Rectovestibular Fistula 15

15.1 Definition/Frequency

Rectovestibular fistula is the most important anorectal malformation in females. This is due to the fact that, by far, it is the most common defect seen in females. Two hundred and ninety of our 1,123 female patients were born with this malformation. Two hundred and seventeen were operated primarily by us, and 73 were reoperations due to a previous failed attempted repair. Interestingly, our series include 531 patients with a cloaca. However, we are convinced that this high number of cloacas in our series is because ours is a referral center. We believe that vestibular fistula is much more common in the general population. Most of the vestibular fistula cases are operated at the place where the babies are born, whereas many cloacas are referred to us due to the complexity of the repair. When this malformation is repaired with a meticulous surgical technique, the recovery of the patients is excellent, and the functional results are also very good. Unfortunately, another characteristic of this malformation is the fact that it is frequently mismanaged. We compared the results obtained in a group of patients that were repaired primarily by us with those of the group that had a secondary procedure due to the fact that the patient underwent a previous failed attempted repair. The difference in terms of bowel control is significantly different. Therefore, we can repeat what other pediatric surgeons through history have said, and that is that "these patients have a single opportunity to have a good repair."

In the old literature [1] one can find that this malformation received different names, including "anovestibular fistula." The authors believed that this was a more benign variant of defect and that these patients had a very short fistula and a very low-lying rectum. They also believed that malformation owed to be distinguished from a "rectovestibular fistula," which has a long, narrow fistula and a rectum located higher in the pelvis, and, therefore, they believed that the prognosis was not as good as the one observed in cases of "anovestibular fistula" [1, 2]. We also found the term "vestibular anus"; the authors believed that some patients were born with an otherwise normal anus located in the vestibule of the female genitalia [2]. We have never seen this type of defect. We do not use those three terms mentioned here, because we found that all our patients with an anal opening located in the vestibule can be repaired with the same surgical procedure and have the same functional prognosis; therefore, we consider the old terminology impractical and misleading. It is true that some patients have a longer fistula than others; those cases may require more dissection to bring the rectum down. However, our results are uniformly good, regardless the type of fistula.

Electronic supplementary material Supplementary material is available in the online version of this chapter at 10.1007/978-3-319-14989-9_15.

A. Peña, A. Bischoff, *Surgical Treatment of Colorectal Problems in Children*,
DOI 10.1007/978-3-319-14989-9_15, © Springer International Publishing Switzerland 2015

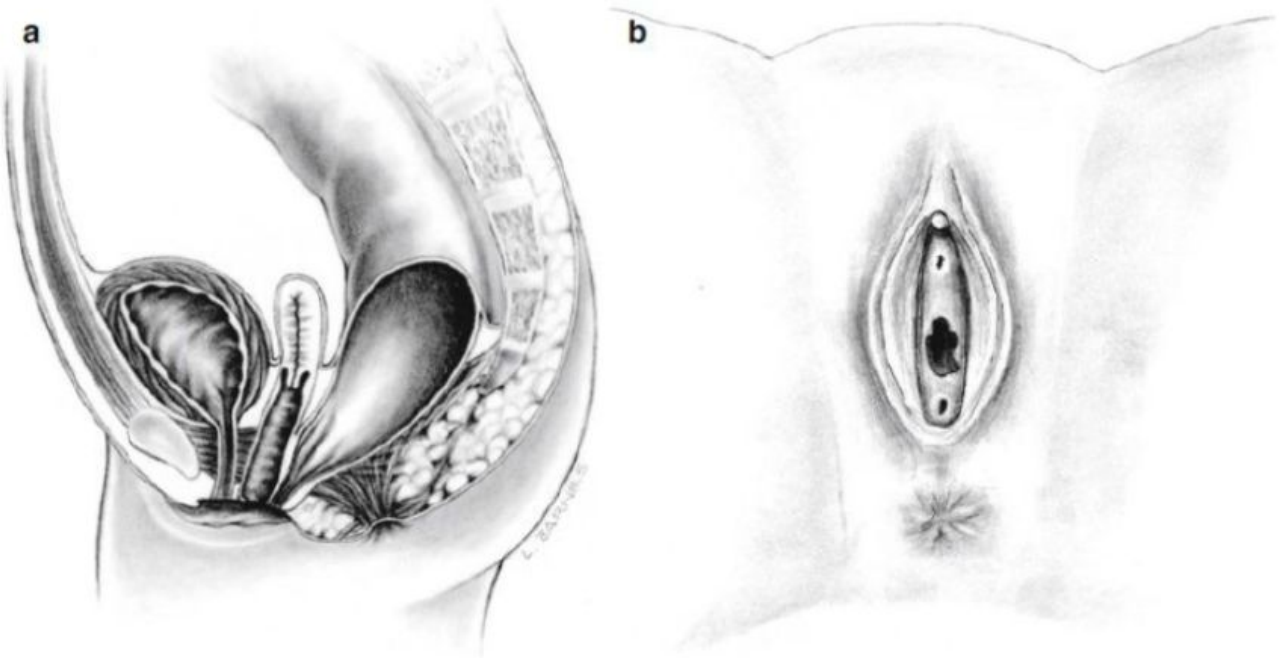

Fig. 15.1 Diagram of vestibular fistula. (**a**) Sagittal view. (**b**) Perineum

Rectovestibular fistula is a defect in which the rectum opens in the vestibule of the female genitalia. This should not be confused with a rectovaginal fistula. In order for us to call a malformation "rectovaginal fistula," one must see the anal opening located inside the vagina, deeper to the hymen. Vestibular fistula patients have a normal hymen, and the anal orifice is located posterior to the hymen (Figs. 15.1 and 15.2). The anal opening is visible most of the time, provided the clinician separates the labia of the baby's genitalia. The newborn female frequently has a significant degree of edema and swelling of that area, considered to be a consequence of the effect of maternal hormones. Therefore, in the newborn baby, it may be a little bit more difficult to see the precise location of the vestibular fistula (Fig. 15.3).

Some cases of vestibular fistula have the anal opening located rather deep and are almost impossible to see it without general anesthesia. In fact, some patients have a rather small-looking genitalia (vulva) similar to what we see in cases of cloaca. The anal orifice is located very deep in the vestibule, and the urethra is also located deeper than normal, which is what urologists call "female hypospadias." This particular variant, we call "cloaca type I," one could also use the term "deep vestibular fistula with a female hypospa-

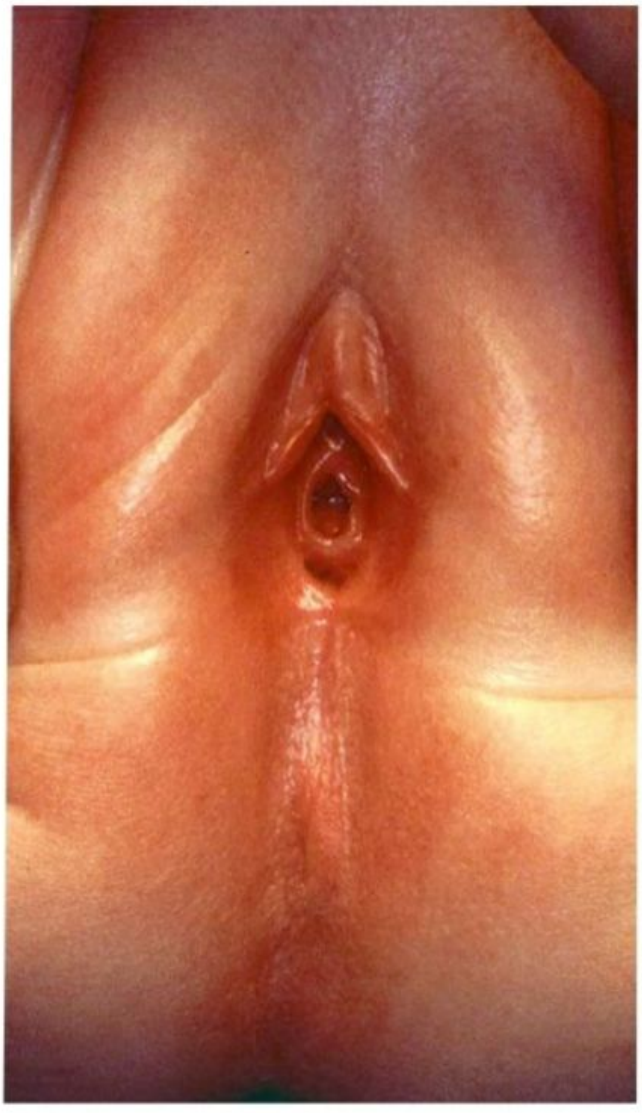

Fig. 15.2 Picture of a vestibular fistula

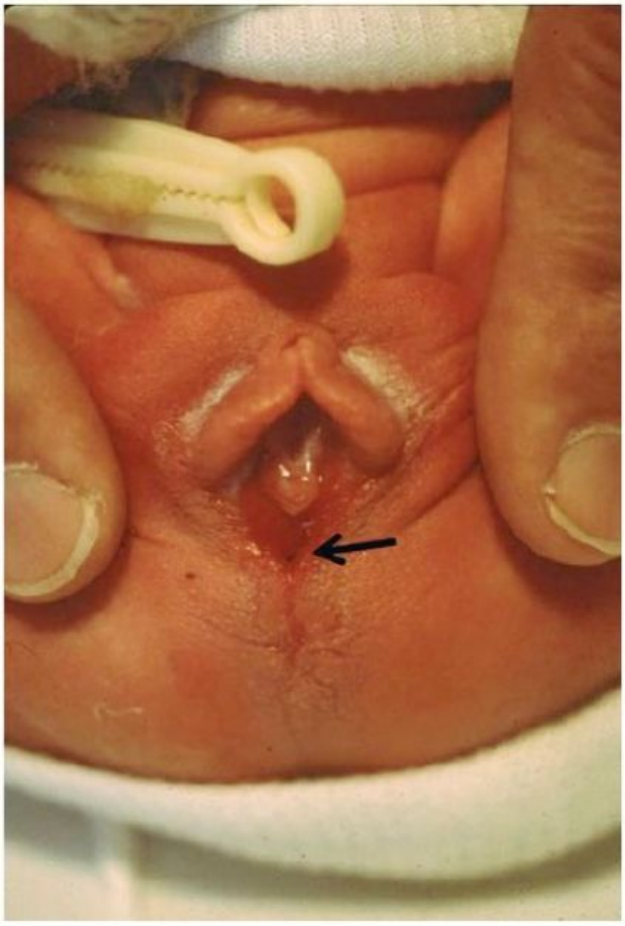

Fig. 15.3 Vestibular fistula in a newborn baby. *Arrow* shows the fistula site

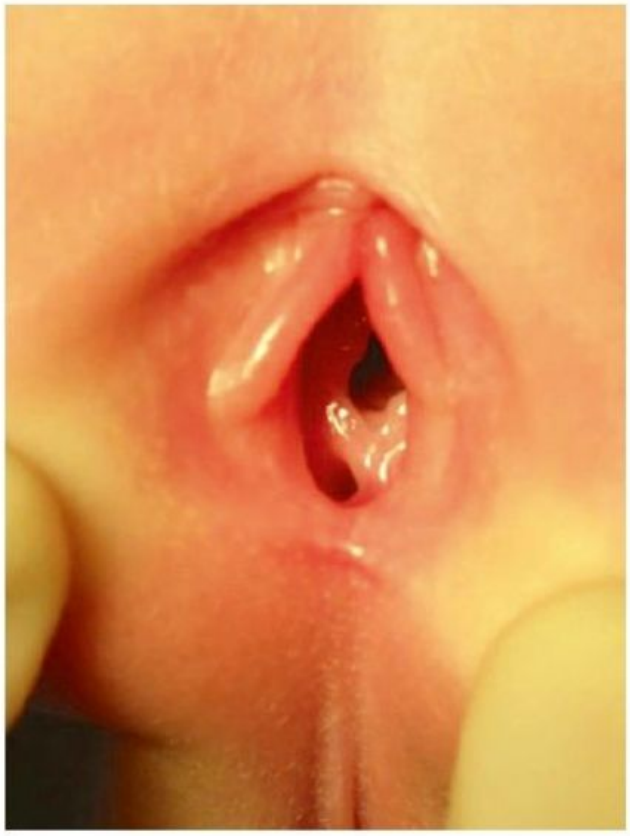

Fig. 15.4 Deep rectovestibular fistula with female hypospadias – observe small vulva

dias" (Fig. 15.4). We consider this particular type of malformation a transition in between a cloaca and a vestibular fistula.

