Recto-perineal Fistula

8

8.1 Definition, Frequency, and Prognosis

Perineal fistula is an anal malformation in which the anal opening is located anterior to the center of the sphincter. In females, the anal opening is located somewhere between the location of the normal sphincter and the female genitalia in the area known as the perineum or perineal body. The anterior mislocation of the orifice could be minimal (a few millimeters) (Fig. 8.1) or severe, becoming borderline with a malformation called vestibular fistula (Fig. 8.2). When the anal orifice is located at the junction of both labia majora, the malformation sometimes receives the French name "fourchette malformation" (Fig. 8.3), which is considered a defect intermediate between the vestibular and perineal area. Yet, most anorectal malformations in females can be clearly differentiated between perineal and vestibular. This is, perhaps, the anorectal defect subjected to more controversies, both in semantics and treatments. In female patients, some surgeons use different terms to refer to this condition, including "ectopic anus" and "anterior displacement of the anus" [1–13]. We prefer the term perineal fistula for the following reasons:

- The anal opening is most frequently strictured or stenotic.
- There is no anal canal.
- The orifice is not surrounded 360° by a sphincter mechanism.

These facts make us believe that this is not a real anus. For similar reasons, we believe that in order to call a malformation "anterior anus," it would be necessary for the patient to have a non-stenotic orifice, with normal anal canal, and surrounded 360° by a sphincter mechanism (as electrically demonstrated). If we accept this, as the definition of an anterior anus, then we must say that we have never seen that specific type of defect. The fact that we have not seen such defect does not mean that it does not exist, but it certainly means that it must be

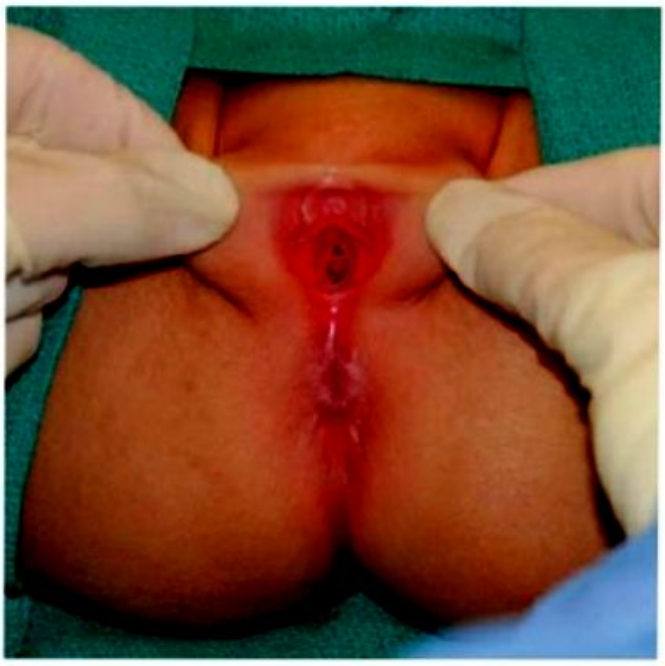

Fig. 8.1 Picture of a perineal fistula with minimal anterior mislocation of the opening

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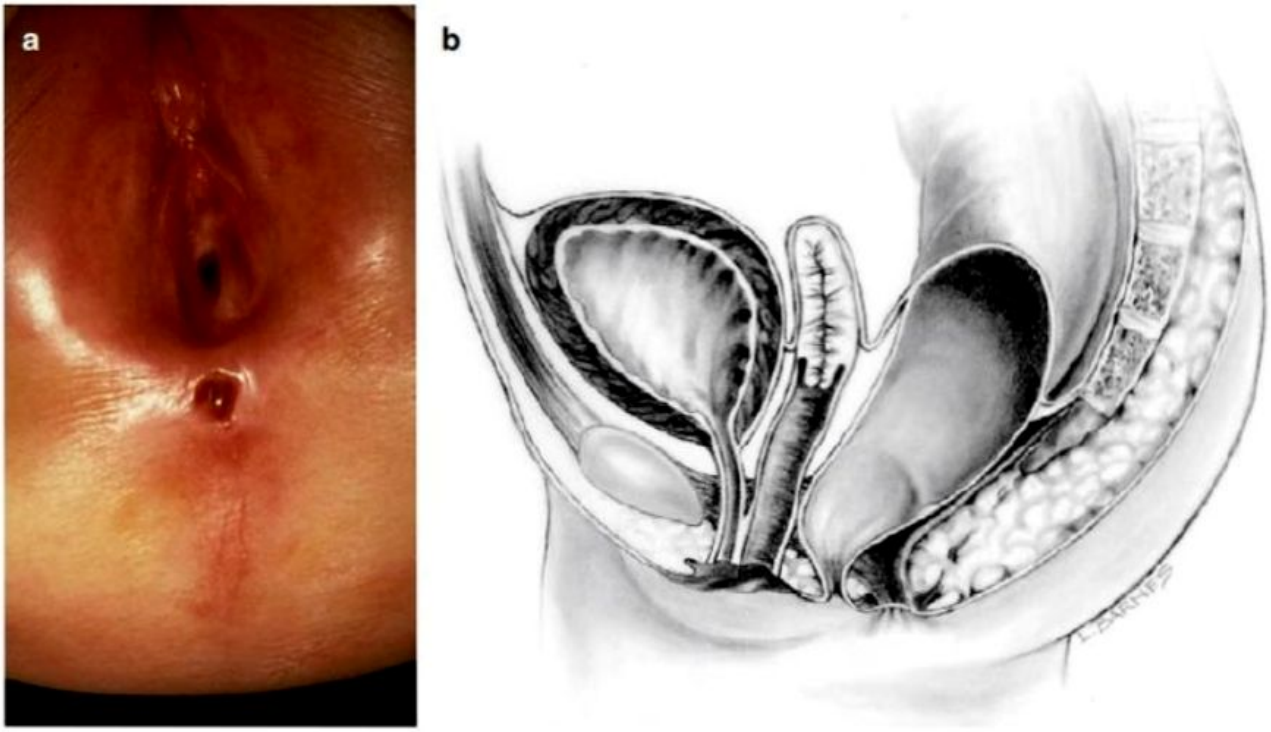


Fig. 8.2 Perineal fistula with a significant anterior mislocation of the anal opening. (a) Picture. (b) Diagram

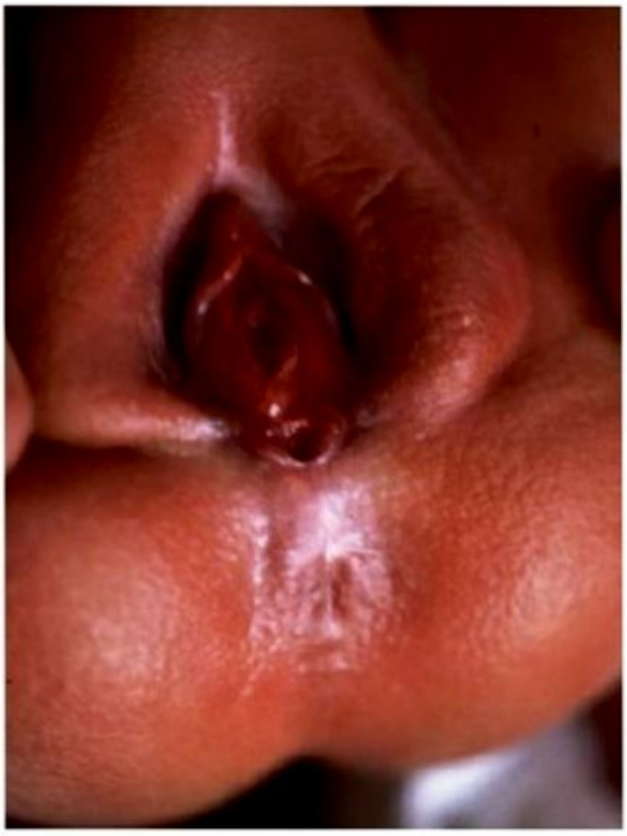


Fig. 8.3 "Fourchette" type of fistula

extremely rare. In addition, we believe that in the event of seeing a case suffering from such condition, we would be very hesitant to operate.

Perineal fistula is one of the most common anorectal defects. Our series includes 90 males and 84 females, occupying the second place in frequency in males after the rectourethral fistulas (bulbar and prostatic) together and the third place in females after vestibular fistula and cloacas. However, we have reasons to believe that the perineal fistula is perhaps one of the two most frequent defects in females. Most likely, the majority of perineal fistula cases are not referred to us, probably because they are considered easy to repair. Ours is a referral center, and we receive mainly complex and (or) complicated patients from other institutions. That may be the reason to explain the elevated number of cloacas that we have, as well as the rather low number of cases with perineal fistula. We conclude that the relative frequency of presentation of these malformations in our series is not representative of the frequency in the general population.

Perineal fistulas are the most benign of all anorectal defects in terms of functional prognosis for bowel control. In fact, in our experience, 100 % of the patients operated by us have bowel control provided the sacrum is normal. The majority of patients have a good sacrum, but there are exceptions, as we will be discussing below.

The chances for these patients to have bowel control are 100 % provided they receive a good operation. Ironically, the incidence of constipation in these patients is the highest of the entire spectrum of anorectal malformations!! In fact, we observed that the higher the malformation, the poorer the prognosis for bowel control, but the better the prognosis for constipation, or the other way around, the lower and more benign the malformation, the highest chances of bowel control, but also the highest possibilities of suffering severe constipation. The reason for this is unknown. Interestingly, higher malformations require much more perirectal dissection in order to bring the rectum down to the perineum. This dissection means that we divide the vessels and the nerves that surround the rectum in order to mobilize it down. In other words, higher rectums require more denervation, yet they have less constipation!! Patients with perineal fistulas require a minimal degree of mobilization and therefore denervation, and yet they have the worst degree of constipation.

Another important characteristic of this particular defect is that there is a group of patients with perineal fistulas that run in families. It is well documented in the literature that about 1 % of all patients with anorectal malformations have a sibling with an anorectal malformation [14]. In other words, traditionally, when the parents ask about the risk of having a second child with an anorectal malformation, the standard answer from geneticists, pediatricians, and us is 1 %. Yet, a deeper and more thoughtful analysis of the question reveals that, actually, we have never seen a family with two cloacas. In contrast, we have several families that have two or more siblings with perineal fistula frequently associated to a presacral mass and sacral defect. The familiar characteristic of this condition has been published many times [15–31]. Some other members of the family may have the sacral defect and the presacral mass and not the perineal fistula. For this reason, in a patient with perineal fistula, it is mandatory to document the integrity of the sacrum with an AP x-ray film of the sacrum to rule out a sacral defect because of the high incidence of sacral defects and presacral masses associated to perineal fistulas. Once we document a case of perineal fistula with a presacral mass, we have to screen the other members of the family for the same defect. This is extremely important because we have seen patients that were diagnosed as having a perineal fistula received an operation focused only in the anal defect, and the patients were left with a presacral mass and sometimes with very negative consequences. In addition, having diagnosed a sacral defect with a presacral mass changes completely the functional prognosis for these patients. This, we believe, it is extremely important to diagnose, to discuss with the parents, and to adjust their expectations in terms of future functional prognosis. In addition, as will be described later, this complex triad requires a specific therapeutic strategy.

