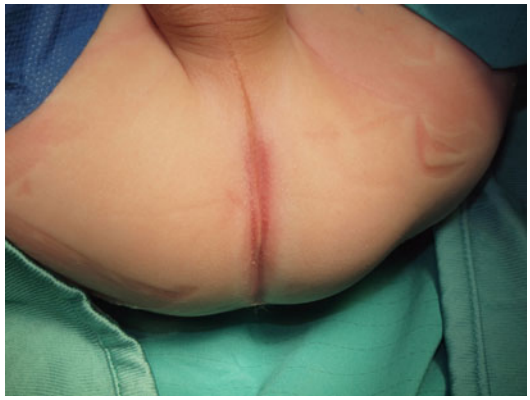

Fig. 24.1 Imperforate anus

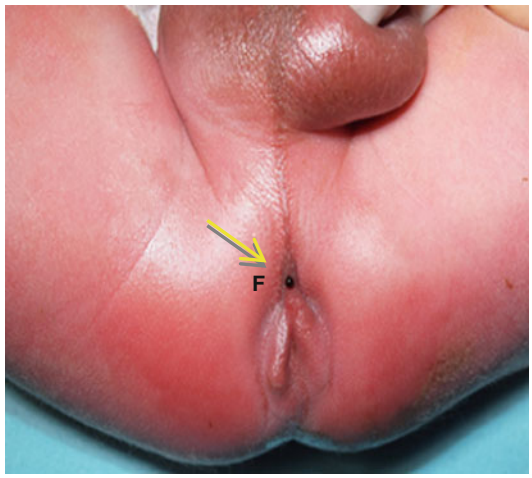

Fig. 24.2 Perineal (cutaneous) fistula in male

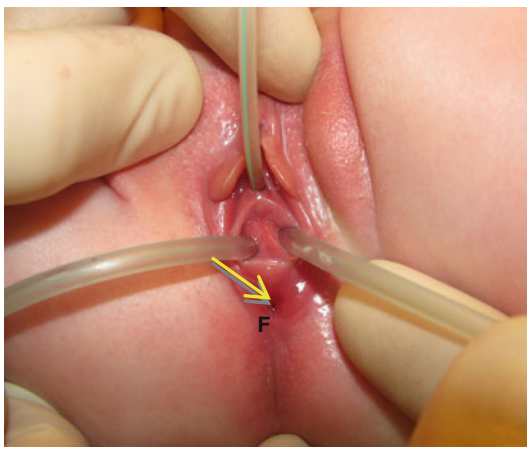

Fig. 24.3 Perineal (cutaneous) fistula in female

abdominal approach as well is needed, laparoscopy can be used in combination with the posterior sagittal approach [3].

24.3 Embryology

The early embryologic development of the anorectum, the primitive urogenital sinus, and the caudal neural tube is closely related, which helps explain the associated malformations of these three systems. In early embryonic life, the distal portion of the hindgut, the primitive cloaca, is divided into dorsal and ventral parts by a coronal sheet of the mesenchyme, the urorectal septum, and separated from the amniotic cavity by the cloacal membrane. Most ARMs result from abnormal development of the urorectal septum. Between weeks 4 and 6 of gestation, both the yolk sac or primitive hindgut and the allantois or primitive urogenital sinus enter into the cloaca. The urorectal septum then develops forklike infoldings (Tourneux and Rathke folds) of the lateral cloacal walls; at the same time, the embryo starts to curve as a result of the longitudinal growth of the developing neural tube and the mesodermal compartment. With these morphologic changes, the distance between the cloacal membrane and the tip of the urorectal septum is progressively reduced. At the end of week 7, the urorectal septum and the cloacal membrane are located at the same level. The cloaca is thus divided into a ventral part (the urogenital sinus) and a dorsal part (the rectum and proximal anal canal). Between them, the tip of the urorectal septum becomes the perineal area. At this time, the cloacal membrane ruptures by apoptosis, thus opening two orifices in the perineum: one ventral or urogenital and one dorsal or anal. Also at the end of week 7, a secondary occlusion of the anorectal canal takes place, initially by adhesion of the walls and later by formation of an epithelial "plug" at the anal level. This secondary closed anal orifice will rupture and recanalize by apoptosis at the end of week 8. Embryologically, ARMs can thus be subdivided into two main groups according to when the disturbances occur: those manifesting as an ectopic anal orifice or

fistula are due to early abnormal development of the dorsal part of the cloaca and the cloacal membrane (at weeks 4–7), whereas those manifesting as an abnormal anus in a normal position are due to later defective recanalization of the secondary occluded anal orifice (at weeks 7 and 8) [4].

24.4 General Considerations

ARMs are found as isolated congenital birth defects, as part of a syndrome or associated with other anomalies. Associated anomalies have been reported to occur in approximately 45–65 % of the patients, mostly of the urogenital tract, central nervous system, skeletal system (vertebrae), or the remaining gastrointestinal tract [1]. ARM has been reported to occur in families suggesting that there is a genetic component in its etiology. There appears to be a low rate of association in families, but some appear to have an autosomal dominant inheritance pattern. Consanguinity has been identified as leading to a higher incidence of ARM, particularly in countries in the Gulf and Middle East regions. In addition, familial Currarino associations are well established, and family members have been shown to have sacral anomalies without the full syndrome. In a small number of patients, genetic factors are clearly associated with ARM. Previous studies have suggested the importance of a locus on chromosome 7q39, which includes three genes: SHH, EN2, and HLXB9. These include Towne-Brock syndrome, FG syndrome, Kaufman-McKusick syndrome, and Lowe syndrome. In addition, ARM has been described in association with trisomy 8 mosaicism, as well as Down and fragile X syndromes. Till date, the accurate embryologic defect causing anorectal malformations still remains undetermined. With recent researches in the pathogenesis of anorectal malformations, the previous theories have been discarded. While in the past, defects in lateral fusion were thought to be causative, there is evidence from animal models and from detailed study of human fetuses with major anomalies that a deficiency in the dorsal component of the cloacal membrane and the adjacent dorsal cloaca is causative. A subsequent malfunction of the primitive streak and tail bud in the early development phase around 3–4 weeks has been proposed (yet to be clearly defined) as causation for associated anomalies of the pelvic floor [1, 5].