Some patients are born with the anal orifice located just in between the perineal body (skin lined) and the vestibule, wet tissue (Fig. 15.5). This type of defect is considered intermediate between the vestibular fistula and perineal fistula. The management of these patients is not different from any other type of vestibular fistula. This defect is also known as "fourchette fistula."

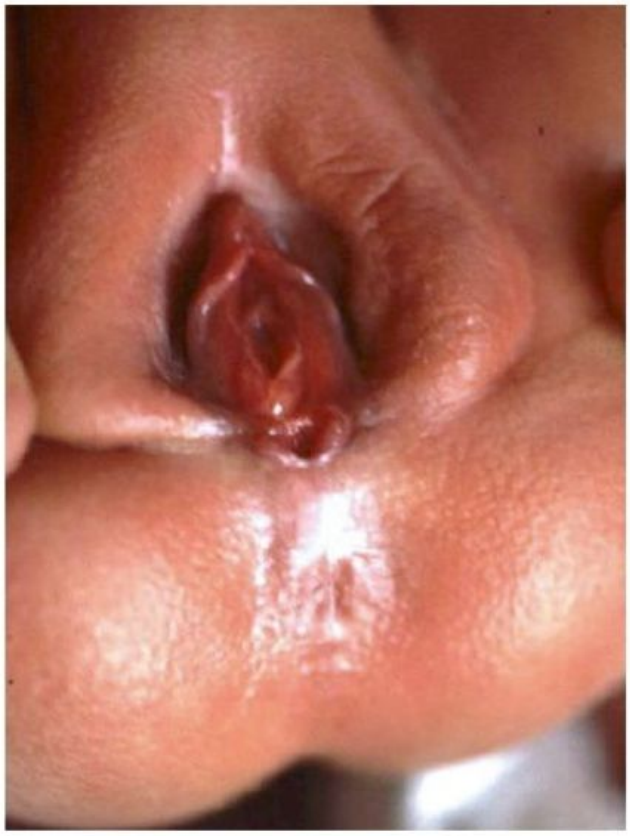

Fig. 15.5 Fourchette fistula

15.2 Associated Defects

A retrospective review of 290 patients with vestibular fistulas operated by us (217 primary and 73 secondary) showed a significant number of associated defects. Since vestibular fistula is considered a malformation representative of the "good side" of the spectrum of anorectal defects

in general, the frequency of association of all the defects is rather low. Yet, it is significant enough to be searched for.

15.2.1 Sacral

We were able to measure the sacral ratio in 113 of our cases and found that the average AP ratio was 0.57 and lateral was 0.7. Six percent of these cases had a ratio lower than 0.4. This is consistent with the fact that we consider this malformation a "benign" one, with good functional prognosis. Fourteen cases had a hemisacrum and a presacral mass, and as previously mentioned, presacral masses occur more frequently in lower defects.

15.2.2 Spinal

Approximately, 9 % of our patients had some form of spinal defect, mainly hemivertebra.

15.2.3 Urologic

Ten percent of vestibular fistula cases had a single kidney, which as we know is the most common anatomic abnormality associated to all anorectal malformations, and 13 % of patients had vesicoureteral reflux, which is consistent with the fact that this disorder is the most common functional urologic abnormality seen in anorectal malformation cases. Hydronephrosis was present in 6 % of the cases.

15.2.4 Gynecologic

There are not many reports in the literature, related to this very important assoc [3, 4]. A retrospective review of our patients with vestibular fistula showed that 17 % of them had associated genital anomalies [5]. Eight percent had absent vaginas or vaginal atresia. Figure 15.6 shows the different types of absent vaginas or vaginal atresias encountered.

Figure 15.6a shows the perineum of one of these patients, and there is no vaginal opening. Figure 15.6b shows a diagram of a sagittal view and the type of repair that we used, consisting in leaving the rectum attached to the urethra, to function as a neovagina and pulling the upper rectum down to the perineum.

Eighty percent of the patients with vestibular fistula and absent vagina are born with agenesis of the internal genitalia (uterus and fallopian tubes). In such cases the vagina is replaced with a piece of colon; this is done only for the patient to have sexual function. Twenty percent of the patients have a uterus and a blind ending of vagina, usually located very high in the pelvis (Fig. 15.6c). In that type of case, the lower vagina is replaced with a piece of colon with dual purpose (sexual and reproductive).

Some cases of vestibular fistula with absent vagina can be repaired without vaginal replacement, but rather pulling down their native vagina. That can only be done in cases with a large blind vagina.

Five percent suffered from some sort of septation disorder of the Müllerian structures. These included a vaginal septum, always associated with the presence of two hemicervices and two hemiuteri (Fig. 15.7). Three patients had a unilateral streak ovary; the rest had two normal ovaries. Two patients had a perineal lipoma, and one patient had a labial hemangioma.

We were able to see patients born with a vestibular fistula that came to us as adolescents; they had a repair in the past, but the surgeons missed the diagnosis of a vaginal septum. These vaginal septa can only be detected when the surgeon suspects their existence. Based on these findings, it is our routine and our recommendation to perform a vaginoscopy with a pediatric cystoscope in all patients with vestibular fistula. The presence of a vaginal septum may, in some cases, interfere with tampon placement and sexual intercourse when the patient grows up. But more important than that is the fact that the presence of a vaginal septum means, by definition, that the patient has two hemiuteri, representing a partial or total septation disorder. Hemiuteri have important gynecologic and obstetric implications. We know that patients with hemiuteri may have a higher degree of infertility, and those patients who become pregnant

have a higher incidence of miscarriages and premature labor. Therefore, it is extremely important to make the diagnosis as early as possible in order to provide these patients with special gynecologic and obstetric care later in life.

It is our routine to do a vaginoscopy in every case of vestibular fistula. We perform that study with a baby cystoscope, during the same anesthesia given for the repair. Figure 15.8 shows the aspect of a normal infant cervix.

15.2.5 Gastrointestinal

Six percent of our patients with vestibular fistula had an associated esophageal atresia, one patient without a fistula, and all the others with a tracheoesophageal fistula; 1 % had a form of duodenal obstruction (atresia or stenosis).

15.2.6 Tethered Cord

Fifty-seven patients were evaluated with an ultrasound (first 3 months of life) or with an MRI, looking for spinal cord anomalies; twenty of them had tethered cord (35 %). These figures are higher than the average of all anorectal malformations, and we believe that this is explained by the fact that the incidence of presacral masses is also high in this malformation. Tethered cord is very common in cases with presacral mass.

15.2.7 Cardiovascular

Twenty-seven patients (9 %) had an atrial septal defect. Twenty-two (8 %) had a ventricular septum defect. Fourteen (5 %) had a patent ductus arteriosus, and four (1 %) suffered from tetralogy

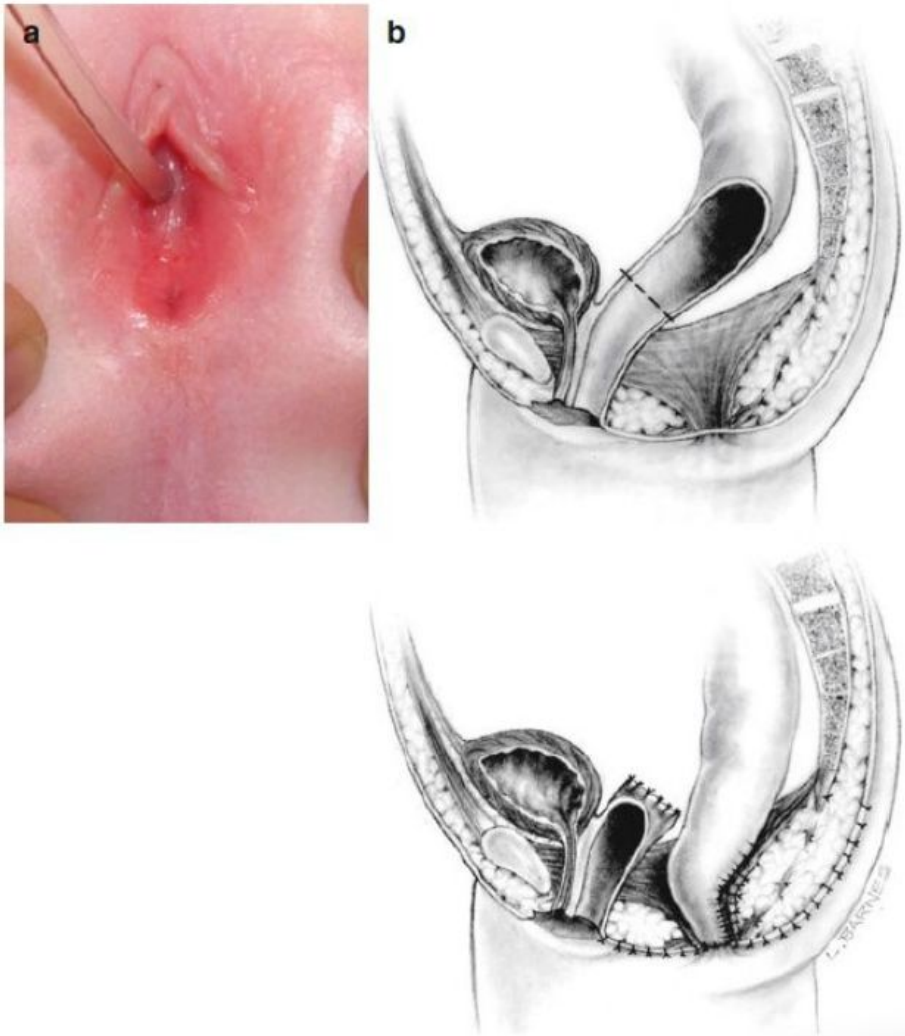

Fig. 15.6 Vestibular fistula with absent or partially absent vagina. (**a**) Photograph. (**b**) Diagram of a sagittal view and one type of repair, using the rectum to replace the vagina. (**c**) Diagram showing the internal genitalia of a patient with a high vaginal atresia, with a piece of colon. (**d**) Diagram showing an absent vagina as well as the uterus, the vagina totally replaced with colon. (**e**) Two types of vaginal atresia. Prepuberty and postpuberty