The association of anorectal malformation, sacral defect, and presacral mass is well known [32–61] and is frequently called Currarino triad [38].

Unfortunately, there is a group of patients who were born with perineal fistula, as previously mentioned, a defect with an excellent functional prognosis, and yet, they end up suffering from fecal incontinence after several therapeutic misadventures and catastrophic events. Those are totally preventable problems consecutive to the lack of knowledge of the management of these patients.

8.2 Associated Defects

Following the same pattern already observed in the entire spectrum of anorectal malformations, perineal fistulas have the lowest incidence of urologic-associated defects or functional disorders. According to our database, 18 % of female patients and 27 % male patients with perineal fistulas suffer from urologic problems. Absent kidney occurs in 2–4 % of the cases, vesicoureteral reflux in 3–5 %, hypospadias in 1 %, ectopic ureters in less than 1 %, vaginal septum in 2 %, hyposadias in 1 %, undescended testicle in 2 %, bifid scrotum in 8 %, and hydronephrosis in 6 %.

Vertebral anomalies also are less common in these defects as compared to the others. Hemivertebra occurs in less than 2–4 % of the cases, and sacral defects are very unusual. The

sacra in these patients are usually normal except in those patients who are born with presacral mass and hemisacrum.

Patent ductus arteriosus occurs in less than 3 %, atrial septal defect in 2 %, ventricular septal defect in 1 %, tetralogy of Fallot in 1 %, and esophageal atresia in 1 %.

Tethered cord is present in 29 % of male patients and 13 % of female patients. This relatively high frequency of tethered cord is explained by the fact that patients with perineal fistula have a high frequency of association with sacral defects and presacral masses.

8.3 Diagnosis

8.3.1 Female Patients

The diagnosis of a perineal fistula in a female patient is a straightforward one. It is enough to see the perineum of the baby to understand that the anal orifice is abnormally located anterior to the center of the sphincter (Figs. 8.1 and 8.2). As previously mentioned, the orifice may be located very close to the female genitalia or very close to the center of the sphincter, and so the degree of mislocation varies from patient to patient. There is also a specific defect called by Stephens [62] "perineal groove" which is a strip of mucosa that runs between the female genitalia and the anus. This defect is frequently associated to perineal fistulas (Fig. 8.4). When this is left untouched, most of the times, that strip of mucosa suffers from metaplasia and changes into something that looks very much like skin. Yet, we have seen patients of 5–7 years old in which that mucosa continued producing some degree of wetness, which prompted us to operate. The operation of the perineal groove requires only removal of a very thin layer of mucosa and suturing together in the midline the skin of the perineal body. This operation is usually done in conjunction with the mobilization of the anal opening back to the center of the sphincter.

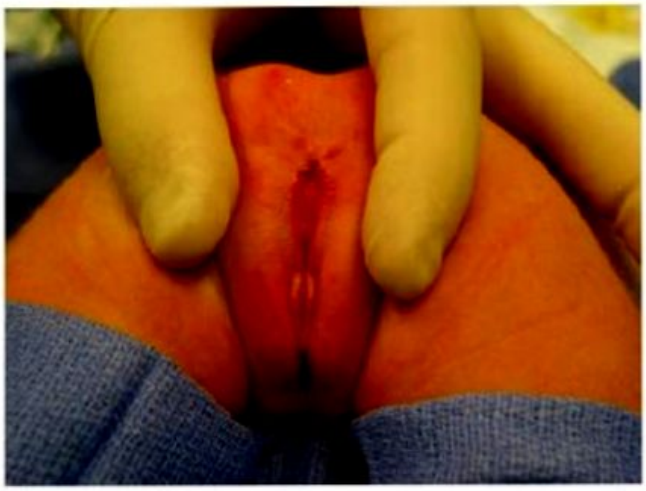


Fig. 8.4 Picture of a perineal groove

Sometimes, the anus looks like it is surrounded by a normal sphincter mechanism. Yet, with an electrical stimulator, under anesthesia, it is extremely easy to demonstrate that it is just an optical illusion. The bulk of the sphincter mechanism is located behind the anal opening and extends like a horseshoe on both sides of the anus, but there is no sphincter whatsoever between the female genitalia and the anal opening.

8.3.2 Male Patients

The diagnosis in the male patient may be a little bit trickier. The baby may have a very obvious orifice in the perineum through which one can see meconium coming out. In such case, the diagnosis is easy (Fig. 8.5). The fistula is always located anterior to the center of the sphincter. We have never seen a case in which the anal opening is located posterior to the center of the sphincter. The orifice, however, could vary in size from patient to patient being always stenotic. Sometimes there is a tiny, almost invisible orifice (Fig. 8.6). The fact that it is a small orifice usually delays the passing of meconium, which is another reason why we believe that in the management of the newborn baby with anorectal malformation, decisions concerning the opening of a colostomy or anorectal repair should not be taken before 24 h. In the first 24 h, it is very unlikely for the patient to pass meconium through a tiny orifice. When the pediatric surgeon is called to see a baby with imperforate anus, it is extremely important to do a meticulous inspection of the perineum. One should not hesitate to use

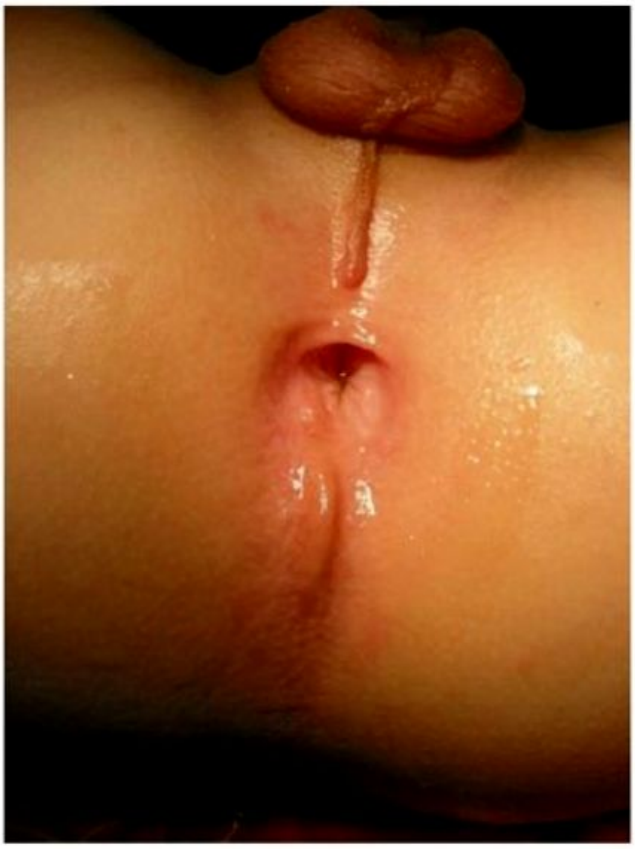


Fig. 8.5 Perineal fistula in a male. The anal orifice is very obvious

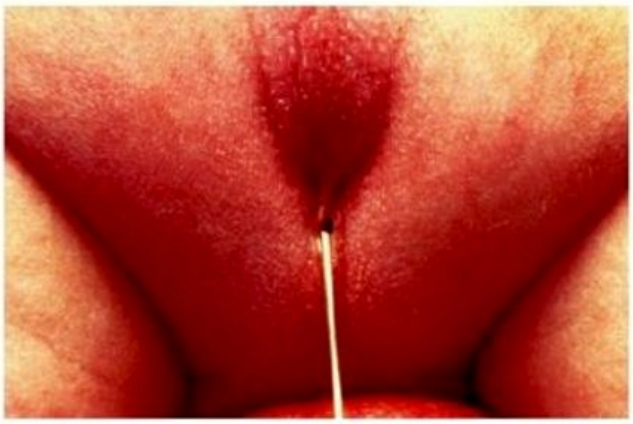


Fig. 8.6 Very small perineal fistula

magnifying glasses if necessary to look carefully for an orifice that sometimes is inconspicuous. Figures 8.7, 8.8, 8.9, and 8.10 show different appearances of the same malformation (perineal fistula).

We intentionally eliminated confusing terms used in the past to refer to this defect. These terms include: "anal membrane," "membranous stenosis," "covered anus," "anocutaneous fistula," and "translevator defect" [63–68].

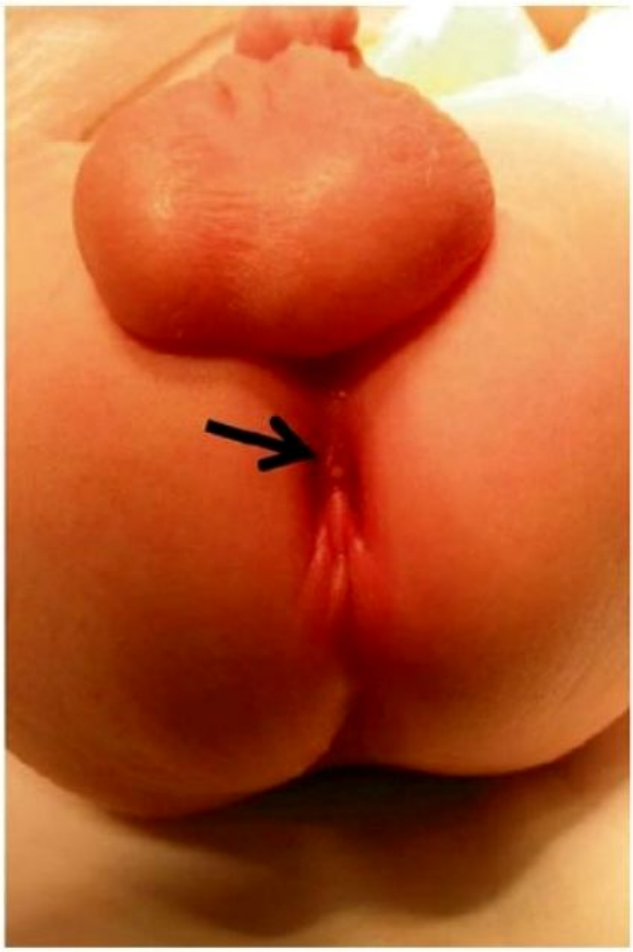


Fig. 8.7 *Arrow* shows fistula opening

The orifice may go undetected. This may have important therapeutic implications. We have seen 17 patients born with this defect in which the surgeon was unaware of the presence of a perineal orifice, believed that the patient had a "high anorectal malformation" opened a non-indicated colostomy. Subsequently, the patient either did not have a high-pressure distal colostogram or had a colostogram without high pressure, and in both circumstances the surgeon "confirmed" his erroneous belief that he was dealing with a case of "high" anorectal malformation. Because of that, he went ahead with an abdominoperineal operation designed for the treatment of more complex malformations, an operation that is not indicated in these cases, and that leaves the patients fecally incontinent. This is something tragic that should never occur if one follows specific steps in the diagnosis of these defects.