24.5 Classification

Based on the anatomy, various classifications have been proposed to define the pathology of these anorectal anomalies. The earliest classification dates back to 1953 when Gross proposed a simple differentiation based on the levator muscle, i.e., supralevator, for those above the levator ani, or infralevator anomalies, for those below the levator ani [6].

With advancement in the understanding of the pathology of the malformations, a need was felt to define these lesions more appropriately. During the centenary of the Royal Children's Hospital in Melbourne, a new international classification was proposed in 1970. This classification utilized the concept of levator ani wherein anomalies above the levator were termed as high and those below were termed as low anomalies, but it also introduced intermediate anomalies which were known as translevator anomalies [7].

The best known classification of ARMs is the Wingspread classification of 1984 (Wisconsin). This classification distinguished between high, intermediate, and low anomalies in the male and female, with special groups established for cloacal and rare malformations. High-type anorectal malformations were agenesis without fistula in both sexes. The low-type malformations were classified as anovestibular fistula in the female and, in both sexes, as anocutaneous fistula and anal stenosis. This classification was widely accepted over the years and was based on detailed embryological and anatomic studies performed especially by Stephens et al. and Kelly on anatomic sections and radiographic investigations. They recognized that the pubococcygeal line extending from the upper border of the os pubis to the os coccyx corresponds with the attachment of levator ani muscles to the pelvic wall, separating high-type malformations lying above the

levator muscle and intermediate and low forms of anorectal agenesis lying below this anatomic line. Furthermore, in healthy individuals, the lowest point of the ischial tuberosity, the so-called I-point, represents the deepest point of the funnel of the levator ani muscles. Therefore, every blind rectal pouch, lying between the puborectal line and the I-point, was classified as an intermediate anomaly and could be treated by a posterior sagittal anorectoplasty (PSARP). Low lesions below the I-point could be easily managed from a perineal approach. Because of these anatomic relations, the Wingspread classification had a significant impact on the choice of surgical approach. However, some details of the Wingspread classification remained questionable. Therefore, in 1995, Peña proposed a classification based on the type of the fistula present. He distinguished between perineal, vestibular, bulbar, prostatic, and bladder neck fistulas, imperforate anus without fistula, vaginal fistulas, cloacal fistulas, and rectal atresia or stenosis (Table 24.1).

This descriptive and fistula-related grouping became widely accepted over the past decade. The advantage of the classification of Peña is that the type of the fistula provides information not only about localization of the blind pouch but also on the anticipated extent of mobilization of the atretic rectal segment necessary to perform a sacro- or abdominosacroperineal pull-through. It is important to remember that the course of the fistula may vary from one individual to another and can be ascending or descending and of shorter or longer length so that the confluence of the fistula with the urogenital tract or perineum may differ from the lowest point of the blind pouch. This is especially true if the fistula arises from a higher level of the blind-ending rectum and not from its lowermost point. Therefore, the classification of Peña does not distinguish between rectovestibular and anovestibular fistulas. By closely comparing both classifications, that is, the Wingspread classification and the suggestions of Peña, it becomes clear that there is no real contradiction between them. Perineal and vestibular fistulas could be regarded as low malformations, bulbar fistulas, and imperforate anus without a fistula, and most of the vaginal fistulas may be regarded as intermediate-type anomalies, and prostatic and bladder neck fistulas are considered high-type imperforate anus. The same is true for rectal agenesis or stenosis. In addition, rare/regional variants, despite being frequent in certain geographic areas of the world, are not alluded to in either classifications. More recently, the Krickenbeck Conference of 2005 established a new classification, which is based mainly on the presence or absence of fistulas and their type and location, as well as the position of the rectal pouch. It has gained overall popularity in the international community of pediatric surgeons. This classification itself seemed a logical sequel to the Wingspread classification. It distinguishes five types of fistulas: rectoperineal, rectovestibular, rectourethral bulbar, rectourethral prostatic, and rectovesical. Cloacal malformations and the absence of fistulas, anal stenosis, and rare regional variants complete this classification. The extremely rare rectovaginal fistula is considered a variant of cloacal anomaly (Table 24.2) [4, 8].

Table 24.1 Peña classification

Males	Females
Perineal fistula (cutaneous)	Perineal fistula (cutaneous)
Rectourethral fistula	Vestibular fistula
Prostatic	Cloaca
Bulbar	Imperforate anus without fistula
Rectovesical fistula	Rectal atresia
Imperforate anus without fistula	
Rectal atresia	

Table 24.2 Krickenbeck classification

Major clinical groups	Rare/regional variants
Perineal (cutaneous) fistula	Pouch colon
Rectourethral fistula	Rectal atresia/stenosis
Prostatic	Rectovaginal fistula
Bulbar	H fistula
Rectovesical fistula	Others
Vestibular fistula	
Cloaca	
No fistula	
Anal stenosis	

Cloacal anomaly is a complex anatomic disorder that manifests as a unique external perineal opening with a short or long common canal for the genital, urinary, and digestive systems. Isolated rectovaginal fistulas are extremely rare and are considered a variant of cloacal anomaly. The Wingspread and Krickenbeck classifications are very similar. The Wingspread classification allows location of the blind rectal pouch. The Krickenbeck classification is more descriptive and is clinically oriented; its most important advantage is the preoperative identification and anatomic evaluation of not only the rectal pouch but also any fistulas. This information allows the surgeon to anticipate the extent of mobilization of the atretic rectal segment required during surgery and helps determine the most appropriate surgical approach for each case (Tables 24.3 and 24.4) [8].