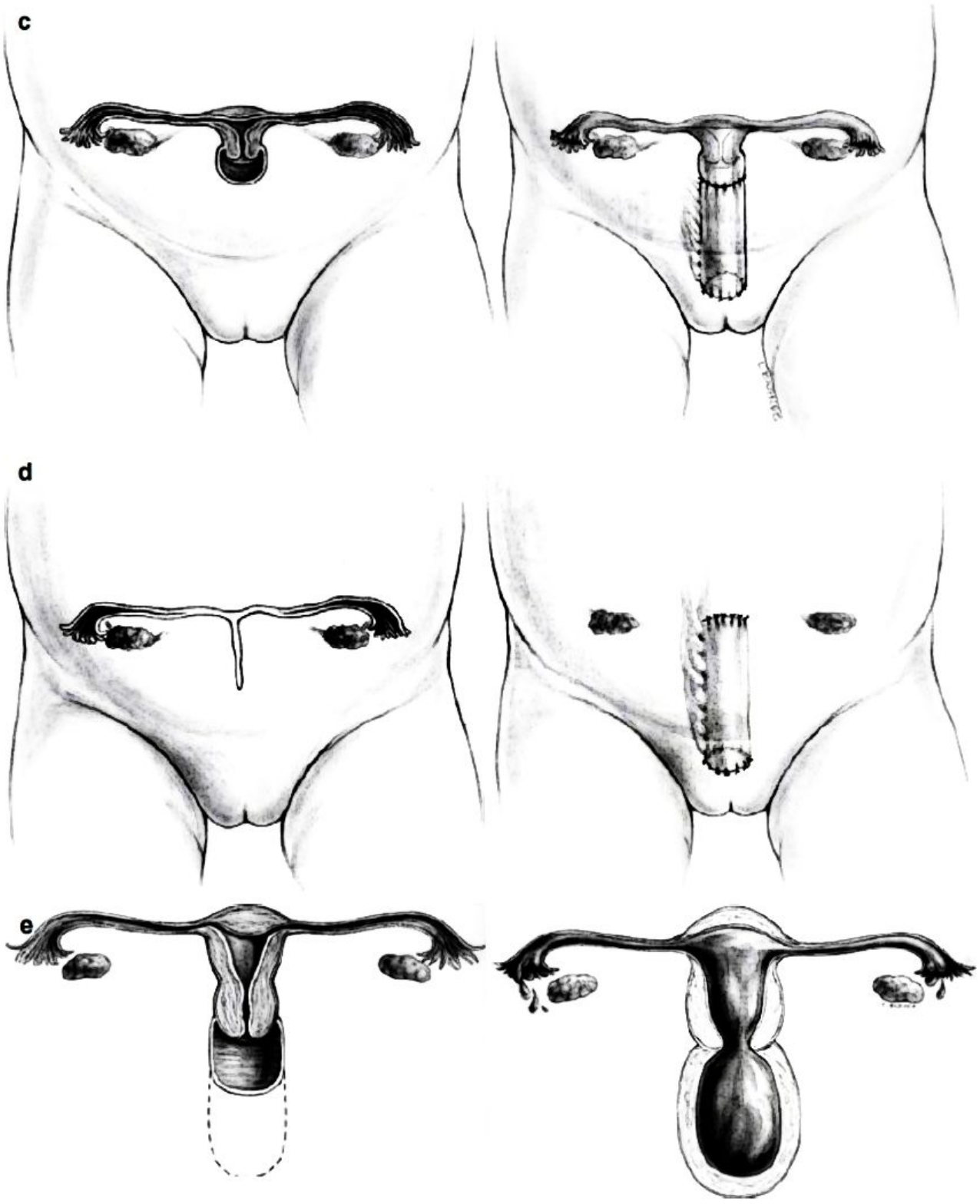

Fig. 15.6 (continued)

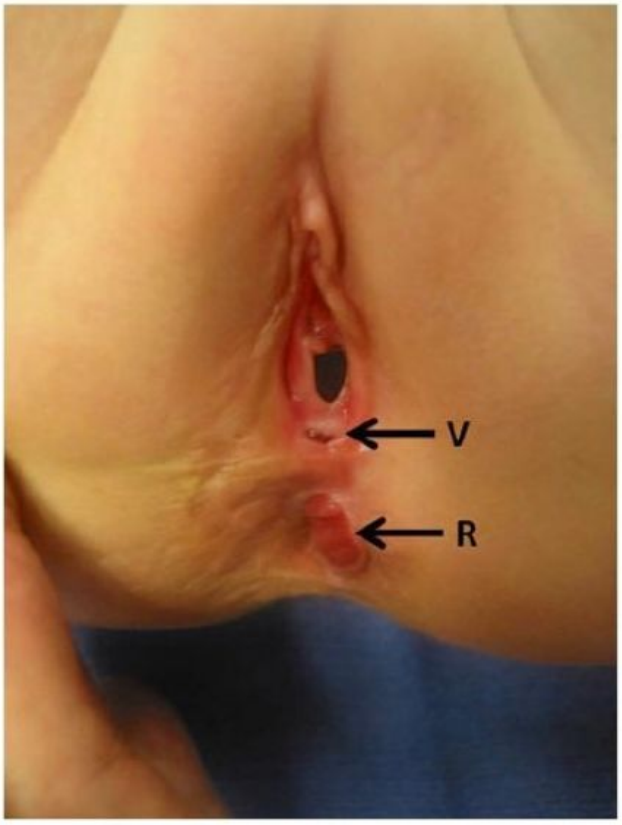

Fig. 15.7 Pocket of the original vestibular fistula in a patient previously operated with the erroneous diagnosis of a "rectovaginal fistula." (*R*) rectum, *V* original fistula

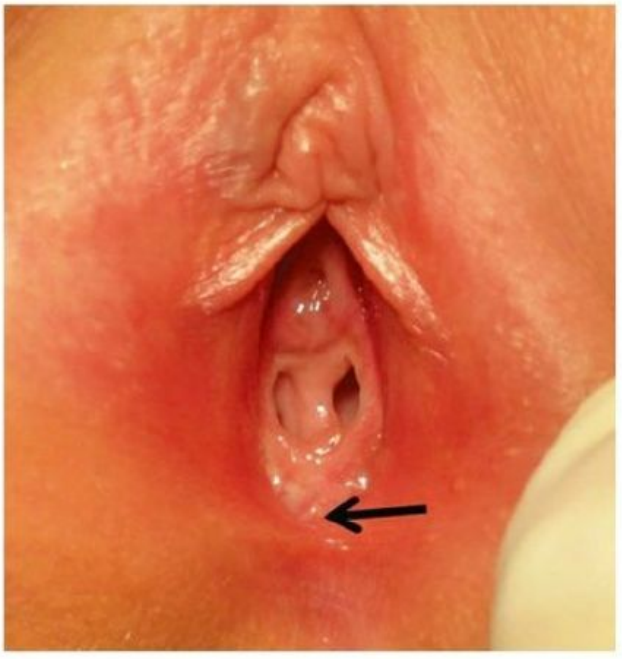

Fig. 15.8 Vestibular fistula with a vaginal septum. *Arrow* shows the fistula site

of Fallot. Most of these defects (approx. 80 %) did not require treatment, since the patients were hemodynamically stable.

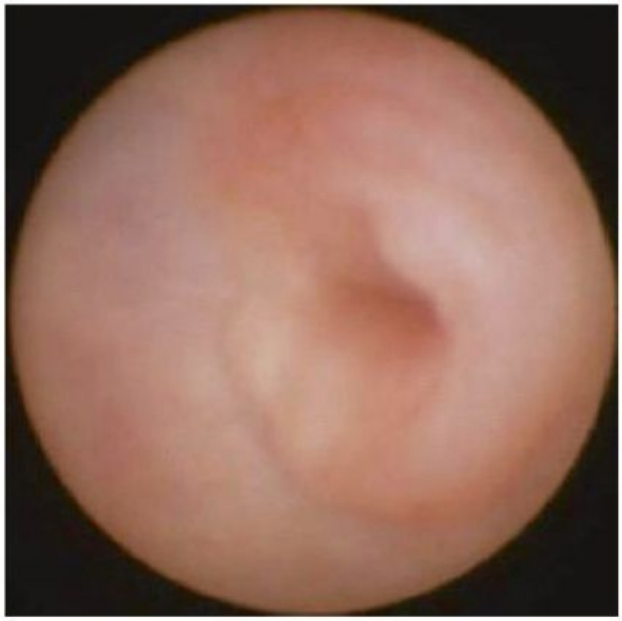

Fig. 15.9 Aspect of a normal cervix in a baby with a vestibular fistula

15.3 Diagnosis

The diagnosis of vestibular fistula is a simple one. It only requires a meticulous inspection of the genitalia of the baby. Yet, amazingly, many patients are not diagnosed or are misdiagnosed as having "rectal vaginal fistula." From our series of 1,123 female patients, we have only seen seven cases of documented real rectovaginal fistula. During the same period of time (over 30 years), we have operated on 290 cases of vestibular fistula. Fifteen of them come to our center with a previous diagnosis of "rectovaginal fistula." Actually, they were born with a vestibular fistula as evidenced by the presence of a little pocket where the vestibular fistula used to be located (Fig. 15.9). Forty-five female patients also came to us after a failed attempted repair of a malformation diagnosed as "rectovaginal fistula." A careful examination revealed that those patients actually had a persistent urogenital sinus, which means that they were actually born with a cloaca and the surgeons only repaired the rectal component of the malformation, because they were thinking that the patient only had a rectovaginal fistula (see Chap. 16, Sect. 16.1.4).

Prior to 1980, the literature [2, 6–13] reported an elevated number of cases of "rectovaginal fis-

tula" in female patients. In contrast, those authors reported very few vestibular fistula cases and very rare cloaca cases. A few publications after 1980 persist reporting "rectovaginal fistula cases." Interestingly, looking at the diagrams of most of those publications, they actually show vestibular fistulas, although they call them "vaginal fistula." The term "vestibular fistula" has been used correctly by some authors with large experience in the management of these defects [14–21].

We believe that this is not a simple semantic problem, but rather has important clinical implications [22]. We have seen patients born with vestibular fistula that were previously misdiagnosed as "vaginal fistula" and underwent a type of repair designed to repair "high" malformations, namely, a contraindicated abdominoperineal (open or laparoscopic) procedure that resulted in fecal incontinence. We also have seen that at least 30 patients born with a cloaca received the wrong diagnosis of "rectovaginal fistula" and underwent a repair only of the rectal component of the malformation, leaving the patient with a urogenital sinus [22].

15.4 Treatment

15.4.1 Colostomy or No Colostomy

This is a frequently debated subject. Many surgeons claim that they routinely repair vestibular fistulas without a colostomy and they have "good results" [12, 23–27]. Many others prefer to open a colostomy in all cases with a vestibular fistula. In the meantime, we see many patients that underwent a repair of a vestibular fistula without a colostomy and suffered from serious complications, including dehiscence and retraction of the rectum as well as reopening of the fistula. However, this recurrent fistula is frequently an acquired rectovaginal fistula, due to the fact that during the attempted repair, the posterior wall of the vagina was damaged.

As discussed in the Chap. 5, we believe that the answer for this question of colostomy or no colostomy is different for every surgeon and his/her different surrounding circumstances. It very much depends on the experience of the surgeon, the clinical condition of the patient, and the infrastructure of the hospital where the patient is treated.

In general, at our institution, if a baby is born with a vestibular fistula, we operate on her within the first 5 days of life without a colostomy, provided the baby is in good clinical condition, is full term, and does not have severe associated defects.

Consider the case of a premature baby with a cardiac condition and vestibular fistula. Under those circumstances, dilatations of the fistula may prove to be useful for the patient to be able to pass stool, eat, and grow. That would allow the surgeon to postpone the decision of colostomy or primary repair. On the other hand, a full-term baby in good clinical condition without associated defects in an institution with a good infrastructure and a pediatric surgeon with experience in the management of this defect, the patient can be operated before starting her feedings, at a time when the patient is still passing meconium, because when it is done in that way, the patient actually does not need any kind of bowel preparation.

Most of our patients come to us after the newborn period and with a colostomy already opened at another institution, sometimes in another country. Many other patients come to us after several months of passing stool with difficulty through the non-operated vestibule, with severe constipation and megacolon. Those patients are also treated without a colostomy at our institution, but our routine includes the admission of the patient 1 or 2 days prior to the main operation, insertion of a nasogastric feeding tube, and administration of GoLYTELY[1] at a rate of 25 mL/kg/h until the colon is completely clean. The patient receives a PICC line and parenteral nutrition for a period of 7–10 days postoperatively.