The perineum of these patients may have different external appearances. Sometimes they

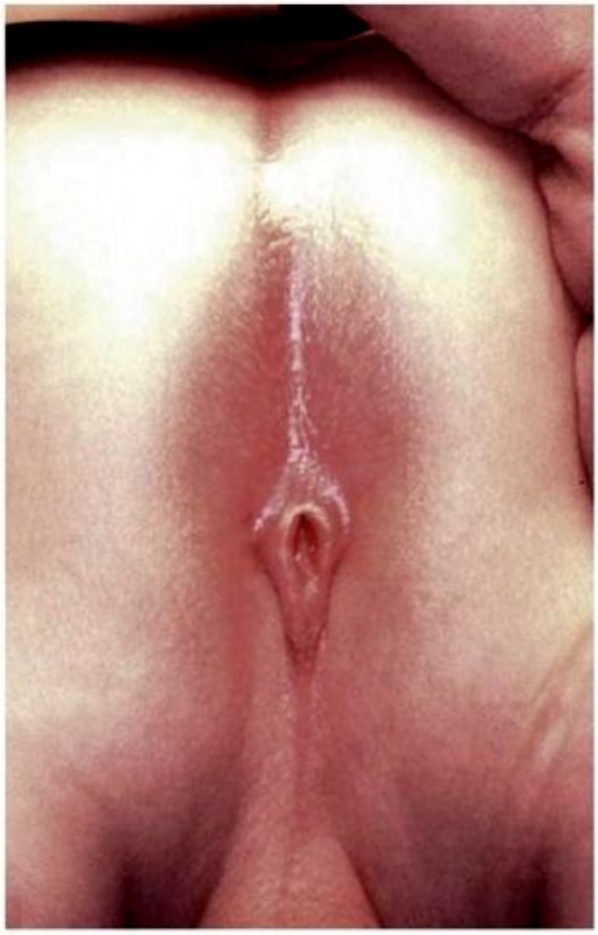


Fig. 8.8 The discoloration of the skin, posterior to the fistula opening represents the location of the sphincter

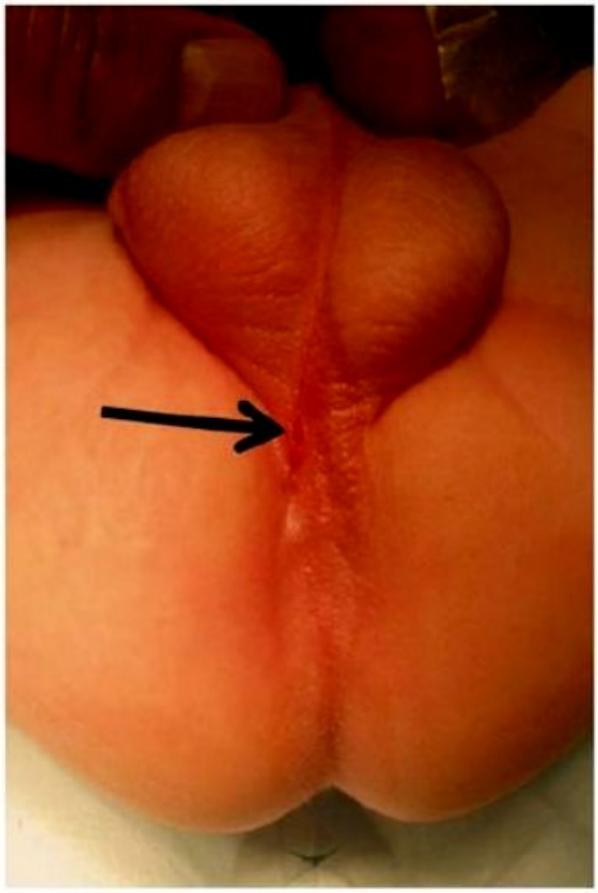


Fig. 8.9 *Arrow* shows fistula opening a little out of the midline

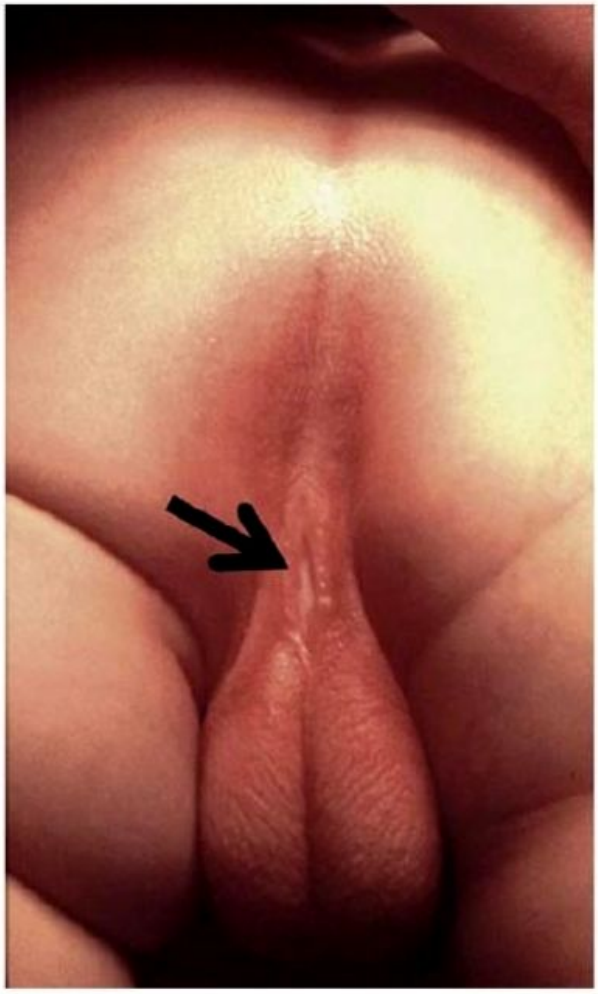


Fig. 8.10 *Arrow* shows fistula opening at the base of the scrotum

have what is called "bucket handle" malformation. Figure 8.11 shows three different variants of a "bucket handle" malformation. Below the strip of the skin (bucket handle), one can find the fistula orifice. Sometimes, the orifice is located at the base of the penis (Fig. 8.12). In other cases the fistula runs subcuticular in the midline raphe, forming a black ribbon type of structure, which represents the presence of meconium running in a subepithelial tunnel (Fig. 8.13). Other times, we do not see a black ribbon type of structure but rather a white ribbon which represents mucous in the subepithelial fistula (Fig. 8.14). The treatment in these patients consists in unroofing that tract until we reach the real fistula and then to continue with the technique described in this chapter.

One must be aware of the fact that sometimes, some patients are born with a fistula located at the base of the penis or in the penis itself, and one believes that it runs at a subcuticular level until it reaches the rectum, and therefore, it can be treated

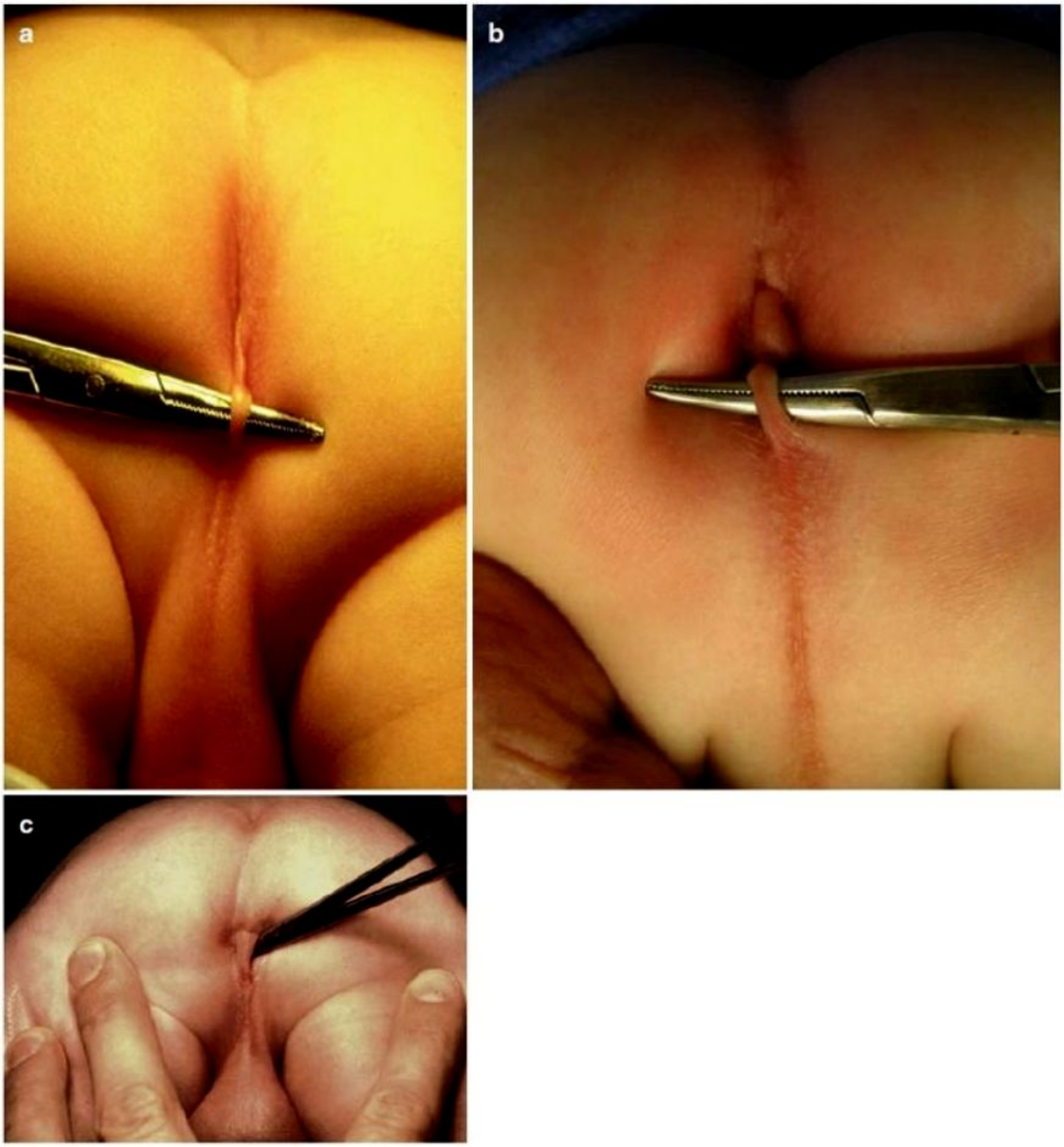


Fig. 8.11 Bucket handle malformations. Three different variants (**a–c**)

like a regular perineal fistula. Yet, surprisingly, one may find that the fistula runs rather parallel to the urethra, deeper and deeper in to the perineum, and through the corpora (Fig. 8.15). As a consequence, trying to follow that structure, the surgeon may provoke significant bleeding. Actually, these specific variants are exceptions to the rule. These malformations are considered rather complex because they require more mobilization of the rectum as well as to be separated completely from the urethra. Fortunately, in most of the cases, one can say that if one is able to see a perineal orifice passing meconium, it means that we are authorized to operate without a colostomy in the way that it was described.