24.6 Prenatal and Neonatal Management

Prenatal diagnosis of ARM remains rare and occurs in only up to 16 % of cases. Currently, the most complex anorectal malformations are the ones that can be most often diagnosed prenatally. The reason for this is the fact that the higher the malformation (recto-bladder neck fistula in males, cloaca in females), the higher is the presence of associated anomalies, and many of these associated defects can be seen in utero. During the prenatal imaging study, one important clue to suspect an anorectal malformation is the finding of multiple systems with abnormalities (digestive, vertebral, genitourinary). The advantages of having a prenatal diagnosis include giving the parents some information about the type of anomaly that the patient will be born with and also giving them the opportunity to make arrangements for the

Table 24.3 Comparison of Wingspread and Krickenbeck classifications in *male patients*

Type of ARM	Wingspread classification (1984)	Krickenbeck classification (2005)
Low	Anal stenosis Anocutaneous fistula	Anal stenosis Imperforate anus without fistula Rectoperineal fistula
Intermediate	Anal agenesis without fistula Anal agenesis with rectourethral bulbar fistula	Anal or anorectal agenesis without fistula Anorectal agenesis with rectourethral bulbar fistula Anorectal agenesis with rectourethral prostatic fistula Anorectal agenesis with rectovesical fistula
High	Rectal atresia anorectal agenesis without fistula Anorectal agenesis with rectourethral prostatic fistula	
	Rare forms	

Table 24.4 Comparison of Wingspread and Krickenbeck Classifications in *female patients*

Type of ARM	Wingspread classification (1984)	Krickenbeck classification (2005)
Low	Anal stenosis Anal agenesis without fistula Anal agenesis with external fistula	Anal stenosis Imperforate anus without fistula Anal agenesis with rectoperineal fistula Anal agenesis with rectovestibular fistula
Intermediate	Anal agenesis without fistula Anal agenesis with rectovestibular fistula Anal agenesis with rectovaginal fistula	Anal or anorectal agenesis without fistula Rectal atresia Cloacal malformations with short (<3 cm) or long (>3 cm) common canal
High	Rectal atresia Anorectal agenesis without fistula Anorectal agenesis with rectovaginal fistula	
	Cloacal malformation Rare forms	

baby to be delivered in a specialized center that is familiar with the neonatal management of patients born with these conditions. Images that can be seen prenatally and should raise suspicions for an anorectal malformation include dilated and or calcified bowel, lack of meconium at the expected rectal level, hydronephrosis, absent kidney, neural tube defects, tethered cord, hydrocolpos, vertebral anomalies, absent radius, and omphalocele in the absence of bladder visualization [9]. During the first 24 h of life, it is important to rule out associated malformations that might be life-threatening. With an echocardiogram, the physician will rule out cardiac conditions, a nasogastric tube should be passed to rule out esophageal atresia, an abdominal x-ray should rule out duodenal atresia, a kidney ultrasound should rule out severe hydronephrosis, and a pelvic ultrasound in females born with a cloaca should rule out a hydrocolpos. A sacral x-ray in anteroposterior and lateral views will allow for the calculation of the sacral ratio, which is an important tool to predict the future prognosis for bowel control. A spinal ultrasound should be ordered to rule out tethered cord. Imaging plays a key role in evaluation of ARM. In the first days of life, clinical and imaging findings facilitate early classification of ARM and allow a decision about whether to perform an immediate colostomy. In children with intermediate and high types of ARM, preoperative pelvic MR imaging after the neonatal period allows accurate evaluation of the morphology and grade of development of the sphincteric muscle complex (Fig. 24.4). This information helps orient the medical and surgical teams as to the postoperative prognosis for continence. During the first 24 h, the surgeon will also have enough information to decide between a primary repair and a descending colostomy. This decision should take into consideration the experience of the surgeon and the condition of the baby. Common indications for a colostomy include flat perineum, meconium in the urine, distal gas on the invertogram taken after 24 h of life above the coccyx, and cloaca (Fig. 24.5). In ARM the ideal colostomy must be completely diverting, leaving enough distal bowel to allow for the future pull-through. Both stomas must be separated enough to accommodate a stoma bag that only covers the proximal stoma.