When the patient has a colostomy, the operation can be done without following this routine,

[1] GoLYTELY… (Polyethylene glycol/electrolytes.) Braintree Laboratories, Braintree, MA, USA

but rather irrigating only the distal stoma of the colostomy the day before surgery. In that case, the baby can eat the same day of the operation; she will stay for 48 h in the hospital receiving intravenous antibiotics. Our experience is that the pain that these patients experience postoperatively is rather minimal. We have operated on primarily without a colostomy in approximately 50 % of our cases.

We use a posterior sagittal approach to repair these malformations. Other approaches do exist, and the most traditional and popular was described by Dr. Potts and is called a fistula transplant [28–32]. Some surgeons describe an operation called "anterior sagittal approach" [33–37]. We found that the word "anterior" in those publications was actually not referring to the incision, but rather to the position of the patient; in other words, the patient is positioned in lithotomy position, rather than prone, but the incision is always posterior to the fistula, because there is no way to make an incision anterior to the fistula site. In other words, the so-called "anterior sagittal approach" is actually a posterior sagittal approach performed in lithotomy position.

Interestingly, Professor Francesco Rizzoli from Bologna, Italy, published in 1869 [38] the technique now referred as "anterior sagittal approach"; his publication includes magnificent illustrations.

The essential components of the posterior sagittal approach described below avoid the flaws observed in those other techniques, and the most common problems seen in our reoperations were retraction of the anoplasty and an inadequate perineal body (anteriorly located anal orifice).

15.5 Main Repair (Animation 15.1)

The patient is brought to the operating room, and we start the procedure with the patient in the lithotomy position in order to perform a vaginoscopy using a baby cystoscope. We do this with the specific purpose to rule out vaginal malformations. Although a vaginal septum can be simply seen by separating the labia without the use of a cystoscope, we prefer to use a cystoscope, because sometimes one can see a vaginal septum that is only present in the lower part of the vagina, and the upper part has a single cervix (Fig. 15.8). Most of the times, however, the septum is complete, and one can see two cervices at the end of the vagina.

The patient is then turned into the prone position with the pelvis elevated, and the perineum, as well as the genitalia and perianal area, is washed, prepped, and draped in the usual manner. Most of the times, one can see the anal orifice in the vestibule, and in such case multiple 5-0 silk stitches are placed at the mucocutaneous junction of the anal opening (Fig. 15.10). These stitches serve the purpose of applying uniform traction to facilitate the separation of the rectum from the vagina. Occasionally, the fistula is located so deep that it is impossible to do this; in such case, we first make the incision and go deep enough to be able to see the edges of the fistula and apply multiple 5-0 silk stitches (Fig. 15.11). We use the electrical stimulation to determine the limits of the sphincter and to guide ourselves to try to stay as much as possible exactly in the midline, dividing the entire sphincter mechanism leaving equal portions of the sphincter in both sides of the midline. For this, we use a needle-tip cautery, changing from cutting to coagulation. The size of the incision usually is shorter than the regular posterior sagittal anorectoplasty. The incision usually runs from the lowest part of the sacrum and coccyx down to the fistula orifice, passing through the sphincter mechanism. We divide the entire sphincter, including the parasagittal fibers, the muscle complex, and the levator mechanism. Deeper to the levator mechanism, one can identify the characteristic white fascia that covers the posterior wall of the rectum (Fig. 15.12).

Figure 15.13 shows the aspect of the rectum with traction sutures. Traction creates the plane. The white fascia is removed from the posterior rectal wall, including the extrinsic blood supply of the rectum. We do this to identify the real rectal wall completely clean. The dissection is then extended to the lateral walls of the rectum (Fig. 15.13). The next step consists of extending the dissection of the lateral walls of the rectum all

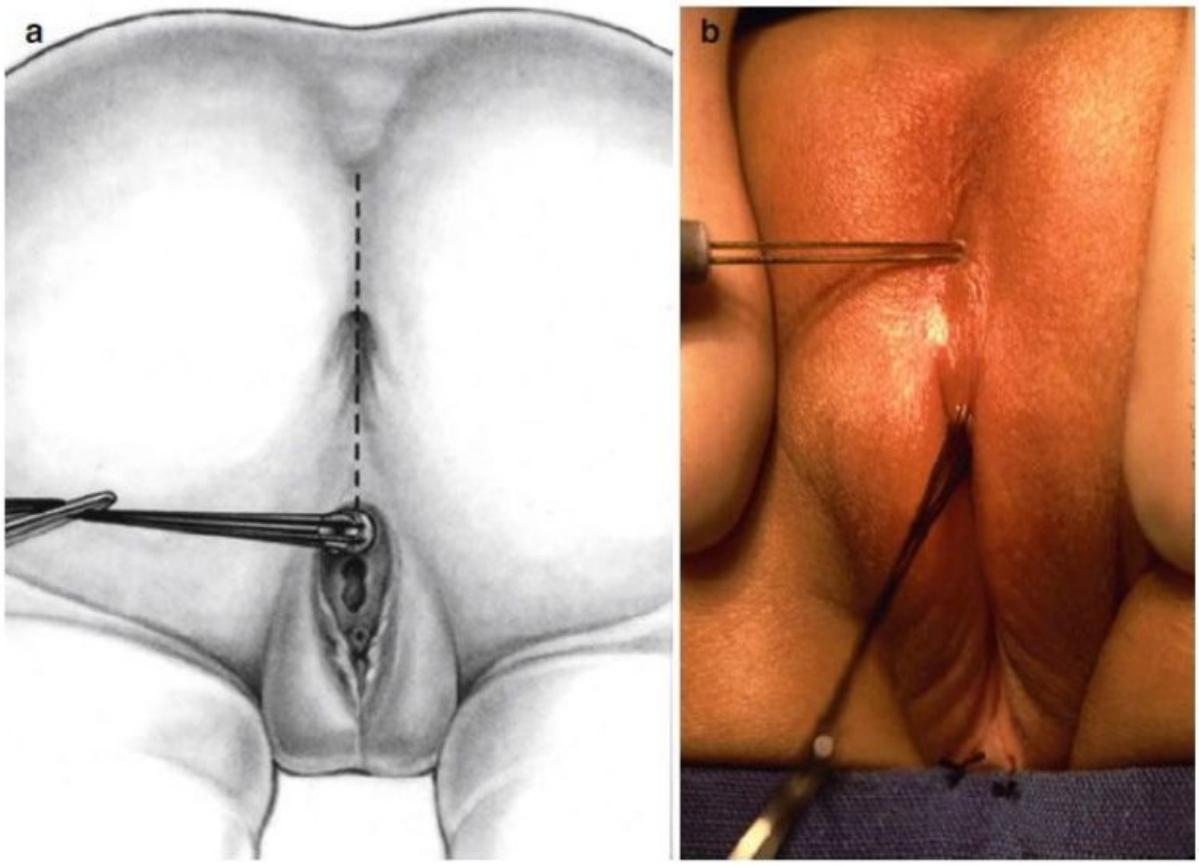

Fig. 15.10 Multiple stitches placed at the anal orifice located in the vestibule. (**a**) Diagram. (**b**) Photograph

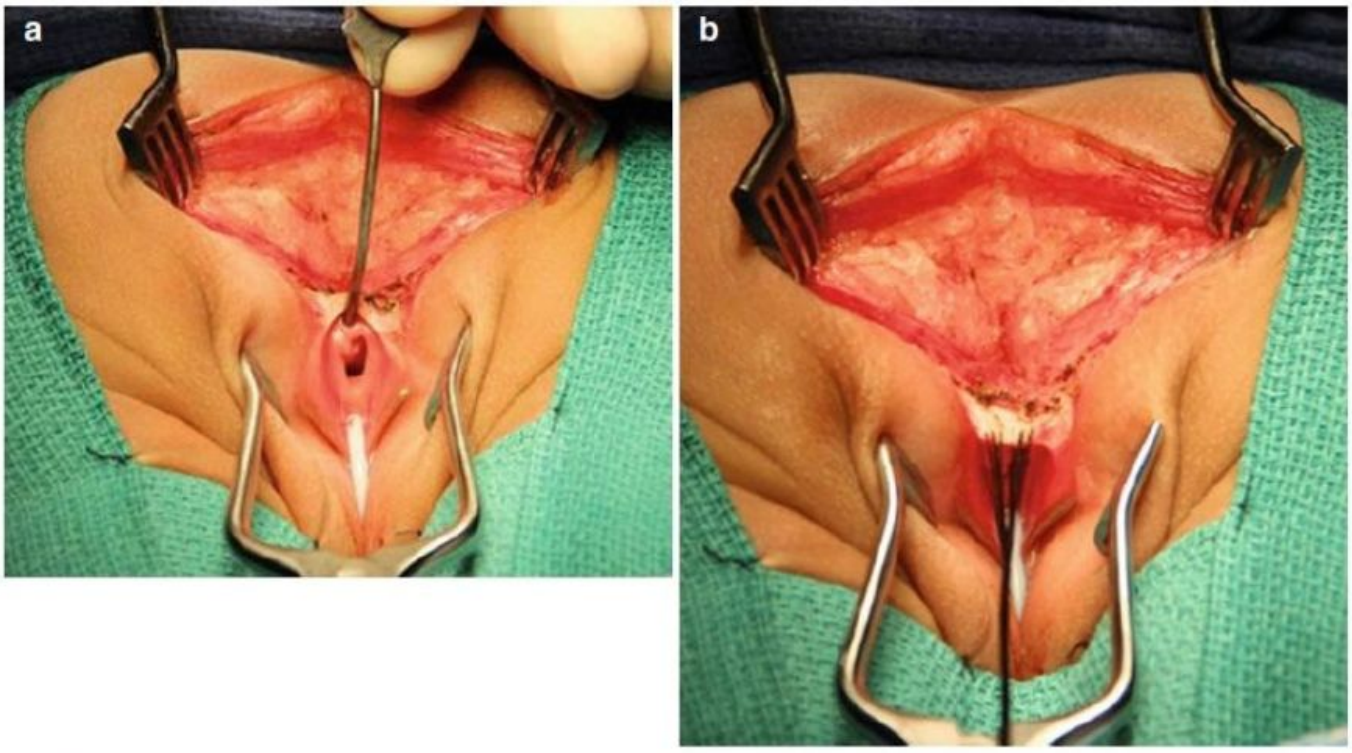

Fig. 15.11 (**a**) Incision – when the fistula is located too deep in the vestibule, we must open first and place the sutures later. (**b**) Multiple stitches in a case of a deep fistula

the way down to the skin. It is important to remember that at the level of the skin, there is no real plane of dissection between the rectal wall and the surrounding tissues. Whereas approximately 1 cm proximal in the rectum, one can clearly identify the plane that separates the rectum from the surrounding tissues, and therefore the recommendation is to follow the steps mentioned