A sacral defect seen in an anterior/posterior view of an x-ray film (Fig. 8.16) is always associated to a presacral mass [38], and the defect can be very small (Fig. 8.17) or giant (Fig. 8.18). It can be located laterally, giving the impression of a

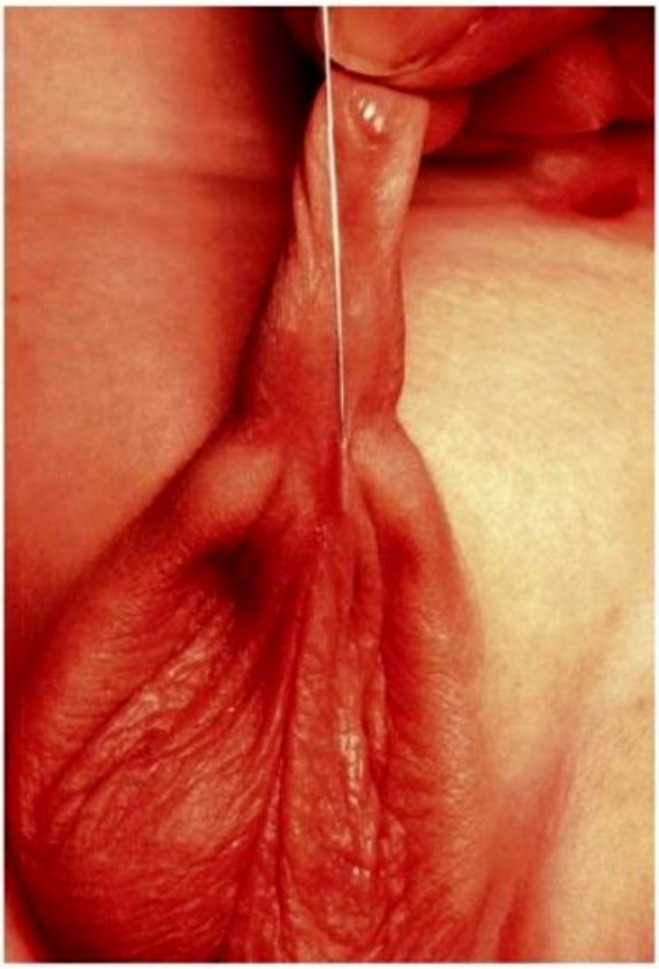


Fig. 8.12 Perineal fistula located at the base of the penis

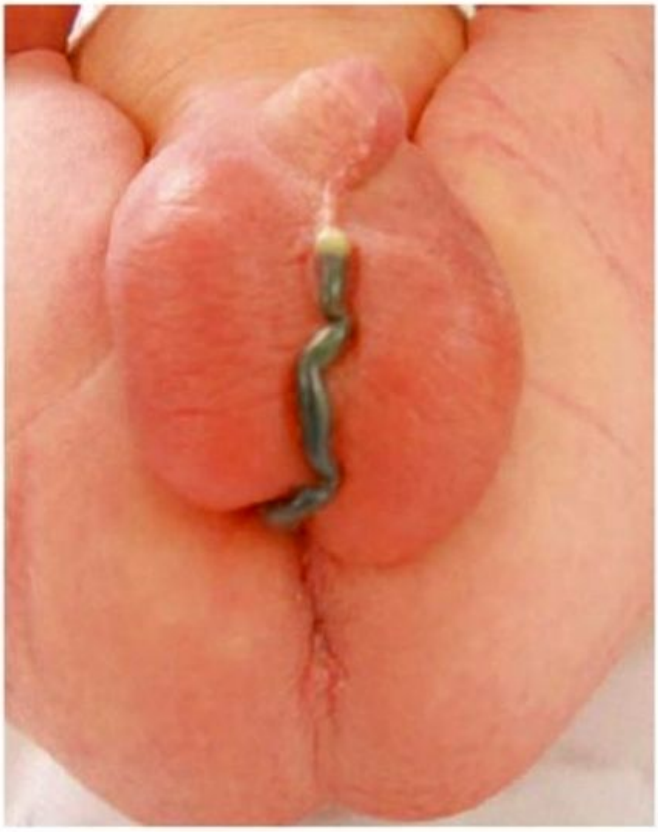


Fig. 8.13 "Black ribbon" malformation. Variant of a perineal fistula (subepithelial with meconium)

Fig. 8.14 "White ribbon" malformation (subepithelial with mucus) – variant of a perineal fistula

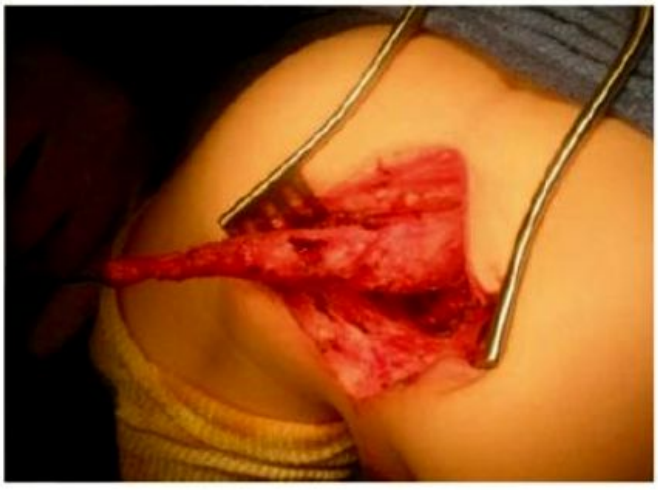


Fig. 8.15 Operative picture of a long narrow perineal fistula. Very rare variant

"hemisacrum" or "scimitar." It can also be located in the midline (Fig. 8.19) giving an image of a "bifid" sacrum. The size and location of the sacral defect usually equals the location and size of the mass. An MRI study will provide more accurate information about the characteristics of the mass,

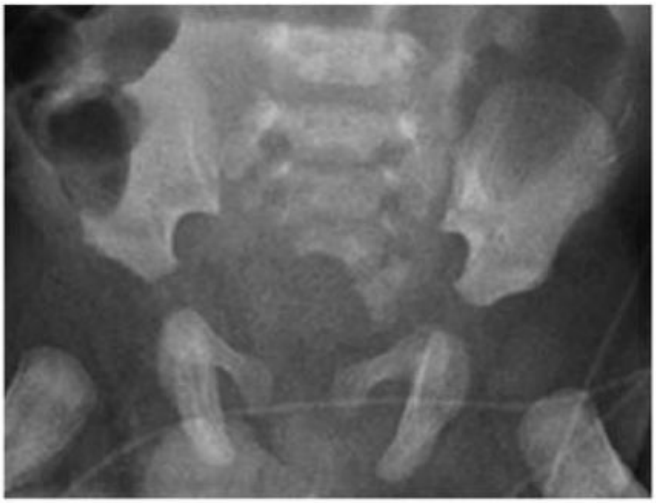


Fig. 8.16 Hemisacrum

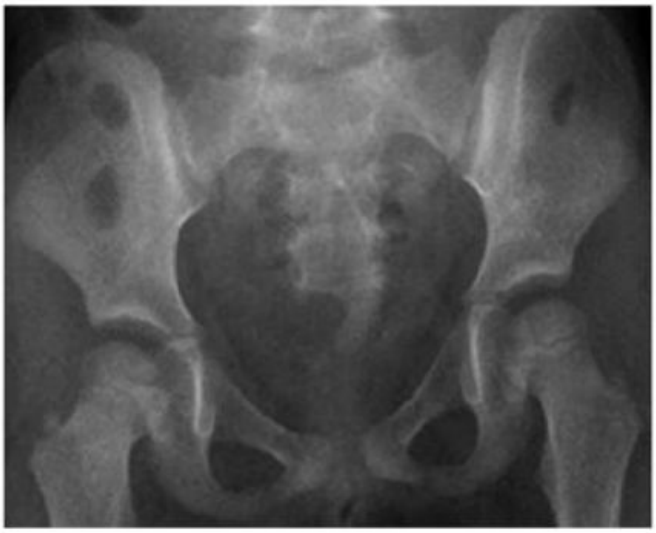


Fig. 8.17 Small sacral defect

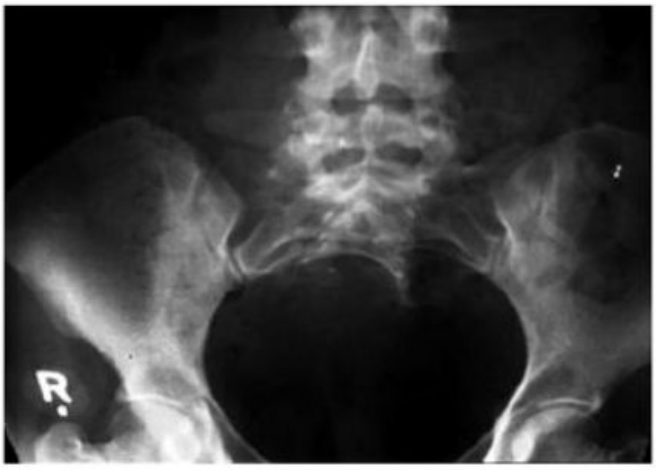


Fig. 8.18 Giant sacral defect. The size of the presacral mass corresponds to the size of the sacral defect

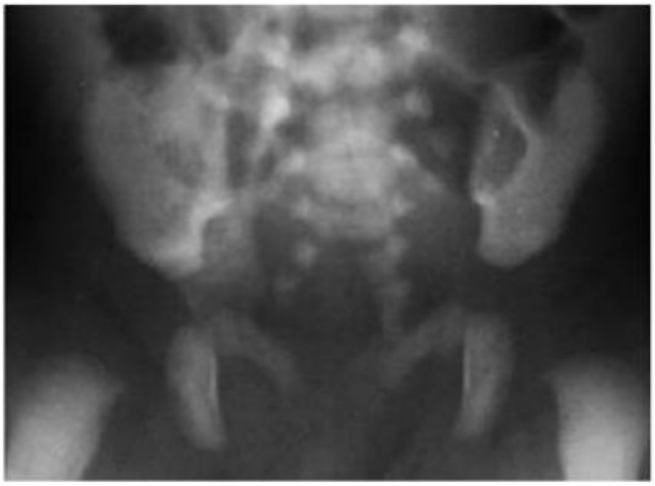


Fig. 8.19 Bifid sacrum, the mass is located in the midline

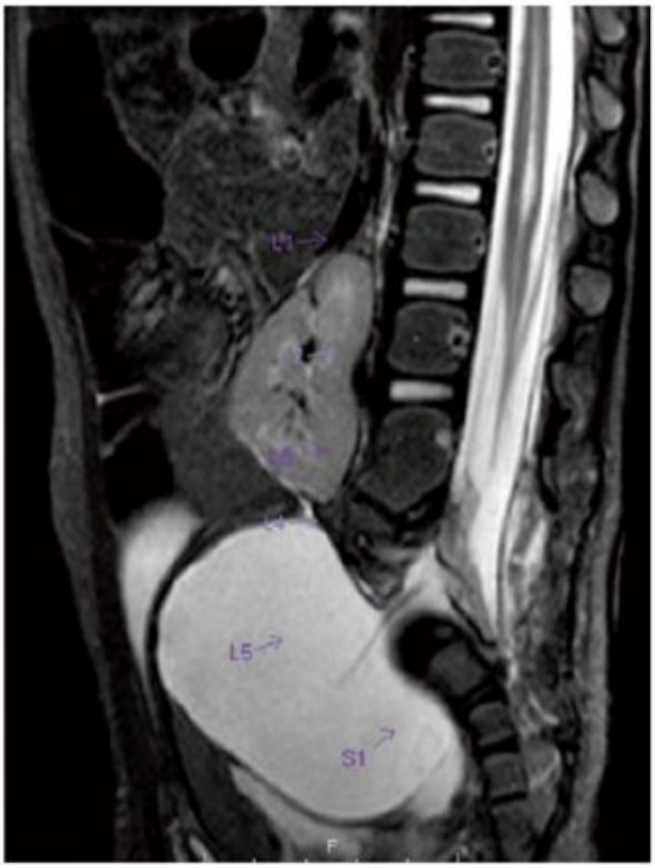


Fig. 8.20 MR image of a presacral mass with an anterior meningocele

as well as the possible communication with the dural space (anterior meningocele) (Fig. 8.20). The most common types of presacral masses are, by far, teratoma/dermoid, lipoma, meningocele, or a combination of all of these. Presacral masses associated to perineal fistulas or to "rectal stenosis" represent an excellent example of the importance of having a good index of suspicion. The presence of the mass may dramatically change the functional prognosis for bowel and urinary function, particularly if the sacral defect is large.