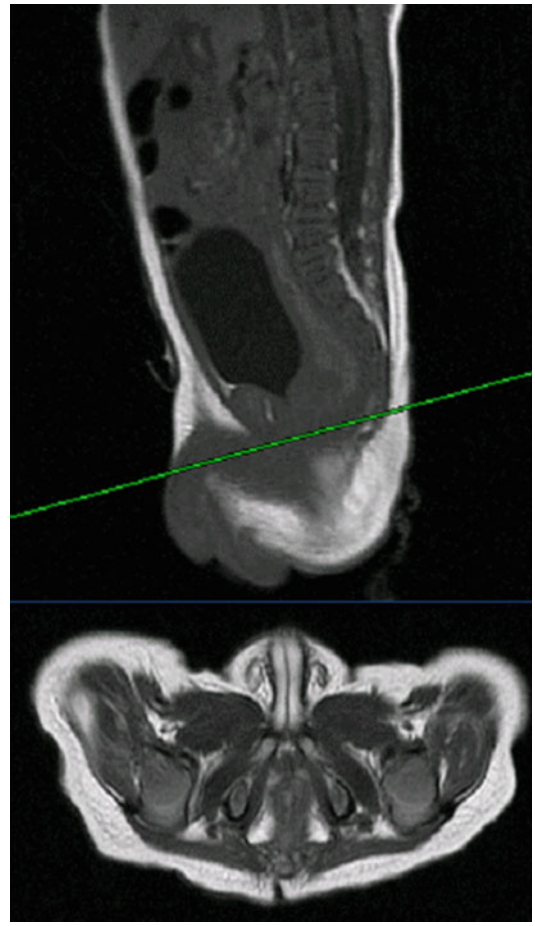

Fig. 24.4 Sphincteric muscle complex

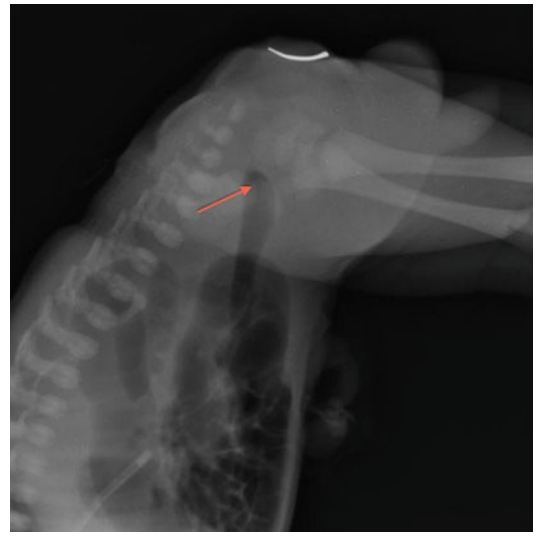

Fig. 24.5 Distal gas on the invertogram above the coccyx

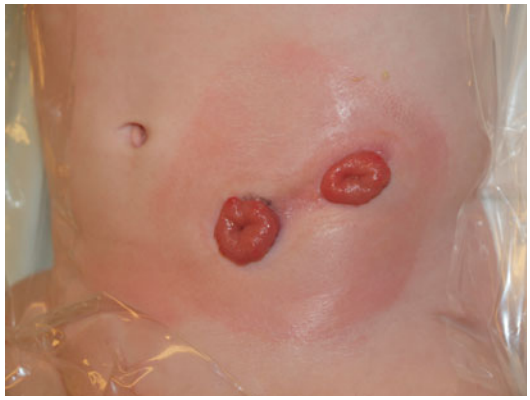

Fig. 24.6 Diverting colostomy in descendent colon

We suggest a descending colostomy taking advantages of the peritoneal attachments of the descending colon to avoid prolapse of the proximal stoma and making the mucous fistula as tiny as possible to avoid prolapse of the distal stoma (Fig. 24.6). During the colostomy opening, the distal bowel should be irrigated with large amounts of saline solution to clear it from any distal meconium. In patients with cloaca, during the first 24 h of life, a pelvic ultrasound should be ordered, specifically looking for a pelvic cystic mass behind the bladder. If a hydrocolpos is diagnosed, it should be drained at the time of colostomy opening with a transabdominal indwelling tube that should be left in place until the time of the main repair (when the patient will have a vaginal opening created). The distal colostogram is the most valuable diagnostic study to determine the specific type of anorectal malformation in male patients (the precise location of the fistula), the length of bowel available for the pull-through, and the relationship between the sacrum, the coccyx, and the rectum (Fig. 24.7). All these informations are important to plan the operation (laparotomy, laparoscopy, or posterior sagittal approach). In addition, it allows for the determination of the future functional prognosis [9].

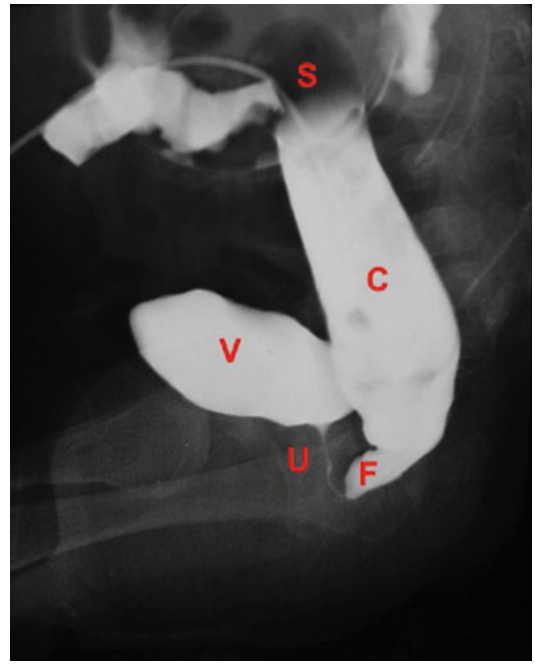

Fig. 24.7 Preoperative distal colostogram

24.7 Surgical Treatment

Almost all ARMs require surgery early in life. The spectrum of malformations sometimes mandates different techniques for different malformations, but the preferred technique is also influenced by the surgeon's preference and surgical education. The most commonly used operative procedures for treatment of ARMs include perineal operations, posterior sagittal anorectoplasty, and laparoscopic abdominoperineal rectoplasty techniques. Cloacal anomaly requires highly specialized reconstructive surgery [1, 2]. ARMs involving a rectal pouch located below the level of the puborectalis muscle, regardless of whether they are associated with a fistula perineal or vestibular, are considered low-type ARM. They may be managed early with a perineal approach involving opening of the rectal pouch and ligature of the fistula, if present. A rectal pouch lying at or above the level of the puborectal sling is considered an intermediate or high type of ARM; it is treated with colostomy in the first days of life and with posterior sagittal anorectoplasty alone or combined with laparoscopic abdominoperineal rectoplasty in a second intervention.

24.7.1 Posterior Sagittal Anorectoplasty (PSARP)

The patient is placed in a prone position with the pelvis elevated (Fig. 24.8).

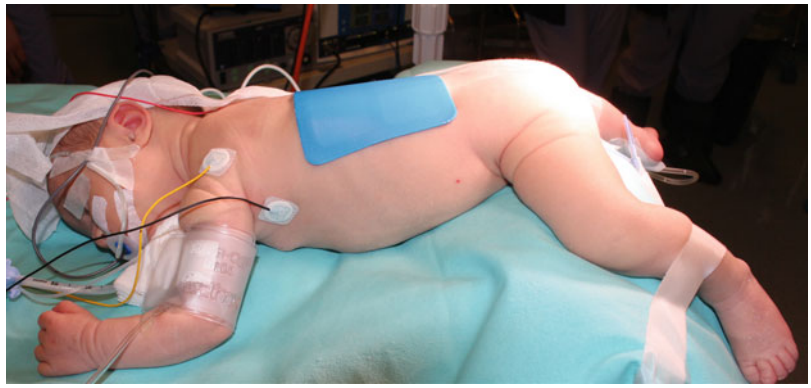

Fig. 24.8 Position of patient in PSARP procedure: prone position with the pelvis elevated