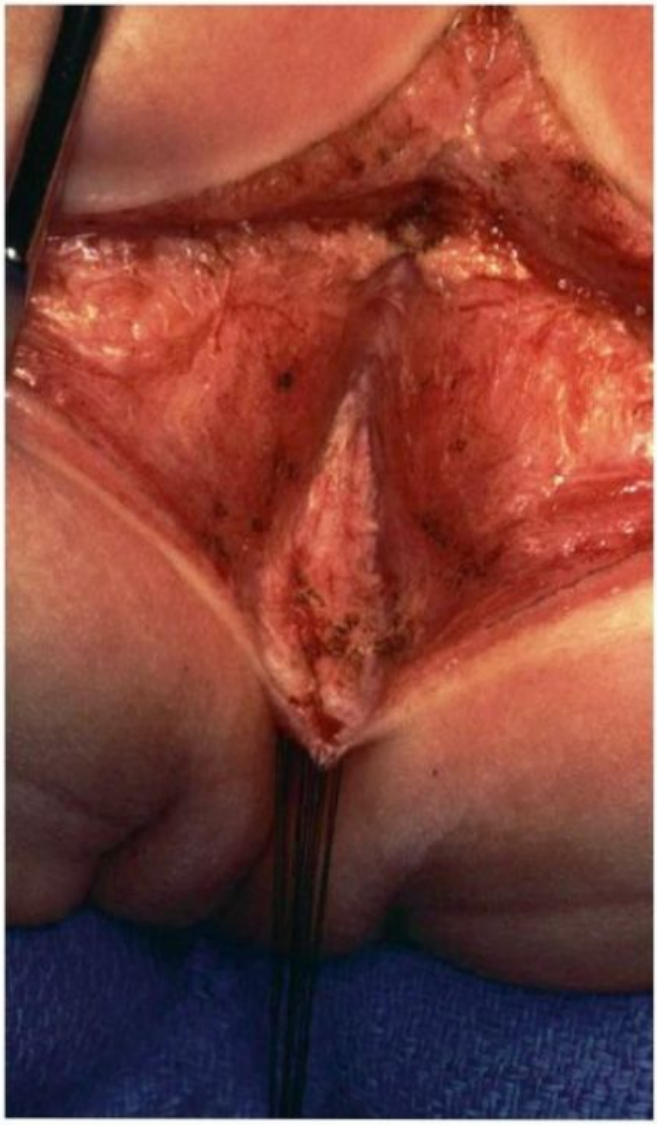

Fig. 15.12 "White fascia" after dividing the entire sphincter mechanism, the rectum is identified covered by the white fascia

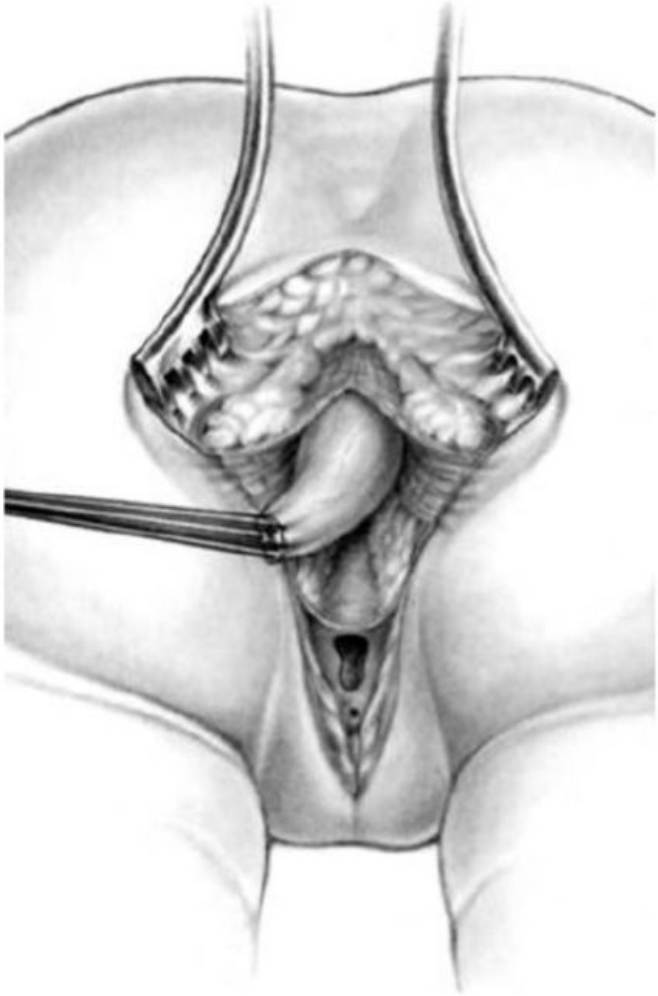

Fig. 15.13 Diagram showing the dissection of rectum

in this description, meaning to identify the posterior rectal wall; continue the dissection to the lateral walls of the rectum and then from there, applying uniform traction on the multiple 5-0 silk sutures; and continue the dissection from the lateral walls of the rectum down to the skin. One must expect to find important vessels that provide the blood supply of the lower rectum while dissecting the lateral walls of the rectum. A Weitlaner retractor is used to achieve adequate, optimal exposure. At this point, we are ready to initiate the most important part of the operation, which is the separation of the rectum from the vagina.

One must keep in mind that the rectum and vagina share a common wall with a variable length from 1 to 3 cm and that there is no real plane of dissection between both structures. In other words, one must make two walls out of one, and very often, this common wall is extremely thin. This happens to be the most important anatomic feature of this malformation. Surgeons must keep in mind that the main challenge in the repair of these defects is the separation of the rectum from the vagina and should take it as a personal challenge. The separation of the rectum from the vagina requires a very meticulous, delicate surgical technique. It cannot be done by blunt dissection. We like to perform this separation using the needle-tip cautery while applying traction on the rectal wall and checking with a lacrimal probe the thickness of the anterior rectal wall and the posterior vaginal wall very frequently, to be sure that we are not getting too close to one or the other (Fig. 15.14). As we progress in this meticulous dissection, the wall of the rectum, as well as the wall of the vagina,

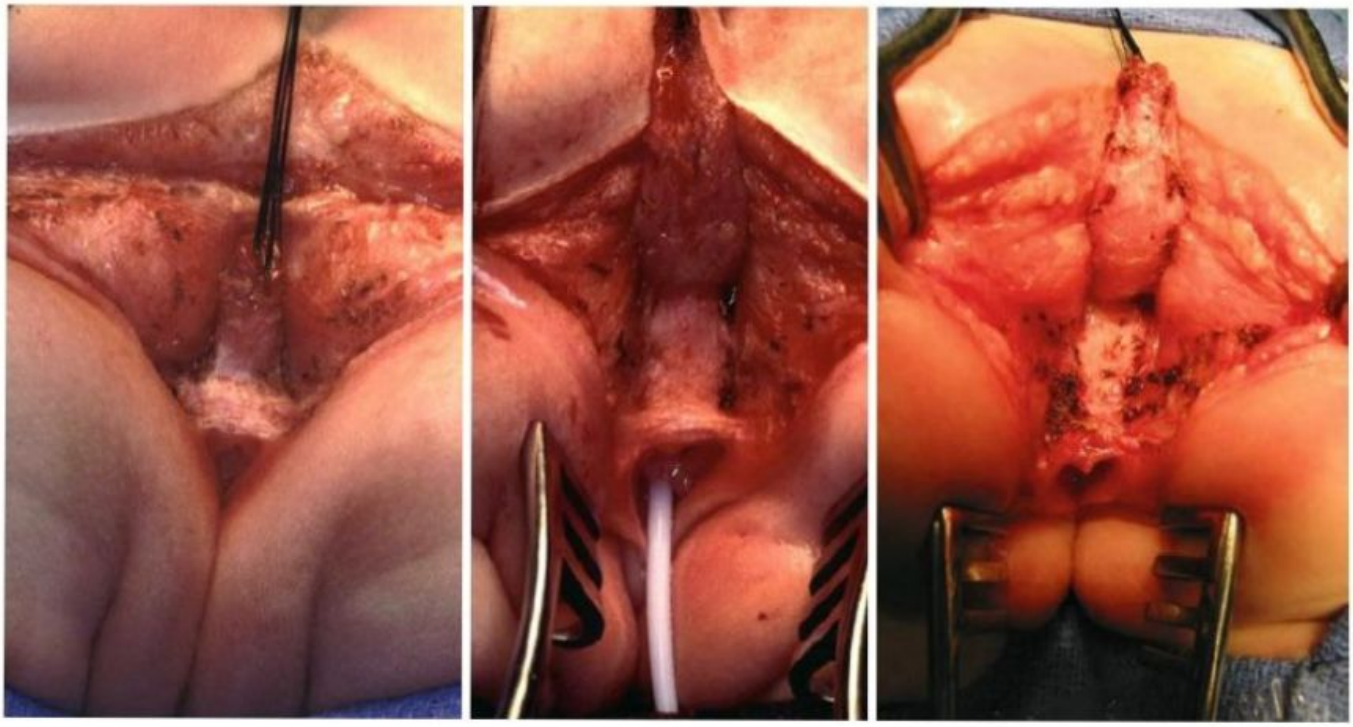

Fig. 15.14 Different stages of the separation of the rectum from the vagina. Posterior vaginal wall and anterior rectal wall intact

starts getting thicker, which indicates that we are getting close to the point where both are expected to be completely separated and have a full thickness. At this time, the surgeon should not be overconfident, because in that point he could injure either the rectum or the vagina (Fig. 15.14). The dissection must continue until the rectum has been completely separated from the vagina (Fig. 15.14).

It is extremely common for surgeons to ask what happens and what to do in the event of accidentally opening either the vagina or the rectum. Our routine answer is as follows: if it happens that we opened the vaginal wall, but maintained intact the rectal wall, one can actually leave the vaginal orifice of the injury open, provided the rectum is intact, and the anoplasty is not under tension, and the patient is going to do alright. Something similar can be said when the orifice is created in the rectum, but the vaginal wall is intact. What is considered nonacceptable is to have an injury of the rectal wall in front of an injury to the vaginal wall, leaving sutures in front of sutures, since that is considered an obvious predisposing factor for the formation of a rectovaginal fistula. Under such circumstances (vaginal injury and a rectal injury), one must continue the dissection of the rectum until we can leave a normal rectal wall in front of the vaginal orifice or suture.

We must always remember that in dealing with anorectal malformations, the real challenge in the surgical repair is represented by the separation of the structures, namely, the rectum from vagina, the rectum from urethra, and the vagina from urethra, because all those structures share a common wall without a plane of dissection. Most of the complications that we have seen in patients who underwent failed attempted repairs of anorectal malformations occur during the separation of these structures.

Sometimes when the rectum has been fully separated from the vagina, we find that we have enough rectal length to do an anoplasty without tension and with good blood supply. However, many other times, the rectum needs further mobilization. To do this, one must continue applying uniform traction on the multiple silk stitches. By doing this, it becomes evident that there are some bands and vessels holding the rectum up in the pelvis. These must be separated from the rectum, independently burned and divided in a circumferential manner, continuing until we have enough rectal length to create an anastomosis without tension.