In other words, a perineal fistula with normal sacrum has an excellent functional prognosis, yet the presence of a mass is a rather devastating finding. The surgical strategy also changes, since it must include the resection of the mass at the same time of the repair of the anorectal malformation. This resection can be a rather easy procedure or can also be a formidable operation that requires the participation of a neurosurgeon.

Some patients are born with a normally located anus and a presacral mass producing a rectal stenosis (Fig. 8.21). The perineal appearance of these patients deserves a special comment. This type of perineum is known as "funnel anus" (Fig. 8.22). It certainly looks like a funnel. The pectinate line is not visible outside because it seems to be located higher, by the pushing effect produced by the mass. This external appearance must prompt us to order an AP x-ray film of the sacrum and an MRI of the pelvis, in order to confirm the diagnosis of the presacral mass and rectal stenosis.

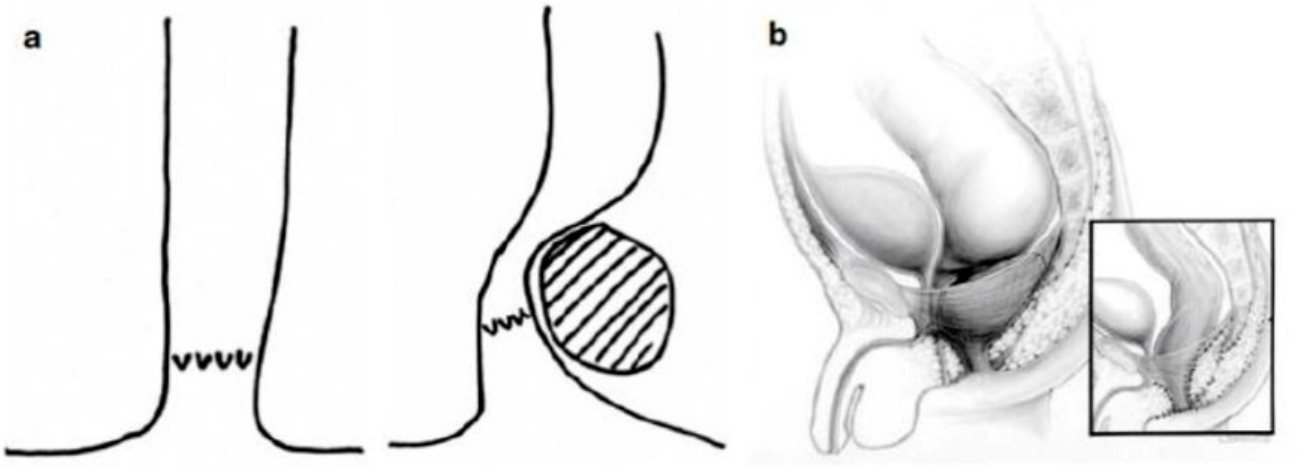
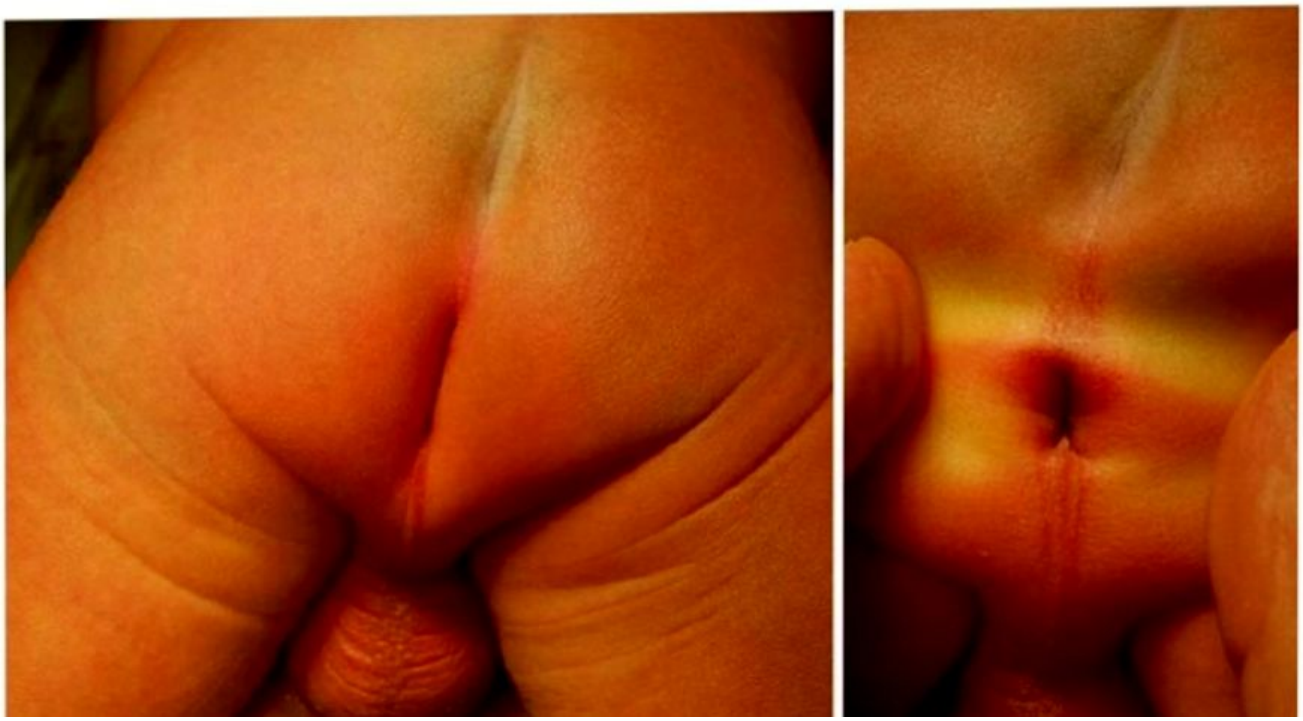


Fig. 8.21 Presacral mass producing a rectal stenosis. (**a**) Diagram showing the rectum compressed by a presacral mass, which pulls up the anal canal. (**b**) Diagram of a perineal fistula associated to a presacral mass (pre- and postoperative)



8.4 Management

Since these defects represent the simplest of all anorectal malformations, the operation designed to repair these defects is also a limited, rather simple, yet meticulous operation. Operations to repair these defects are performed without a protective colostomy. The exceptions to this rule include cases of misdiagnosis. More specifically, some male patients are born with no anal opening; they rather have a tiny perineal orifice that goes unrecognized, and the surgeon erroneously believed that the patient had a "high" malformation and opens a non-indicated colostomy.

When the patients are born full term at our institution, with no serious associated defects, we operate on them without bowel preparation within the first 72 h of life. Unfortunately, however, patients frequently come to us weeks or months after they were born, and in that type of case, we feel it is safer to prepare the gastrointestinal tract with GoLYTELY as described in the chapter related to colonic preparation. In the case of a newborn operated in the first hours of life, we feed the patient usually 3–5 days after the operation. We believe that those babies operated early, without bowel preparation, have a better postoperative course and less chance of infection, perhaps due to the fact that the bacterial proliferation in the bowel is nil or very limited. However, in older patients, we are stricter in the protocol of management. We use GoLYTELY for total bowel preparation (see Chap. 7), insert a central line, and keep the patients 10 days with nothing by mouth.

During the newborn period, there are several therapeutic options for these patients. The first one is simple dilatation of the perineal orifice.

8.5 Dilatations

Dilatations usually help the patient to eliminate the stool and avoid abdominal distention. This treatment modality is preferred by many doctors all over the world, based on the fact that these patients will have bowel control with and without an operation. In fact, misadventure and catastrophic events in these patients occur when somebody performed the wrong type of operation or technically incorrect procedure that ended up in a series of complications that will be described later. Therefore, we can say that it is preferable for the patient to receive anal dilatations rather than a poor operation. The fact is that these patients will have bowel control with and without an operation. We explain that, by the fact that the mislocation of the anal orifice is only present in the lowest part of the rectum (Fig. 8.2), most of the rectum, however, is well located traversing within the funnel-like muscle structure and is only deviated in the lowest part. It is this type of case that exemplifies the fact that a perfect location of the anus, within the sphincter mechanism, is not a precondition to have bowel control. Bowel control then seems to depend on many other factors besides anal location.

Anal dilatations, from our point of view, are then indicated under specific circumstances. One of those could be a patient that has multiple serious associated defects and abdominal distention. Premature babies with severe cardiac conditions that are not in good condition to be taken to the operating room are also good candidates to receive anal dilatations. The protocol of dilatation should be as the one described here (Chap. 18). Another reason to do dilatations is the case in which the surgeon has no experience with performing a formal posterior sagittal anoplasty for these patients. In that case, the chances of hurting the patient are much less with anal dilatations than with an operation. The operation also could be done later. Anal dilatations in cases of perineal fistula are sometimes rather difficult and painful. The reason is that one has to dilate a congenital stricture type of anomaly. This is not the same as in cases of postoperative dilatations after a technically correct operation; in those cases usually the dilatations are uncomfortable, but not painful. Very painful dilatation usually means a congenital stricture or ischemia that occurred during the performance of an anoplasty.

8.6 Cutback Operation

The cutback operation is another therapeutic alternative for these patients. The indications are similar to the ones of the anal dilatation. In fact,

it is a procedure that can be done with local anesthesia. Again, we do not think that it should be the final ideal operation for these patients, but it is certainly an alternative for patients who are extremely sick or in situations in which the surgeon does not have the necessary training to perform a formal posterior sagittal anoplasty. The cutback procedure consists of making an incision in the posterior rim of the anal opening and suturing the skin to the mucosa. In other words, it is a kind of Heineke-Mikulicz type of procedure. The cosmetic appearance after the cutback is less than optimal, yet this operation does not really hurt the sphincter mechanism or the innervation of the bowel, and therefore it is considered a good contemporizing preliminary operation, designed to facilitate the passing of stool and alleviate the abdominal distention.

8.7 Minimal Posterior Sagittal Anoplasty

We believe that the ideal procedure to repair a perineal fistula is what we call "minimal posterior sagittal anoplasty." Our experience with this operation includes 174 cases with excellent results from the functional and cosmetic point of view (Fig. 8.23). We recognize, however, that even when it is a small procedure, it is technically demanding. This is the kind of operation that requires experience, meticulous technique, familiarity with the delicate handling of tissues, and avoiding excessive burning. There is a significant difference in the surgical technique for perineal fistulas in male and female patients.