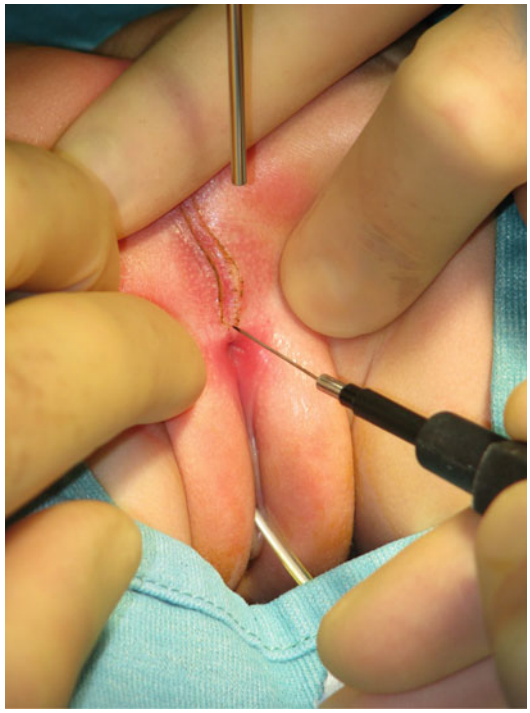

Fig. 24.9 Midline incision from the tip of the coccyx to the perineum

A strictly midline incision is then made from the tip of the coccyx to the perineum. Throughout the procedure, muscles are identified with the help of a muscle stimulator. All muscle groups are separated and opened as if paging through a book, without cutting them, until the rectal pouch is located. The levator ani muscle must then be divided to reach the rectal pouch (Figs. 24.9 and 24.10).

The rectum is then mobilized until a sufficient length is obtained for anal reconstruction.

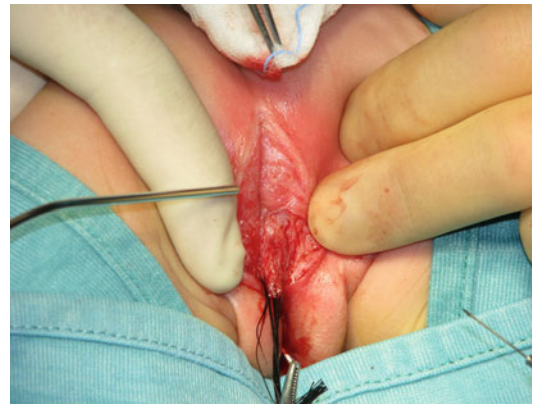

Fig. 24.10 Isolation of rectal pouch

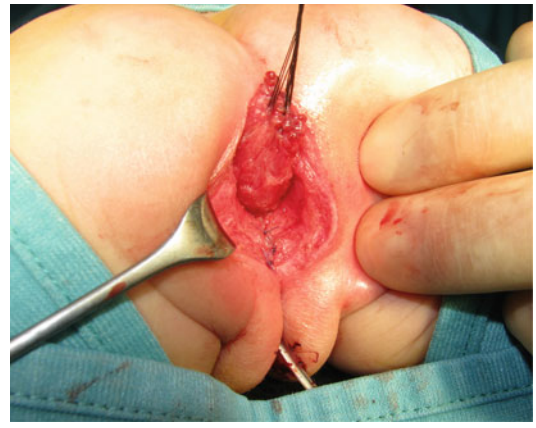

Fig. 24.11 Anterior suturing of the muscular plane

After that, the levator ani muscle is repaired, followed by repair of the muscle complex and external anal sphincter (Figs. 24.11, 24.12, 24.13, and 24.14).

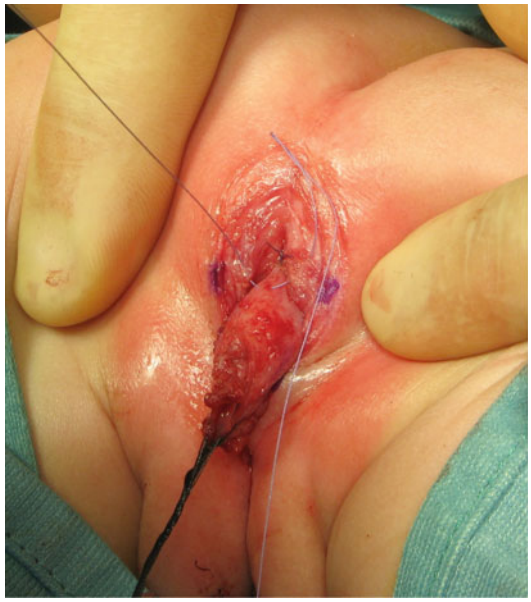

Fig. 24.12 Posterior suturing of the muscular plane

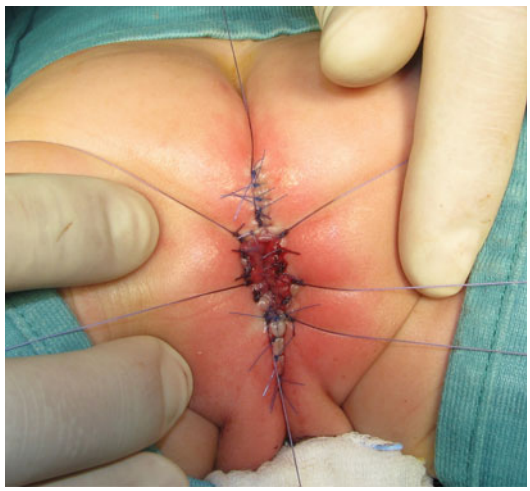

Fig. 24.13 Final step: anoplasty

Very high fistulas, mainly rectourethral prostatic or rectovesical fistulas in boys, are sometimes impossible to visualize exclusively through a perineal sagittal approach, and a laparotomy or laparoscopy (abdominoperineal rectoplasty) is also required. If an abdominal approach is needed, the patient is then positioned faceup, allowing the surgeon to work simultaneously from the abdomen and the perineum.