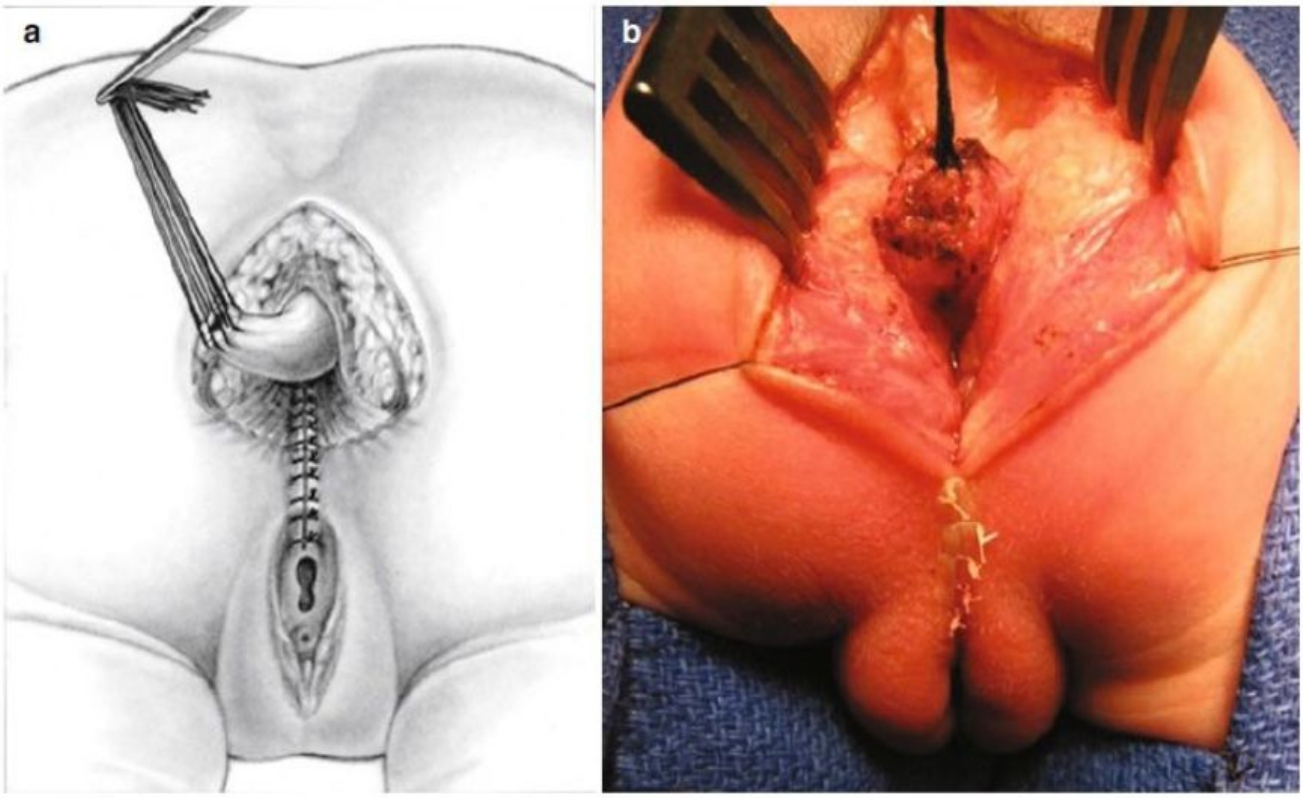

Fig. 15.15 Perineal body reconstructed. (**a**) Diagram. (**b**) Intraoperative diagram

The incision required to repair rectovestibular fistulas includes the opening of the muscle complex and part of the levator mechanism. Sometimes, it is not necessary to open completely the levator mechanism, and therefore we call this a limited posterior sagittal anorectoplasty. However, we are convinced that the size of incision does not affect, in any way, the future functional prognosis, provided all of the other important surgical steps are done correctly.

Once the rectum has been separated from the vagina and mobilized, in preparation for the reconstruction, the limits of the sphincter are electrically determined and marked with temporary silk stitches. The goal at this stage is to bring together the anterior limits of the sphincter and by doing that to reconstruct the perineal body of the patient (Fig. 15.15). This is the space that separates the vagina from the rectum. It is extremely important to use strong sutures (5-0 or 4-0 long-term absorbable sutures depending on the patient's age) to approximate both sides of the perineal body. There, we usually find a fibrous tissue that surrounded the original vestibular fistula. We use this tissue to anchor our stitches. These deep stitches must relieve most of the tension of the perineal body to be sure that the skin edges in the perineal body come together with no tension. We close the skin of the perineal body with 6-0 Vicryl sutures, only to be sure that the edges of the skin have come together, but those sutures hold no tension. Figure 15.15 shows the repaired perineal body. The rectum then is located within the limits of the sphincter immediately behind the perineal body. The posterior edges of the muscle complex and levator are sutured together in the midline using 5-0 long-term absorbable sutures, including a bite to the posterior rectal wall to anchor it in normal location (Fig. 15.16). These stitches are aimed to avoid retraction and prolapse. The ischiorectal fossa, as well as the subcutaneous tissue, is obliterated using 5-0 long-term absorbable sutures, and the skin is closed either with subcuticular 5-0 monofilament, absorbable, or interrupted 6-0 long-term absorbable sutures.

The anoplasty is done as previously described, using two layers of interrupted 5-0 or 6-0 long-term absorbable sutures (Fig. 15.17). We try to trim off as little as possible rectal tissue, but we do not hesitate to remove all of the tissue that is

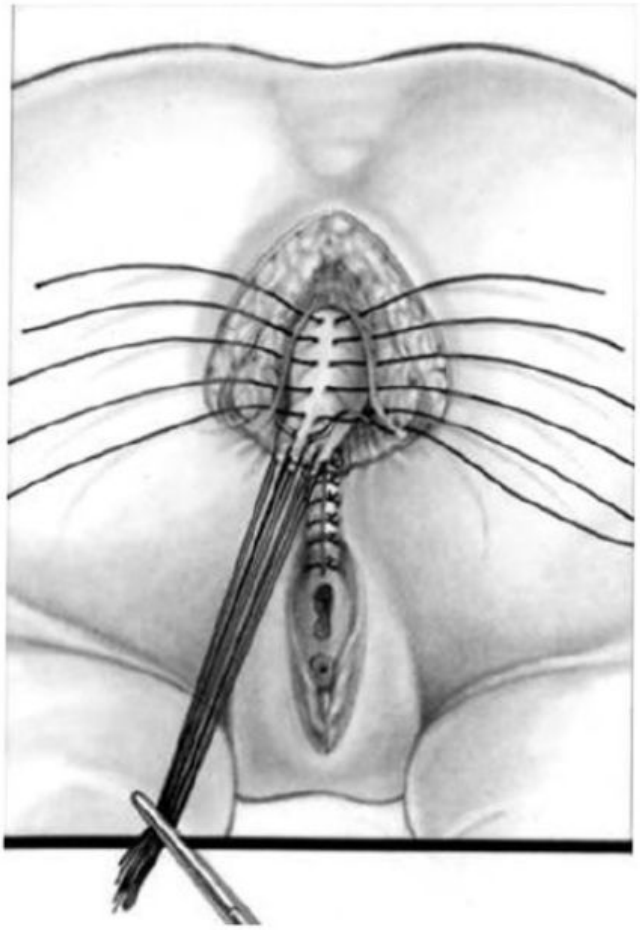

Fig. 15.16 Diagram showing sutures taking the posterior edges of the muscle complex, including a bite to the posterior rectal wall to anchor it

considered damaged, to be sure that we have healthy rectal tissue with good blood supply to create a healthy anoplasty (Fig. 15.18).

Patients operated without a colostomy are kept on parenteral nutrition with nothing by mouth for a period not shorter than 7 days. After 7 days, we look into the external aspect of the perineum, and if it looks that it has healed nicely, we allow the patient to eat and discharge her. Anal dilatations start 2 weeks after surgery following our protocol (see Chap. 18). Occasionally, when one examines the patient's perineum 1 week after the operation, one may find that there is an area of partial dehiscence of the perineal body or the posterior sagittal incision, and this is a good opportunity to take the patient to the operating room and resuture that, taking advantage of the fact that the patient has been with nothing by mouth. Under those circumstances we keep the patient 2 or 3 more days fasting, receiving parenteral nutrition.

At the time of colostomy closure (in those patients with a colostomy) 2 or 3 months after the operation, the external aspect of the anus and the vagina, as well as the perineum in these patients, is remarkably normal looking (Fig. 15.19).

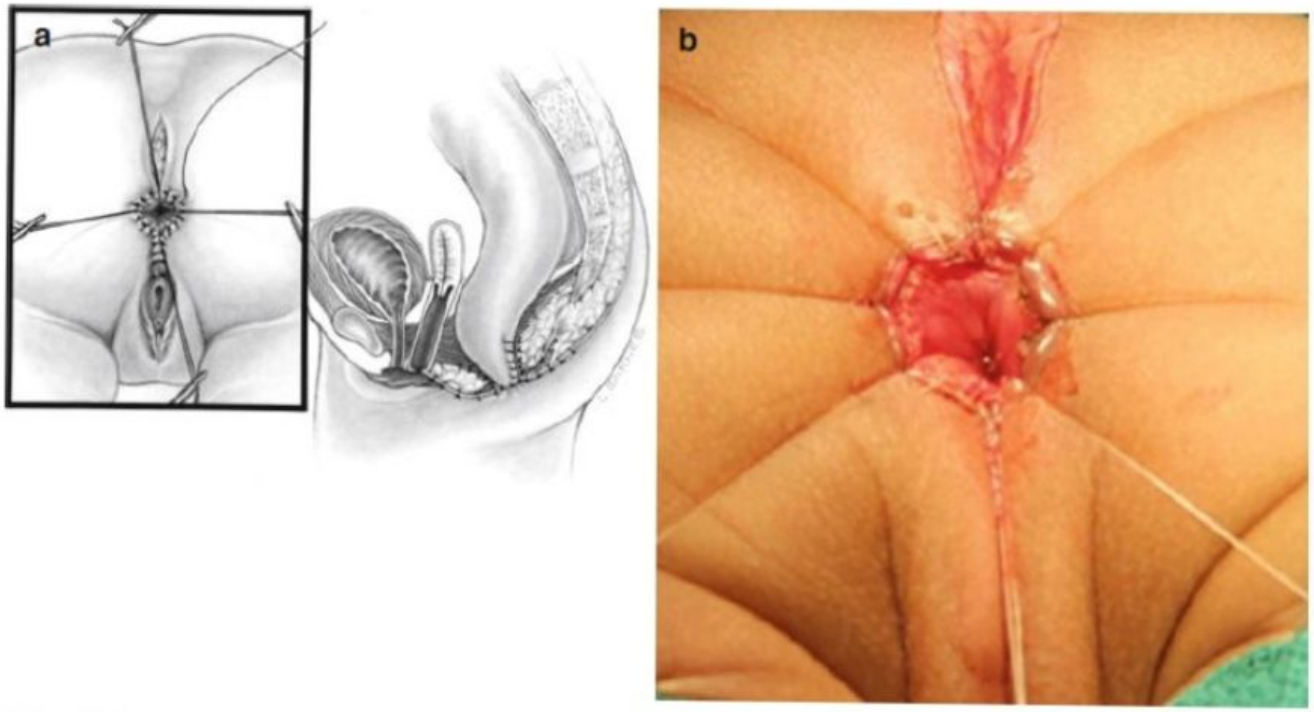

Fig. 15.17 Anoplasty and wound closed. (**a**) Diagram. (**b**) Intraoperative picture

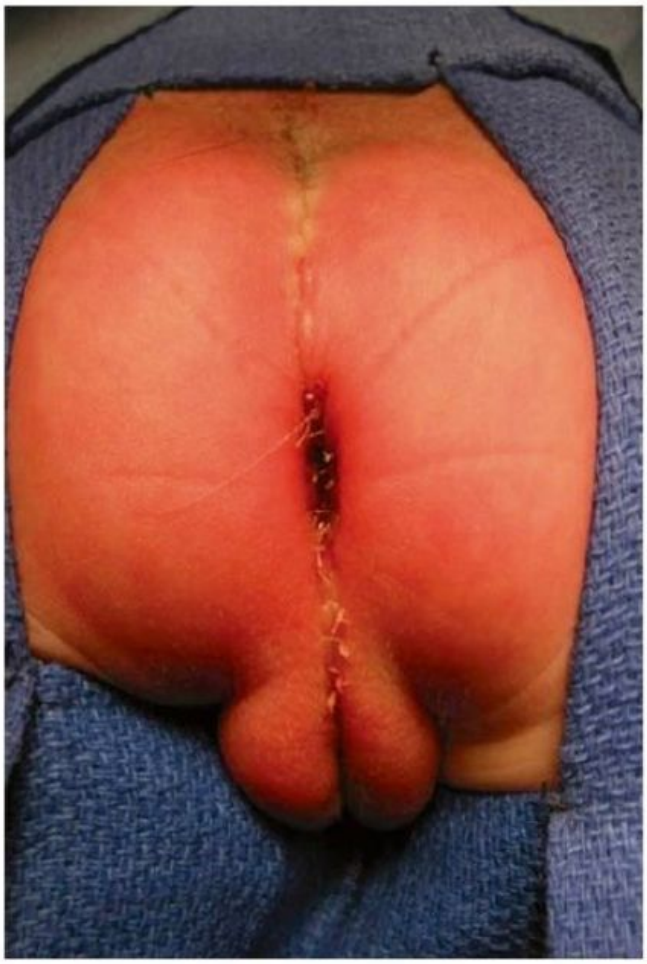

Fig. 15.18 Photograph of a finished operation

15.6 Complications

Five of our patients suffered from a dehiscence requiring a reoperation.