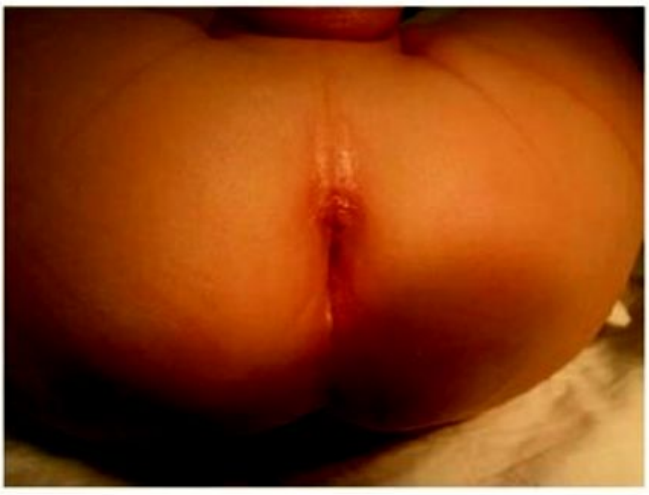


Fig. 8.23 External appearance of an excellent operative result

Obviously, when dealing with patients with perineal fistulas associated to a presacral mass, it is necessary to use a full posterior sagittal incision [47, 69–71].

8.7.1 Male Patients

A few authors referred to this malformation using the terminology that we used, calling this defect "recto-perineal fistula" [72–75]. The real challenge in male patients with perineal fistula is the separation of the rectum from the urethra (Fig. 8.24). In fact, the most common and feared intraoperative accident of this procedure is the urethral injury. This happens when the surgeon underestimates the complexity of this operation and takes the baby to the operating room without a Foley catheter in the bladder. Many surgeons operate on these babies in the lithotomy position; we do it in prone position. One must keep in mind that the urethra of an infant or a newborn male baby is an extremely delicate thin structure and can be divided inadvertently. That is why we cannot overemphasize the need to use a Foley catheter in all of these cases. In addition, during the repair, one must keep thinking very specifically in not injuring the urethra. Figure 8.25 shows a suprapubic cystogram and retrograde urethrogram in a patient that underwent an attempted failed repair of a perineal fistula and suffered a complete division of the urethra. The patient was passing urine through the perineum, and no urine was coming out through the penis. The percutaneous cystogram showed that the urethra had been divided. We had reoperated on several patients like this; we were able to separate the urethra from the rectum and put together both ends of the urethra successfully. This complication is a serious one, and that is why we insist that if the surgeon does not have experience with this kind of operation, it is better to subject the patient to anal dilatations and/or cutback type of procedure.

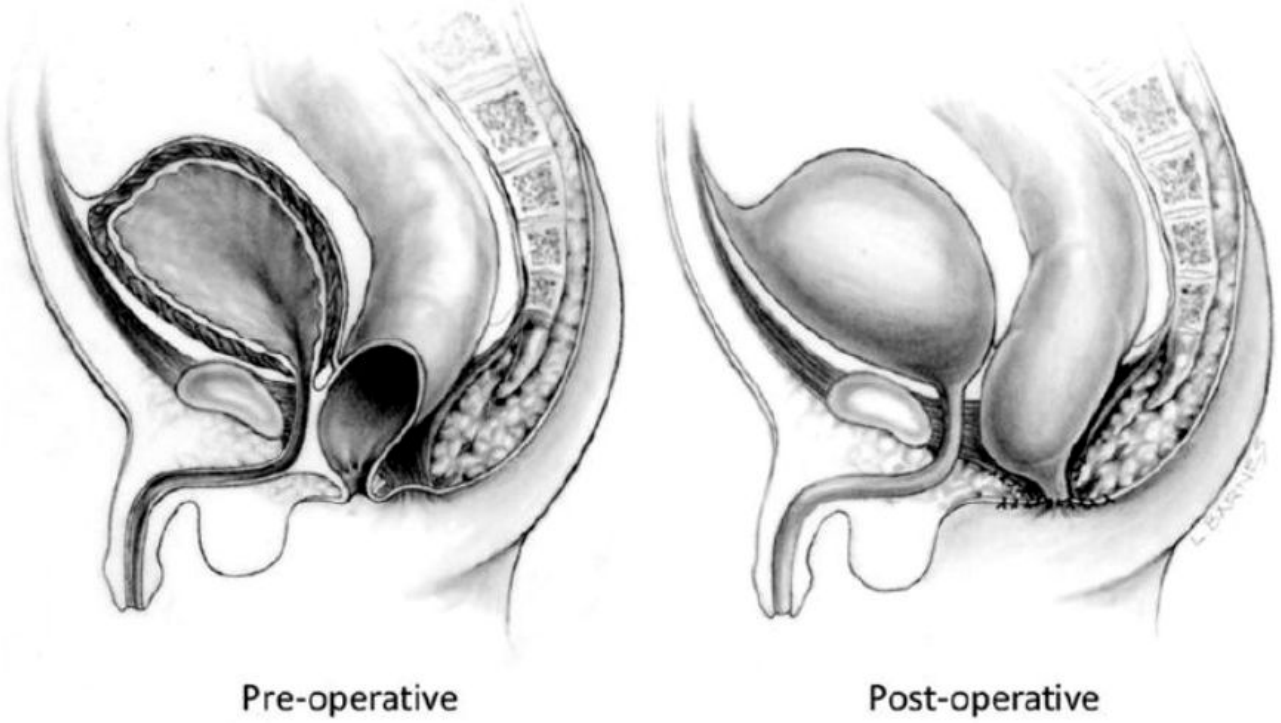


Fig. 8.24 Diagram of a perineal fistula in a male patient, pre- and postoperative

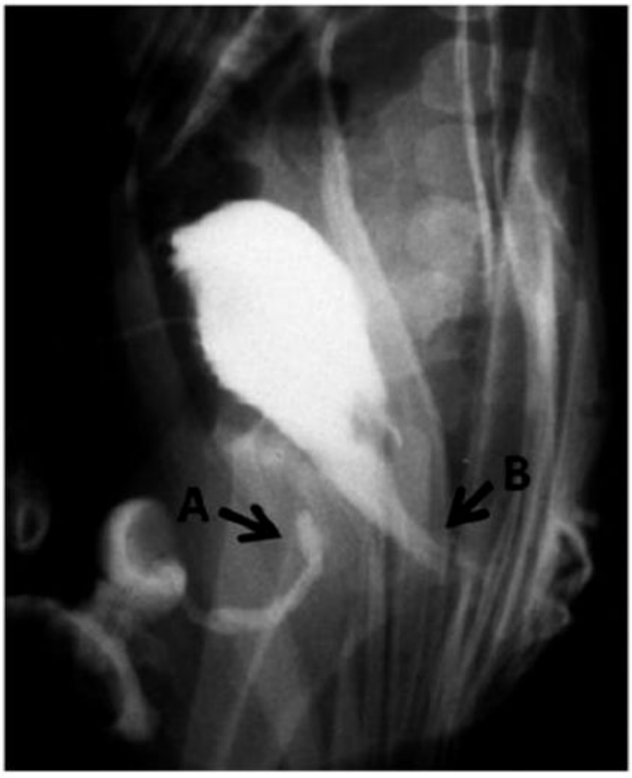


Fig. 8.25 Suprapubic cystogram and anterior urethrogram in a patient who suffered from an accidental complete division of the urethra, during the repair of a perineal fistula. (**a**) Blind end of the distal urethra. (**b**) Distal urethra draining next to the anus

8.7.2 Surgical Technique

Usually, a no. 8 Foley catheter goes well for a full-term newborn baby, but if the baby is smaller than that, we use a no. 6 Foley catheter. The baby is then turned into the prone position with the pelvis elevated. The skin of the perineum is washed, prepped, and draped in the usual manner. Multiple 6-0 silk stitches are placed in a circumferential manner taking the edges of the fistula site (Fig. 8.26). These multiple stitches are used in order to apply uniform traction on the rectum to facilitate the separation of the rectum from the peripheral tissues, particularly from the urethra. We recommend making a small posterior sagittal incision that includes the entire sphincter mechanism (Fig. 8.27). We have been learning from all these cases that the sphincter mechanism can be easily seen, particularly in white babies because it is represented by a characteristic, dark discoloration of the skin. In a full-term baby, the anterior posterior diameter of the sphincter mechanism is approximately 2–3 cm. The incision is continued deeper through the entire sphincter mechanism. The surgeon must keep in mind the

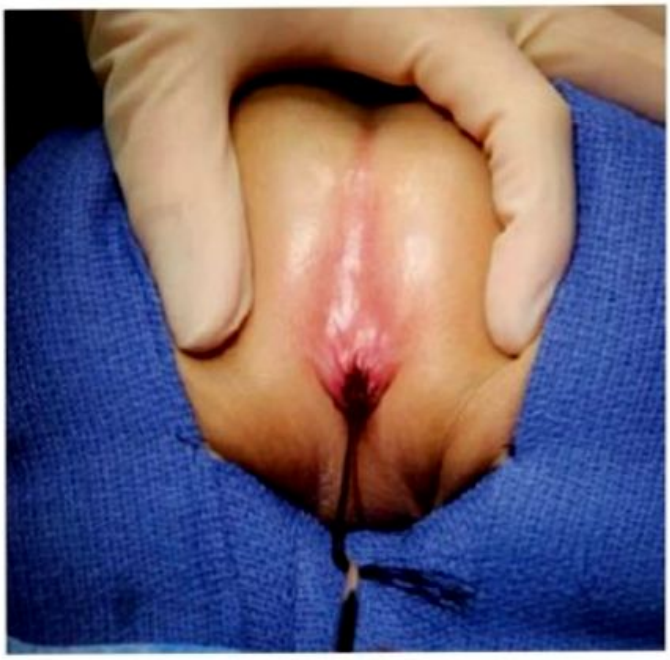


Fig. 8.26 Surgical repair of a perineal fistula in a male patient – multiple silk stitches on the fistula site

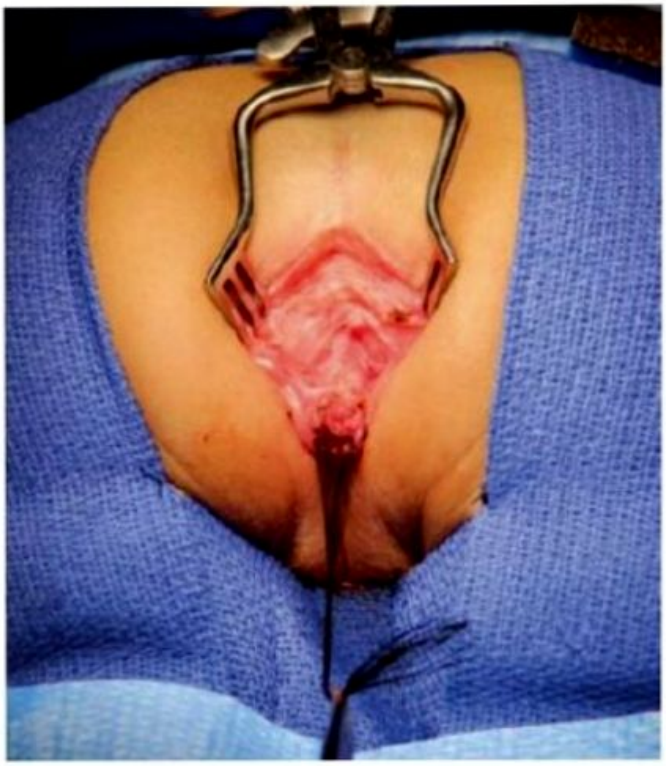


Fig. 8.27 Posterior sagittal incision, dividing the sphincter mechanism

fact that near the fistula site (most distal rectum), the rectal wall is almost in intimate contact with the skin; as we become more proximal in the bowel, the distance between the skin and the bowel is greater. The entire sphincter mechanism is divided until we are able to identify the characteristic white