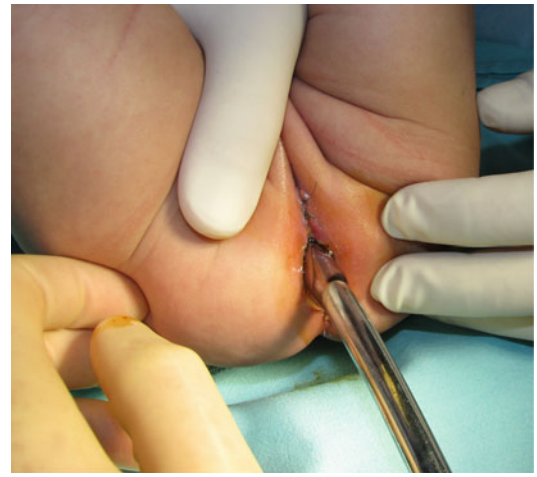

Fig. 24.14 Anal calibration

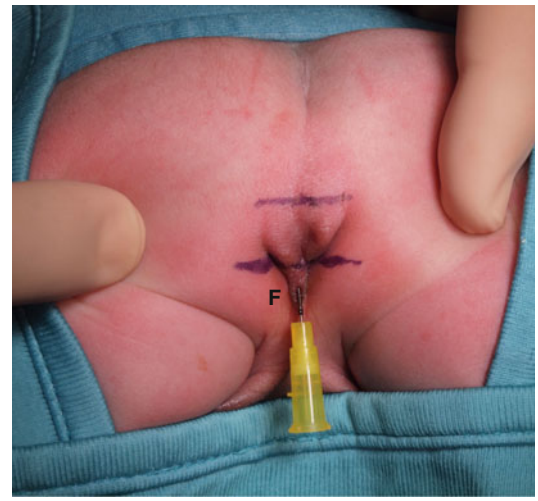

Fig. 24.15 Rectoperineal fistula in male. *F* fistula

24.7.2 Posterior Sagittal Anoplasty for Rectoperineal Fistula

The repair of these defects consists of a small posterior sagittal incision with enough mobilization of the rectum, sufficient to be transposed and placed within the limits of the sphincter (Figs. 24.15, 24.16, 24.17, and 24.18). This is a meticulous operation and can be done during the neonatal period without a colostomy. The most common complication during the repair of this defect in male patients is a urethral injury, which can be avoided by placing a urethral catheter and taking particular care during the dissection of the anterior rectal wall.

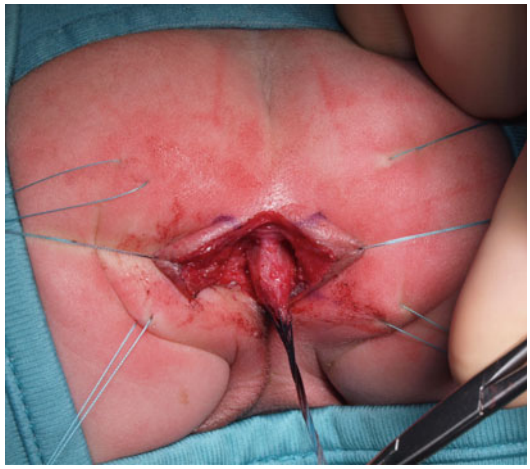

Fig. 24.16 Mobilization of distal pouch

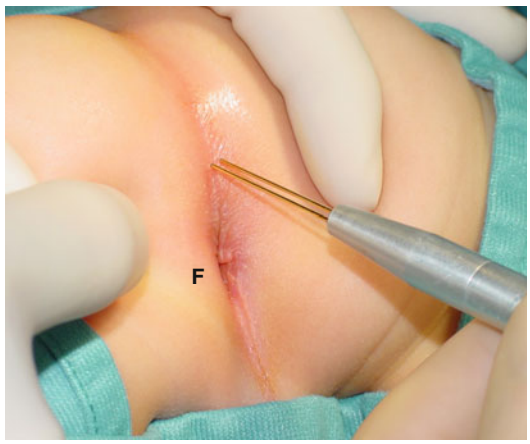

Fig. 24.17 Localization of neoanum with Peña electrostimulator

24.7.3 Posterior Sagittal Anorectoplasty for Rectovestibular Fistula, Rectourethral Bulbar Fistula, Rectourethral Prostatic Fistula, and Imperforate Anus Without Fistula

The key anatomic characteristics that should be kept in mind are that in rectovestibular fistulas the rectum shares a common wall with the vagina, and in rectourethral fistulas and imperforate anus without fistula, the rectum shares a common wall with the urethra. The surgeon has to make two

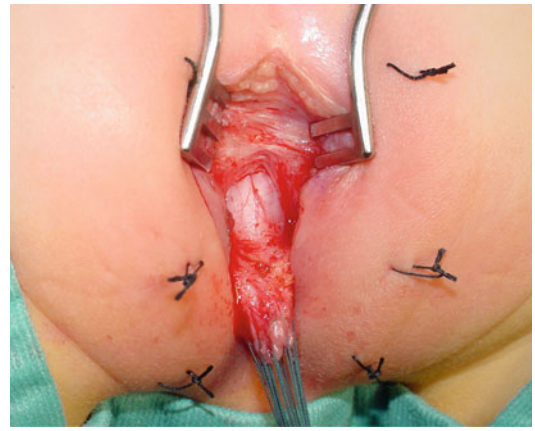

Fig. 24.18 Mobilization of distal pouch

walls out of one with a careful and meticulous separation of these structures. The posterior sagittal incision in these cases should be long enough to allow for adequate rectal mobilization. The posterior rectal wall should be identified, the lateral walls should be dissected, and then the surgeon should concentrate on the most delicate portion of the operation: the separation of the anterior rectal wall, without damaging the urethra in males and the vagina in females.

24.7.4 Laparoscopic-Assisted Posterior Sagittal Anorectoplasty (LAARP)

In 10 % of the male patients, the abdominal cavity has to be entered either through laparoscopy or laparotomy to repair the anorectal malformation. We consider the recto-bladder neck fistulas the ideal indication for laparoscopy as well as some selected rectourethral prostatic fistulas. In 2000, Georgeson proposed a new technique that combines the laparoscopic approach [10]. Yamataka et al. proposed and others confirmed the laparoscopic use of the Peña electrostimulator [11]. The LAARP technique allows treatment of high malformations by pulling down the rectum under direct vision close to the perineal plane. The levator muscles are clearly identified, thanks to intra-abdominal and external electrostimulation, so the surgeon can be sure of the