We have seen patients born with vestibular fistula that underwent a "laparoscopic repair." It must be very difficult for the surgeon to work in this common wall through the abdomen with a laparoscope. What they rather have done is to amputate the rectum at a "convenient" location, leaving the distal piece of the rectum attached to the vagina. We consider this an inappropriate way of management.

15.7 Functional Results

Ninety percent of our patients have voluntary bowel movements by the age of three. This is, provided they have a normal sacrum, not tethered cord, and they had received a good operation. Over 50 % of the patients suffer from significant constipation that deserves special attention. Not taking good care of the constipation will provoke chronic fecal impaction and overflow

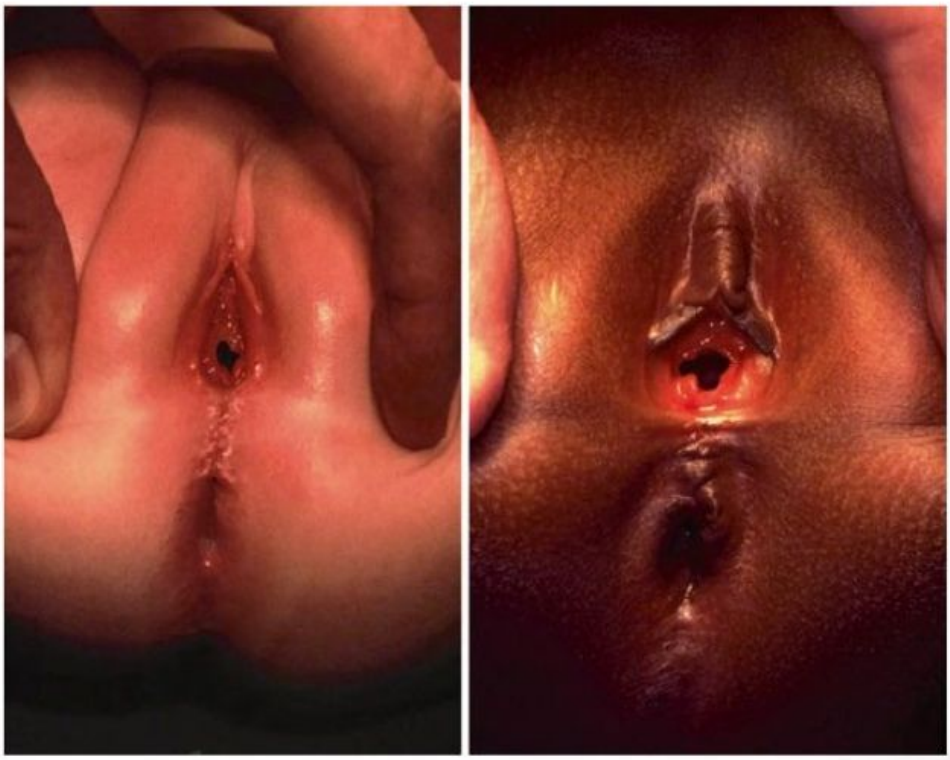

Fig. 15.19 External appearance 2–3 months postoperatively

pseudoincontinence. We usually have a long conversation with the parents and explain in detail the importance of taking care of the constipation. We emphasize the fact that the constipation that these patients suffer from is much more severe than the common idiopathic constipation of the general pediatric population. The amount of laxatives that these patients need sometimes is two, three, four, or five times higher than in other types of patients. We try to make the parents paranoid against the problem of constipation. We also emphasize the fact that the amount of laxatives that these patients require to empty the colon every day cannot be predicted. We determine the amount of laxatives by trial and error over a period of several days, taking abdominal x-ray films to be sure that the patient empties the colon. If the patients are receiving breast feedings at the time of our operation, most likely they will not need laxatives until they start decreasing the amount of breast milk and receiving another type of formula.

Sexual life in these patients is normal, and as we have seen, many of our patients are becoming adults and are getting married. They also can deliver babies vaginally, since we did not actually injure the vagina which preserves a normal elasticity in most of its circumference, since we only dissected the posterior vaginal wall.

In the past, some surgeons [2] claimed that these patients could have a normal life without an operation or simply doing a "cutback" type of procedure to enlarge the anal opening. We have seen that this is not true. First of all, the bowel control under those circumstances is rather poor. In addition, when these patients grow up, they feel very unhappy about the fact that they have the anus located immediately behind the vagina with no perineal body. This gives them insecurity and psychological problems, and in addition, a vaginal delivery is contraindicated, because it will produce severe rectal damage.

15.8 Reoperations in Patients with Vestibular Fistula

From all anorectal defects treated by us, it is the vestibular fistula type of case that most frequently came to us after a failed attempted repair at another institution. In fact, from our total series of 290 patients with vestibular fistula, 73 of them are reoperations. We believe that this is a reflection of the fact that surgeons in general probably underestimate the complexity of the repair of this defect. As previously mentioned, vestibular fistula is by far the most common anorectal malformation seen in females. The functional prognosis in girls when they are born with a good sacrum, have no tethered cord, and receive a good operation is excellent. Unfortunately, patients who underwent a failed attempted repair followed by a reoperation do not have the same good functional prognosis. Eighty percent of them have voluntary bowel movements as compared to 90 % for those operated primarily.

Probably, the surgeons find it relatively easy to imagine that the orifice of the rectum located in the vestibule could easily be moved back to the normal location of the anus. In reality, the repair of this malformation is a delicate and technically demanding procedure.

The most common scenario in dealing with reoperations for vestibular fistulas is a patient that was operated without a protective colostomy and soon after suffered from dehiscence and retraction of the rectum, followed by opening of the rectum into the posterior vaginal wall. In other words, the original malformation was a vestibular fistula, but the patient comes with a real acquired rectovaginal fistula secondary to a poor initial operation. During the re-exploration, our most common finding in this specific type of problem has been an intact common wall between the rectum and vagina. In other words, the surgeons try to repair the malformation but failed to separate the rectum completely from the vaginal wall. They still tried to pull the rectum down which was left, we think, under tension, because it was still attached to the vaginal wall. As a consequence, the rectum retracted. We assume that during the attempt to separate the rectum from the vagina, the lower part of the vaginal wall was injured, and therefore when the rectum retracted, it reopened into the posterior vaginal wall creating an acquired vaginal fistula.

Another common scenario in reoperations for vestibular fistula is a group of patients that underwent a previous operation called cutback

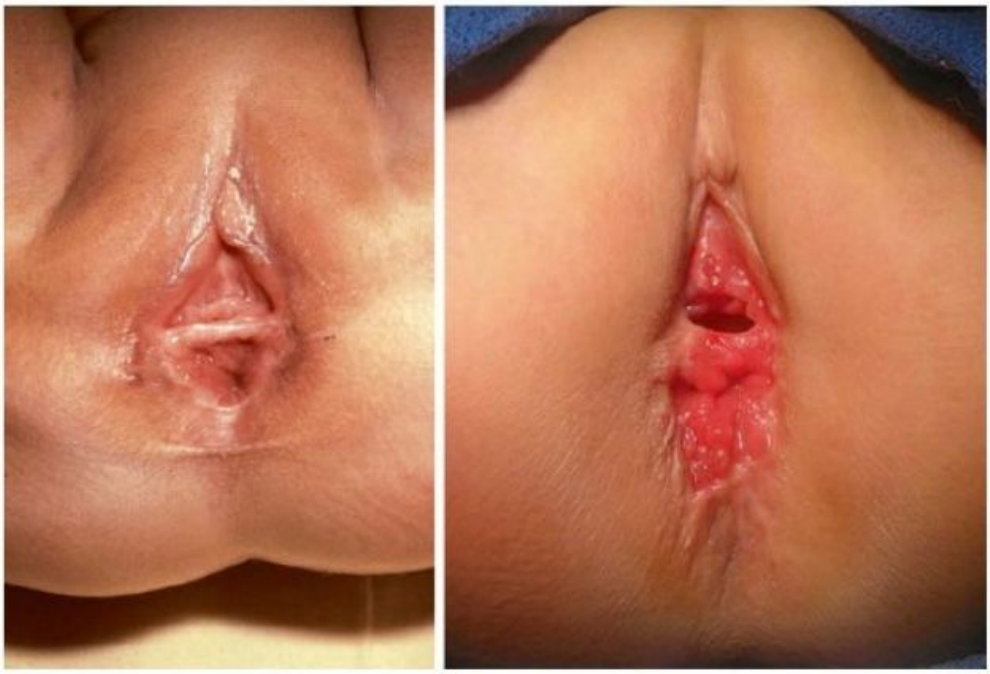

Fig. 15.20 Pictures of two patients born with a vestibular fistula and underwent a cutback procedure prior to coming to our center

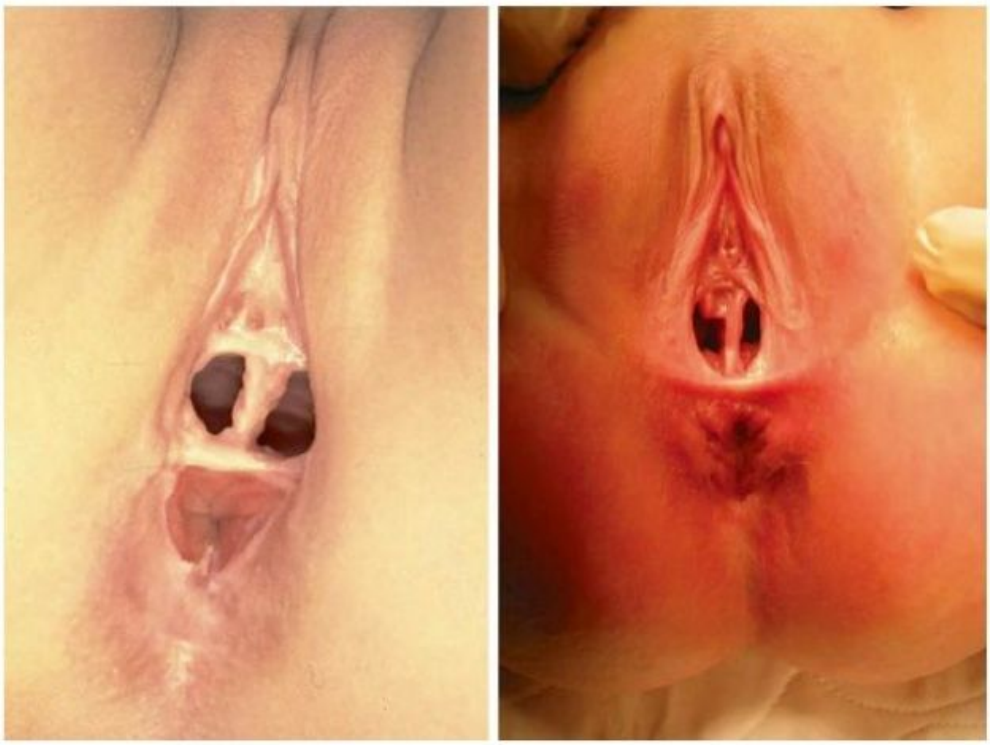

Fig. 15.21 External aspect of perinea of two patients born with a vestibular fistula and two hemivaginas. They underwent a poor attempted repair and were left with no perineal body and two hemivaginas

procedure at another institution [39, 40]; these consisted in making a posterior slit in the posterior edge of the anal opening in the vestibule and suturing it horizontally like a Heineke-Mikulicz type of procedure. That procedure only enlarges the anal opening and leaves the rectum attached to the vaginal wall with no perineal body (Fig. 15.20). We believe that, perhaps, in cases of perineal fistula, the cutback procedure could be considered an acceptable therapeutic alternative, but we strongly believe that this type of operation is contraindicated in patients with vestibular fistula. There was an old belief that went from generation to generation that by leaving the rectum attached to the vagina, as time went by, the perineal body would grow, which is definitely not true.