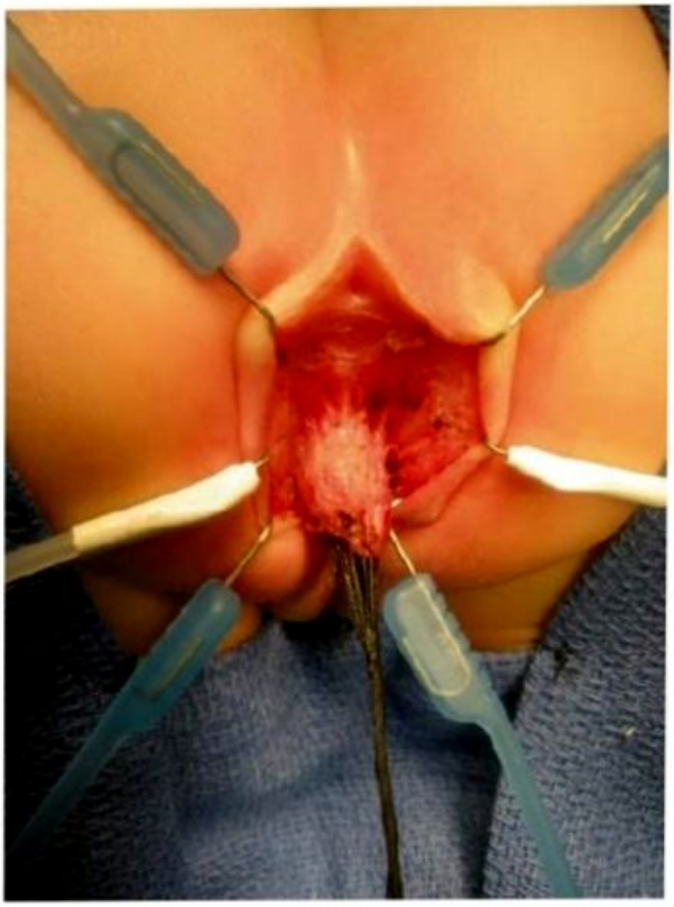


Fig. 8.28 Picture showing the characteristic "white fascia" covering the posterior rectal wall

fascia that covers the posterior rectal wall. This becomes more evident when we apply traction on the rectum (Fig. 8.28). The fascia is removed from the posterior rectal wall in order to create a plane of dissection in intimate contact with the rectal wall. The dissection of the rectal wall is continued in both lateral sides and from there; the dissection is extended down to the skin. At this point, the next step is the most delicate part of the operation which is the mobilization and dissection of the anterior rectal wall, with special emphasis in not injuring the urethra. The surgeon must keep in mind that while applying traction to the rectum, we are also putting traction on the urethra that may be kinked in an acute angle and to be injured, particularly if the patient does not have a urethral catheter. One important sign that indicates that the surgeon is dangerously getting close to the urethra is to find the kind of bleeding that is seen when working in spongiosum/cavernosum type of tissue of the penis. This indicates that we are dissecting very close to the urethra (Fig. 8.29). If one finds that

kind of bleeding that is difficult to stop with the cautery, that means the dissection is being done too close to the urethra and far away from the rectum. The dissection must be carried out closer to the rectum and away from the urethra. The bleeding originated in the spongiosum tissue is better controlled by suturing the spongiosum capsule with fine 6-0 absorbable sutures. The dissection of the rectal wall and separation from the urethra is indispensable if one wants to really mobilize the rectum to move it back to be placed within the limits of the sphincter without tension. Fear to injure the urethra may provoke that the surgeon does not mobilize the rectum enough, leaving a tension anoplasty with high chances to suffer from dehiscence.

Once the rectum has been completely separated from the urethra and mobilized, the limits of the sphincter are electrically determined and marked with temporary silk stitches. The perineal body then is reconstructed with interrupted 5-0 long-term absorbable sutures. The skin of the perineum (where the fistula used to be located) is sutured with interrupted 6-0 long-term absorbable sutures bringing together the anterior limits of the sphincter. Usually the levator muscle is not touched. Yet, the posterior edge of the muscle complex in both sides of the midline is sutured together with 5-0 long-term absorbable sutures taking with the same sutures a bite of the posterior rectal wall in order to anchor the rectum in a good position. Excessive, damaged tissue from the rectum is resected, and a circumferential anoplasty is performed with 16 stitches of 6-0 Vicryl sutures (Fig. 8.30). As we mentioned before, these patients usually have no pain after this kind of operation (Fig. 8.31). When this operation is performed in a newborn baby passing meconium and no real stool, the babies can be fed 2 or 3 days after the procedure. On the other hand, when the operation is performed, without a protective colostomy weeks or months after the baby is born, we believe that it is safer to insert a central line, keep the baby fasting, and administer parenteral nutrition for 10 days. These babies receive intravenous antibiotics for 48 h.

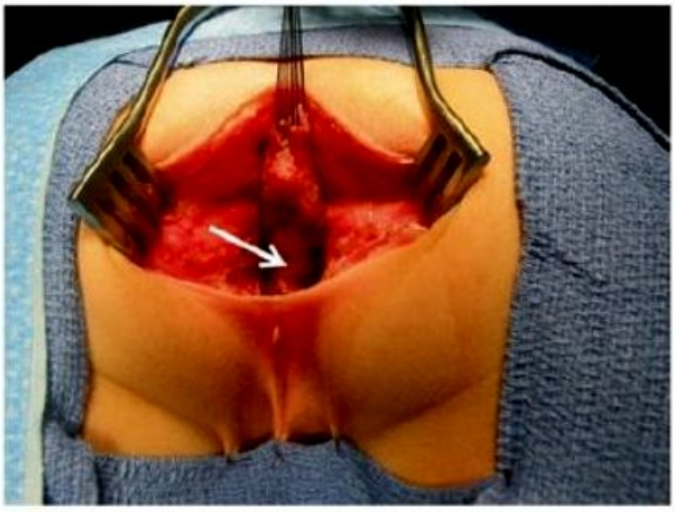


Fig. 8.29 Picture showing the dissection of the anterior rectal wall. The spongiosum tissue of the urethra can be clearly seen

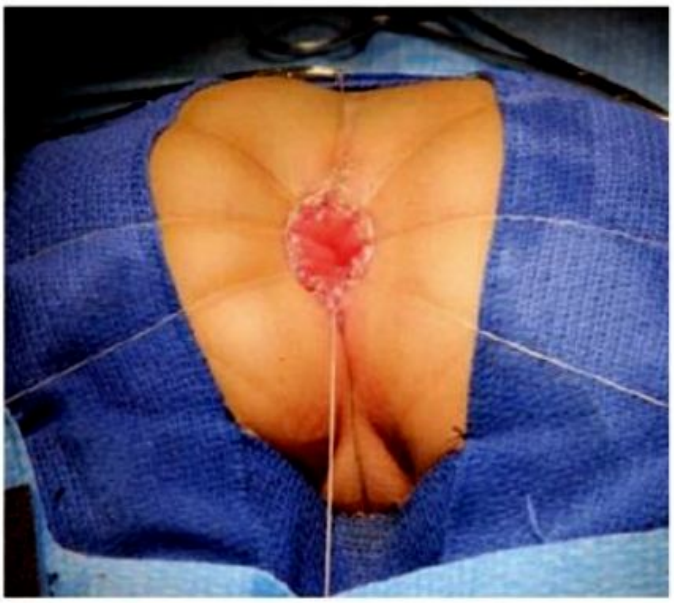


Fig. 8.30 Anoplasty

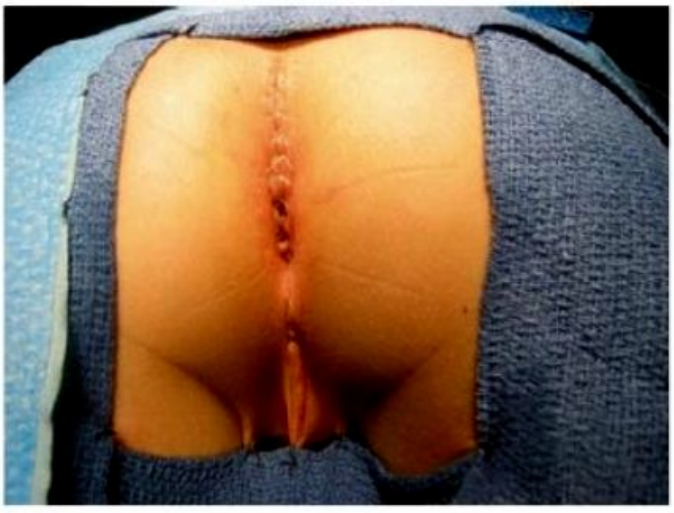


Fig. 8.31 Finished operation

8.7.3 Female Patients

The operation in female patients is usually simpler than in the male patients because there is no risk of injuring the urethra. The equivalent to the urethra would be the vagina, yet it is extremely unusual to see cases of vaginal injuries during the repair of a perineal fistula. This is due to the fact that the vagina and rectum are usually significantly separated although one should not underestimate the possibility of injuring the vagina. The technique is the same as described for male patients except for the fact that there is no possibility of urethral injury and therefore no need of a Foley catheter (Fig. 8.32).