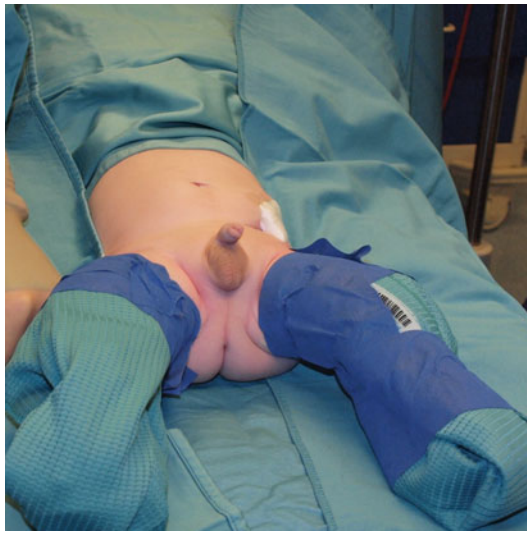

Fig. 24.19 Supine position in LAARP procedure

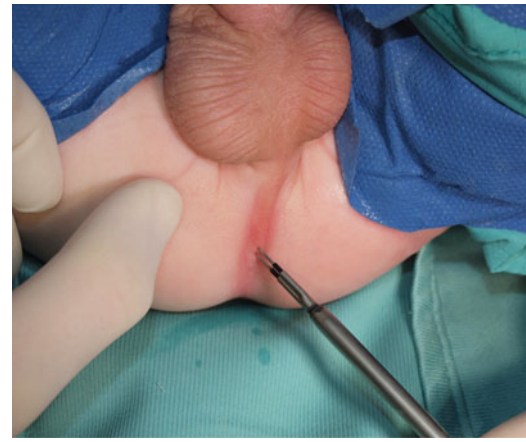

Fig. 24.20 Peña electrostimulation

correct position of the anus, thus avoiding the risks of sagittal dissection. Some studies seem to demonstrate better anorectal manometric findings in patients who underwent LAARP. Although the primary pull-through without colostomy has been described, we prefer to perform the LAARP after a diverting colostomy. We want to emphasize the importance of good positioning of the colostomy in order to avoid problems in mobilizing the rectum. Finally, we want to state the advantage of the intra-abdominal use of the Peña electrostimulator. The bellies of the puborectalis sling are clearly seen, and the contractions indicate the exact site of the pull-through. This can be particularly useful in cases of immature and unclear levator muscles [12] (Figs. 24.19–24.26).

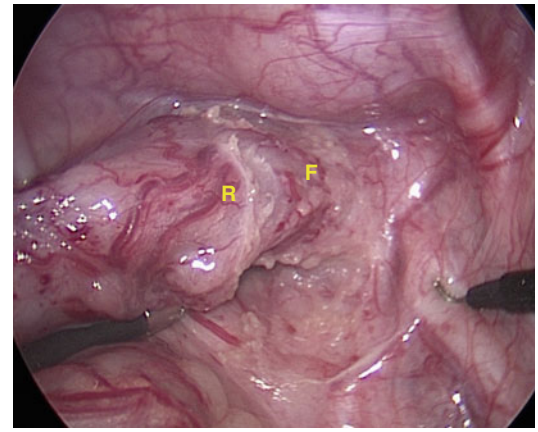

Fig. 24.21 Laparoscopic identification of fistula

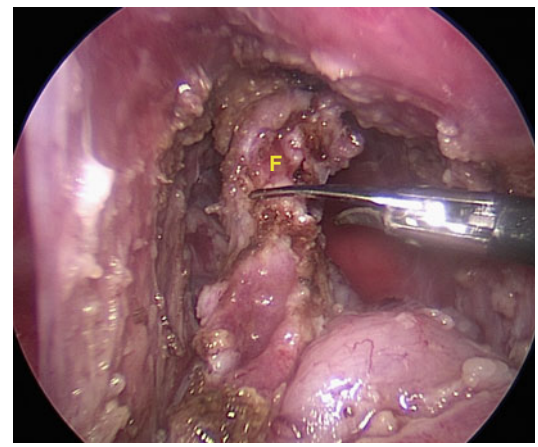

Fig. 24.22 Resection of fistula

24.7.5 Posterior Sagittal Anorectal–Vaginal–Urethral Plasty with Laparotomy for Cloaca with a Common Channel Length of More Than 3 cm

The repair of these complex defects requires the implementation of a rather complicated decision making algorithm. When the total urogenital mobilization (TUM) is not enough for the urethra

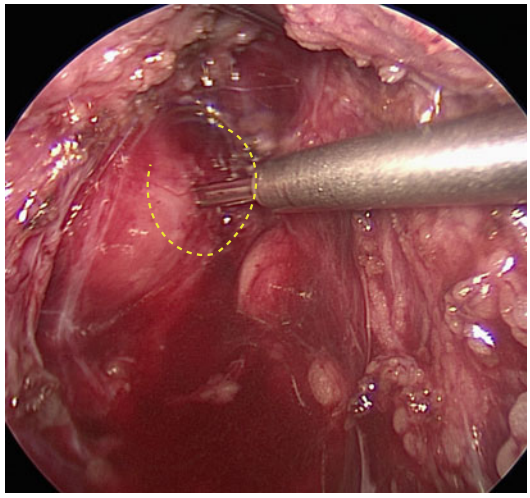

Fig. 24.23 Laparoscopic electrostimulation

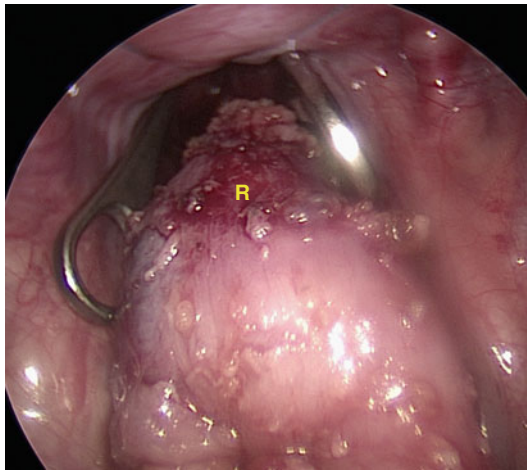

Fig. 24.24 Video-assisted rectal pull-through

and vagina to reach the perineum, carving the pubic cartilage and making a Heineke-Mikulicz maneuver in the vagina may give extra millimeters; when that is not enough, an extended transabdominal total urogenital maneuver is performed. If the structures still do not reach the perineum, the most challenging maneuver should be done, and it consists in the separation of the vagina from the bladder. To do that, the bladder must be open, and catheters have to be inserted into the ureters. At this stage, if the vagina still does not reach, depending on the anatomy, a vaginal switch maneuver can be performed or a partial vaginal replacement using the rectum or colon [6].