Another finding that is interesting to mention is the fact that in some of these patients, we found that they had two hemivaginas, and such malformation was never mentioned in the operative reports of the previous surgeons (Fig. 15.21). Again, we like to say that "our eyes see only what our mind suspects."

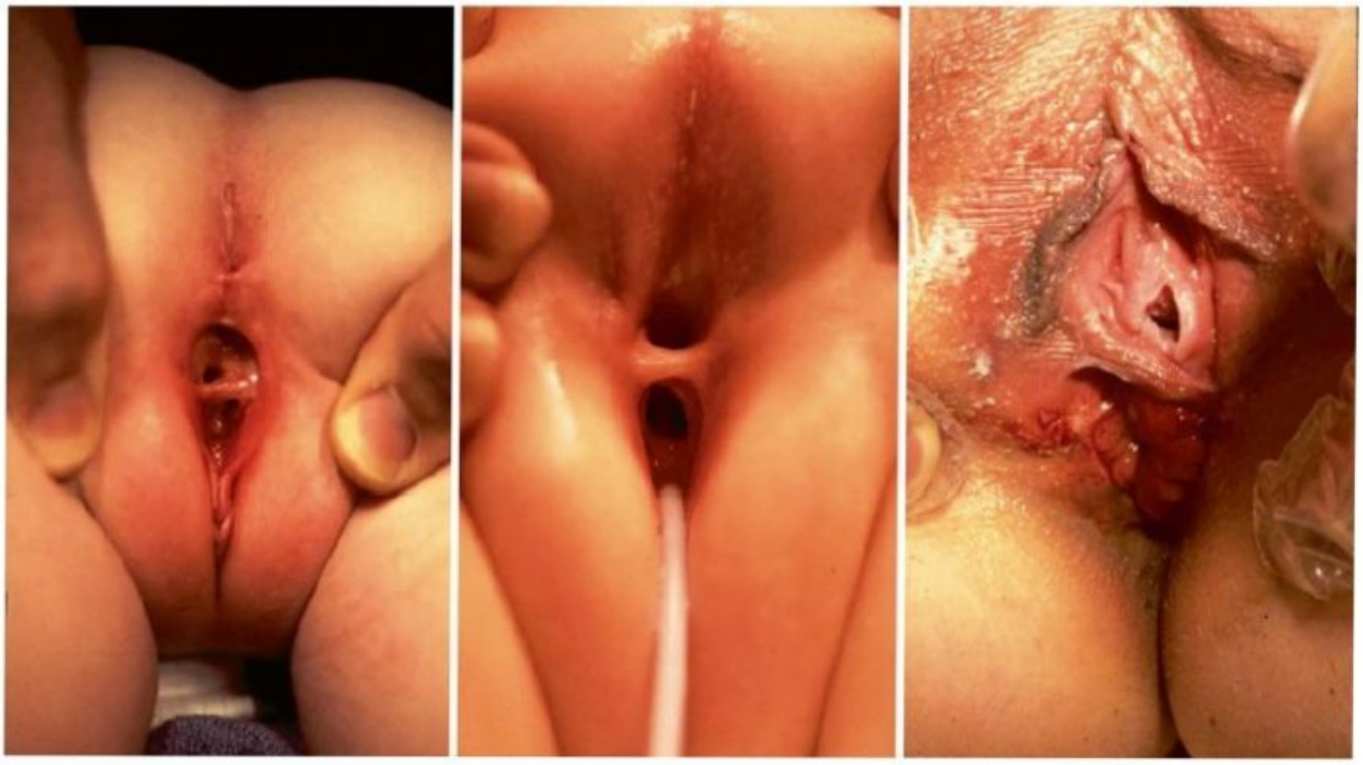

Fig. 15.22 External appearance of the perineum of different patients referred to us, after failed attempted repairs

15.9 Surgical Technique

Reoperations for recurrent or dehiscent, retracted vestibular fistulas are currently done by us without a protective colostomy. Figure 15.22 shows examples of cases that came to us after a failed attempted repair of their malformation. However, we follow the precautions already mentioned in the chapter related to bowel preparation. We take the baby to the operating room with the bowel completely clean. As part of our routine, we perform vaginoscopy and cystoscopy to rule out the presence of associated defects (mainly vaginal septum). We place the patient in prone position with the pelvis elevated and make a posterior sagittal incision following the specifications already described. Multiple 5-0 silk stitches are placed at the mucocutaneous junction of the rectovaginal fistula or the rectal opening in order to apply uniform traction. Through the posterior sagittal incision, all structures are divided in the midline until the posterior rectal wall is identified and then the dissection of the rectum proceeds, first on the lateral walls and eventually in the common wall between the rectum and the vagina. As previously mentioned, we have been impressed by the fact that most of these patients have an intact common wall between the rectum and vagina, which reflects the fact that the surgeons did an incomplete mobilization of the rectum. We go ahead and make two walls out of one. In other words, we separate the rectum from the vagina as previously described in the primary procedure. We must suture the defect of the posterior vaginal wall. Once the rectum has been completely separated, we then mobilize the rectum enough to guarantee that an intact anterior rectal wall is left in front of the vaginal sutures. We are convinced that the vaginal defect can even be left unsutured, and it will heal normally provided the rectal wall left behind is intact. We dissect the rectum enough to guarantee that the rectal wall in front of the vagina is completely normal and also to be sure that the anastomosis between the rectum and the skin of the anal dimple is performed without tension. Before we do the anoplasty, we repair the posterior vaginal wall with long-term absorbable sutures, determine the limits of the sphincter, and continue the operation as described for primary cases, reconstructing the perineal body and doing the anoplasty. The patients remain 10 days fasting and receiving parenteral nutrition.

We were impressed by the fact that many patients had a failed operation early in their life, remained incontinent during childhood, and searched for help only when they became teenagers and had decided to become sexually active. We believe that they had become aware of their defective anatomy and felt very upset about the fact that their rectum and vagina were located one next to the other, with no perineal body. In other words, they felt embarrassed at considering sexual life with that kind of anatomy.

15.10 Rectovestibular Fistula with Normal Anus

See Chap. 27, Sect. 27.2.

References

1. Cigarroa FG, Kim SH, Donahoe PK (1988) Imperforate anus with long but apparent low fistula in females. J Pediatr Surg 23(1 Pt 2):42–44
2. Stephens D, Smith D (1971) Chapter 4: Individual deformities in the female. In: Anorectal malformations in children, vol 4. Year Book Medical Publisher, Chicago, pp 81–117
3. Digray NC, Mengi Y, Goswamy HL, Singh N, Atri MR, Sharma R, Thappa DR (2001) Complete vaginal prolapse: an unusual presentation of anovestibular fistula. Pediatr Surg Int 17(2–3):226–227
4. Banu T, Hannan MJ, Aziz MA, Hoque M, Laila K (2006) Rectovestibular fistula with vaginal malformations. Pediatr Surg Int 22(3):263–266
5. Levitt MA, Bischoff A, Breech L, Peña A (2009) Rectovestibular fistula–rarely recognized associated gynecologic anomalies. J Pediatr Surg 44(6):1261–1267. doi:10.1016/j.jpedsurg.2009.02.046
6. Hanley PH, Hines MO, Stephens JE (1954) Anal sphincter-preserving operation for congenital low rectovaginal or rectoperineal fistula. Am J Surg 88(5):737–745
7. Stone HB (1936) Imperforate anus with rectovaginal cloaca. Ann Surg 104(4):651–661
8. David VC (1937) The treatment of congenital openings of the rectum into the vagina—atresia ani vaginalis. Surgery 1(2):163–168
9. Donovan EJ, Stanley-Brown EG (1958) Imperforate anus. Ann Surg 147(2):203–213
10. Rosenblatt MS, Gustavson RG (1958) The treatment of congenital malformations of the anus and rectum. Am J Surg 96(2):343–350
11. Patil UB, Kavouksorian JK (1980) Unusual imperforate anus. N Y State J Med 80(1):87–88
12. Aluwihare AP (1990) Primary perineal rectovaginoanoplasty for supralevator imperforate anus in female neonates. J Pediatr Surg 25(2):278–281
13. Simmang CL, Paquette E, Tapper D, Holland R (1997) Posterior sagittal anorectoplasty: primary repair of a rectovaginal fistula in an adult: report of a case. Dis Colon Rectum 40(9):1119–1123
14. Duhamel B (1960) Le Traitement des anus vulvaires. Ann Chir Infant 1:53–70
15. Bill AH, Hall DG, Johnson RJ (1975) Position of rectal fistula in relation to the hymen in 46 girls with imperforate anus. J Pediatr Surg 10(3):361–365
16. Salamov KN, Dultsev YV, Protsenko VM (1987) Surgical treatment of the vestibular ectopia ani in adults. Acta Chir Plast 29(4):209–215
17. Heinen DFL, Bailez M, Solana J (1992) Malformaciones anorectales I. Fístula vestibular. Area Cirugía. Hospital de Pediatría J.P. Garrahan, Buenos Aires, pp 148–154
18. Sawicka E (1995) Results of surgical treatment of girls with ano-vestibular fistula. Surg Childh Intern 3(2):94–98
19. Heinen FL (1997) The surgical treatment of low anal defects and vestibular fistulas. Semin Pediatr Surg 6(4):204–216
20. Javid PJ, Barnhart DC, Hirschl RB, Coran AG, Harmon CM (1998) Immediate and long-term results of surgical management of low imperforate anus in girls. J Pediatr Surg 33(2):198–203
21. Martín RS, Molina E, Cerdá J, Estellés C, Casillas MAG, Romero R, Vázquez J (2002) Manejo del ano vestibular en niñas mayors. Cir Pediatr 15:140–144
22. Rosen NG, Hong AR, Soffer SZ, Rodriguez G, Peña A (2002) Rectovaginal fistula: a common diagnostic error with significant consequences in girls with anorectal malformations. J Pediatr Surg 37(7):961–965
23. Demirbilek S, Atayurt HF (1999) Anal transposition without colostomy: functional results and complications. Pediatr Surg Int 15(3–4):221–223
24. Upadhyaya VD, Gopal SC, Gupta DK, Gangopadhyaya AN, Sharma SP, Kumar V (2007) Single stage repair of anovestibular fistula in neonate. Pediatr Surg Int 23(8):737–740
25. Kumar B, Kandpal DK, Sharma SB, Agrawal LD, Jhamariya VN (2008) Single-stage repair of vestibular and perineal fistulae without colostomy. J Pediatr Surg 43(10):1848–1852. doi:10.1016/j.jpedsurg.2008.03.047
26. Menon P, Rao KL (2007) Primary anorectoplasty in females with common anorectal malformations without colostomy. J Pediatr Surg 42(6):1103–1106
27. Upadhyaya VD, Gangopadhyay AN, Pandey A, Kumar V, Sharma SP, Gopal SC, Gupta DK, Upadhyaya A (2008) Single-stage repair for rectovestibular fistula without opening the fourchette. J Pediatr Surg 43(4):775–779. doi:10.1016/j.jpedsurg.2007.11.038