We have been impressed by the fact that one of the anorectal malformations for which the parents have more problems in making a decision about to operate or not to operate is precisely a perineal fistula in female patients. We feel morally obligated to tell the parents that babies with perineal fistulas will have bowel control with and without an operation. Another very important fact in these cases is that the patients will suffer from severe constipation with and without an operation. In other words,

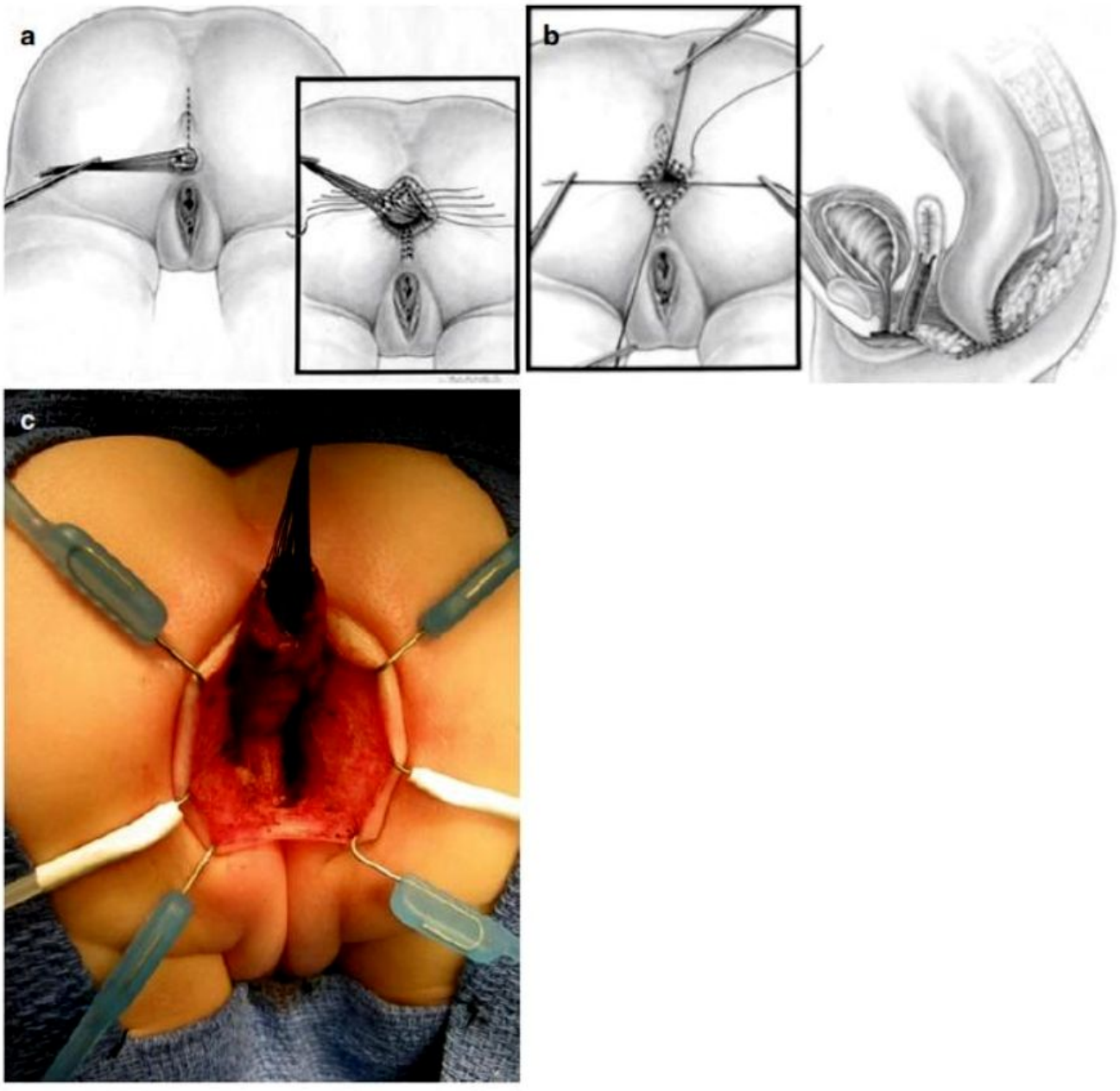


Fig. 8.32 Repair of a perineal fistula in a female patient. (**a**) Traction sutures. Relocation of anal opening. (**b**) Anoplasty. Finished operation. (**c**) Separation of the rectum from the vagina

we should not let the parents develop wrong expectations thinking that the operation will prevent these patients from suffering constipation. In fact, we are not sure if the constipation is going to get worse after an operation and the unpleasant experience of anal dilatations. It is a fact also that not relieving a stenosis (when present) will make the constipation much more severe. Therefore, if the patient has a perineal fistula with stenosis (like most cases have), then something must be done to avoid the exacerbation of the constipation. When one tells the family that the patient will have bowel control with and without an operation and constipation with and without an operation, then they reasonably ask why to operate? There is a reason why we believe that these female patients will benefit from this operation, and that is the fact that leaving the patient with an anteriorly located anal orifice means that they have a very short perineal body. We have seen teenage patients that when they discover the characteristics of their perineal anatomy, they express their dissatisfaction, and they demand a repair later in life, which is a more uncomfortable experience than when it is done in the newborn period. Also, a vaginal delivery of a baby may have higher chances of a rectal injury in a mother with a short perineum. Because of this, we believe that it is indicated to do the operation in female patients provided the surgeon is familiarized with the surgical technique and is delicate enough to do it. In male cases, we cannot argue that the operation will be good for those kinds of reasons, and therefore we simply tell the parents that the advantage of the operation is a better cosmetic effect as compared with cutback or simple dilatations. We respect the parents' decision.

Cases with perineal fistulas associated to a presacral mass usually have a very long narrow fistula with a dilated rectum above the location of the mass (Fig. 8.33). The operation is a sophisticated one because it requires resection of the mass. Most presacral masses do not have an anterior meningocele component. However, many have and therefore if the pediatric surgeon has no experience in dealing with anterior meningoceles must go to the operating room accompanied by a neurosurgeon because it is a rather unpleasant surprise to find that the dura has been open and the patient runs the risk of suffering meningitis and (or) cerebrospinal fluid fistula. After the mass has been resected, the anterior meningocele is resected and the dura is closed with nonabsorbable sutures. Sometimes it is necessary to develop muscle and cartilage patches to be sure that the dura space is well sealed. The mass must always be resected because we have seen patients later in life that suffer from an infection of the mass, and also there is a certain degree of risk of malignancy [41].

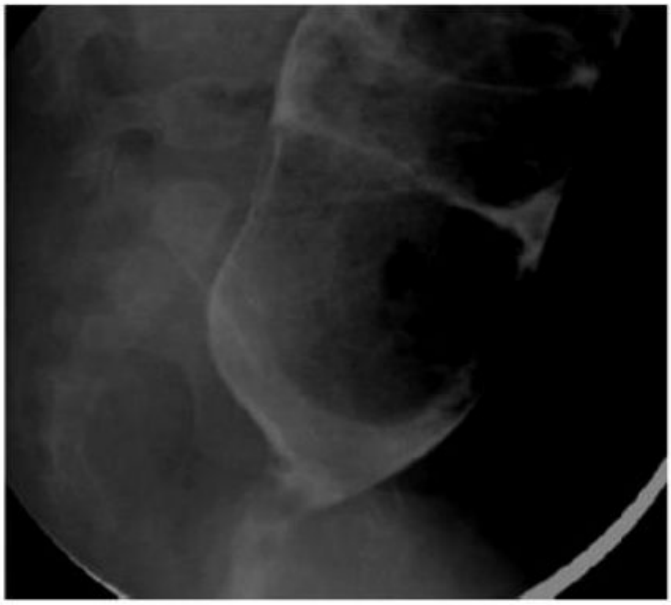


Fig. 8.33 Contrast study showing a dilated rectum and a narrow lower portion compressed by a presacral mass

Once the mass is resected, we can mobilize the rectum down. We should not try to dilate the long narrow fistula that is the result of the compression of the presacral mass. Usually that portion of rectum is non-dilatable and therefore must be resected. Attempts to dilate this kind of rectum have failed in several cases and were referred to us after many attempts of doing that. One must go and find the upper rectum and pull it down to the perineum.

8.8 Postoperative Care

As we previously described, these babies, when operated in the newborn period with meconium in the bowel, can be fed after 3–5 days, and they do not need parenteral nutrition. When the babies

are operated weeks or months later with real stool in their colons, we keep them 10 days with nothing by mouth, receiving parenteral nutrition.

We must anticipate a high degree of constipation in these patients. We cannot overemphasize the importance of assuming that the patient is going to be severely constipated and assuming that he will need an amount of laxatives much larger than what other patients need. The magnitude of constipation is worse when the defect is associated with a presacral mass.

Constipation must be treated aggressively to prevent severe consequences. We must keep in mind that these babies are born with an abnormal rectum that does not have the normal peristalsis, and therefore these patients need help to empty their rectum. We must also remember that every patient has a different laxative requirement that is not what it says in traditional books for the management of constipation. These patients need 2, 3, 4, 5, and 10 times larger dosages of laxatives than other patients need. Constipation means incapacity to empty the rectum which results in accumulation of stool that leads to formation of a fecal impaction that produces megacolon. All pediatric surgeons are familiar with the fact that hollow viscus subjected to an abnormal dilatation loses its peristaltic efficiency. This phenomenon has been observed in cases of colonic, duodenal, and small bowel obstruction, as well as in megaureters. A dilated bowel loses it peristalsis, and therefore, constipation produces retention, retention produces dilatation, dilatation produces lack of peristalsis, lack of peristalsis produces more constipation, creating a vicious cycle that ends up with patients that behave like being incontinent when actually they suffer from overflow pseudoincontinence (see Chap. 18). This is highly inconvenient because we must keep in mind that we are dealing with patients that have an excellent prognosis for bowel control. We have seen 25 patients that came to our clinic suffering from "fecal incontinence" and severe constipation. When evaluated for the purpose of providing bowel management, we found that they were born with one of these malformations, they received a technically correct operation, and yet they suffered from "fecal incontinence." A contrast enema revealed that they had a huge rectosigmoid with fecal impaction. We went ahead with our protocol of disimpaction (see Chap. 25, Sect. 25.7.1). Following that, we determined, by trial and error and radiologic monitoring, the laxative requirement of the patient. We usually found that it was a very large amount of laxative, what the patient required to allow the emptying of the rectosigmoid as radiologically demonstrated. Once we reached that amount of laxative, we found that the patient was actually fecally continent and all the patient required from the beginning was the administration of the right amount of laxatives. Those are very rewarding experiences in dealing with that kind of patients.

Unfortunately, we also have been exposed to about 35 patients that had the same symptomatology just described, but unfortunately a surgeon suspected that the patient could have Hirschsprung's disease. We strongly believe that Hirschsprung's disease is not more common in patients with anorectal malformations than in the general population. Yet, the incidence of constipation in patients with anorectal malformation is extremely high, and most of us pediatric surgeons were trained to suspect Hirschsprung's disease whenever we deal with a child with constipation. Consequently, those patients received a rectal biopsy, and occasionally, those rectal biopsies show no ganglion cells which, from our point of view, do not necessarily make the diagnosis of Hirschsprung's.

In a patient with Hirschsprung's disease, we expect to see not only absent ganglion cells, but we also request an accurate description of the site where the biopsy was taken from, and also we expect from the pathology department to tell us whether or not there was an increase in the activity of acetylcholinesterase as well as the presence of hypertrophic nerves. If we do not find that kind of abnormalities and in the absence of symptoms of enterocolitis, we simply do not believe that these patients have Hirschsprung's. Some of these patients, as we said, had a biopsy done at other institutions that showed unfortunately absent ganglion cells as a single finding with a dilated rectum, which the surgeons considered enough evidence for the diagnosis of Hirschsprung's and went ahead with an abdominoperineal resection. An abdominoperineal resection in a constipated patient with

perineal fistula will certainly cure the problem of constipation, but will also make the patient totally fecally incontinent for life!! A patient with Hirschsprung's disease has a normal sphincter and normal anal canal and therefore is capable of preserving bowel control after the resection of the rectosigmoid provided the operation is performed in a technically correct manner, preserving the anal canal. Yet, patients with anorectal malformations do not have anal canal or have a very abnormal one and therefore cannot tolerate that kind of operation. In summary, one must be very careful before making the diagnosis of Hirschsprung's disease in patients with anorectal malformations.

Another scenario includes patients born with a perineal fistula, suffering from severe mistreated constipation, megacolon, and overflow pseudoincontinence. Some patients are seen by enthusiastic adult colorectal surgeons who offered them a creation of an artificial sphincter or the creation of a sphincter using gracilis muscle or gluteus muscle. Those operations were performed without a good preoperative selection of the patient, and they actually make the patients worse.

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