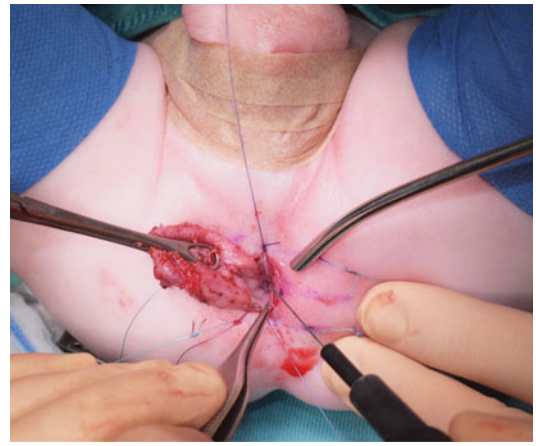

Fig. 24.25 Perineal approach: rectal pull-through

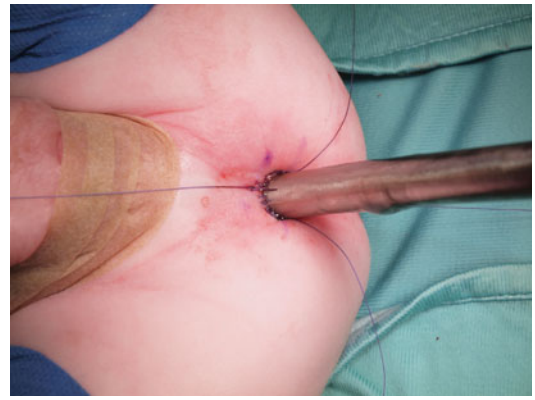

Fig. 24.26 Calibration of neoanum

24.8 Postoperative Management

At 2 weeks postsurgery, anal calibration is performed, followed by a program of anal dilatations to avoid an anoplasty stricture. The anus must be dilated twice daily, and the size of the dilator is increased every week. The final size to be reached depends on the patient's age (Table 24.5).

24.9 Treatment of the Functional Disorders (Constipation and Fecal Incontinence)

Constipation Constipation is the most important problem to avoid after definitive repair. Patients with good prognosis for bowel control

Table 24.5 Postoperative program of anal dilatations

Guidelines for sizing anal dilators	Suggested timing
1–4 months of age: 12	Dilate 2–3 times a day for 1–2 weeks Dilate once daily for 1–2 weeks Dilate once every other day for 1–2 weeks Dilate once every 3–4 days for 2 weeks Dilate once weekly for 4 weeks
4–8 months of age: 13	
8–12 months of age: 14	
1–3 years of age: 15	
3–12 years of age: 16	
>12 years of age: 17–18	

(rectoperineal fistula, rectovestibular fistula, rectourethral bulbar fistula, imperforate anus without fistula, with normal sacrum, and no tethered cord) are the ones that suffer from the most severe type of constipation. These patients usually require laxative dosages much higher than what is conventionally recommended. Patients must be regularly monitored, and laxatives and dietary manipulations are begun at the first sign of constipation. If surgical treatment to restore anatomy as normal as possible is indispensable, postoperative care is essential for these patients whose defecation mechanisms are altered, to reach if not continence, at least a socially acceptable cleanliness.

Fecal incontinence Patients with poor prognosis for bowel control (recto-bladder neck fistulas, cloaca with common channel more than 3 cm in length and tethered cord) should be kept artificially clean with a daily enema. Rectal administration of this daily enema allows the patient to be clean of stool in the underwear for a 24 h period, until the time for the next enema. Patients may complain of soiling. This may represent fecal incontinence in patients with very high ARMs or in those with poor muscles and an abnormal sacrum. These patients require a proper bowel management program. However, in a patient with a good prognosis, soiling may represent overflow incontinence, and constipation must be treated [2].

24.10 Complications

Iatrogenic complications include *dehiscence* and *infection*, which may be avoided with colostomy before the main repair. The *anoplasty stricture* is a possible postoperative complication that may be avoided by a program of dilatations. Posterior *urethral diverticulum* may develop from a fistula remnant.

References

1. De Blaauw I, Wijers CH, Schmiedeke E et al (2013) First results of a European multi-center registry of patients with anorectal malformations. J Pediatr Surg 48(12):2530–2535
2. Cretolle C, Rousseau V, Lottmann H et al (2013) Anorectal malformations. Arch Pediatr 20(Suppl 1):S19–S27
3. Levitt MA, Peña A (2012) Correction: anorectal malformations. Orphanet J Rare Dis 7:98
4. Alamo L, Meyrat BJ, Meuwly JY et al (2013) Anorectal malformations: finding the pathway out of the labyrinth. Radiographics 33(2):491–512
5. Iwai N, Fumino S (2013) Surgical treatment of anorectal malformations. Surg Today 43(9):955–962
6. Gross RE (1953) Malformations of the anus and rectum. The surgery of infancy and childhood. WB Saunders, Philadelphia, pp 348–357
7. Santulli TV, Kiesewetter WB, Bill AH Jr (1970) Anorectal anomalies: a suggested international classification. J Pediatr Surg 5:281–287
8. Holschneider A, Hutson J, Peña A et al (2005) Preliminary report on the International Conference for the Development of Standards for the Treatment of Anorectal Malformations. J Pediatr Surg 40(10): 1521–1526
9. Bischoff A, Levitt MA, Peña A (2013) Update on the management of anorectal malformations. Pediatr Surg Int 29(9):899–904
10. Georgeson KE, Inge TH, Albanese CT (2000) Laparoscopically assisted anorectal pull-through for high imperforate anus: a new technique. J Pediatr Surg 35:927–931
11. Yamataka A, Segawa O, Yoshid R et al (2001) Laparoscopic muscle electrostimulation during laparoscopy-assisted anorectal pull-through for high imperforate anus. J Pediatr Surg 36:1659–1661
12. Lima M, Tursini S, Ruggeri G et al (2006) Laparoscopically assisted anorectal pull-through for high imperforate anus: three years'experience. J Laparoendosc Adv Surg Tech 16:63–66