# Cloaca, Posterior Cloaca and Absent Penis Spectrum

# 16

## 16.1 Cloaca

### 16.1.1 Definition and Management

A cloaca is a malformation that affects the rectum and urogenital tract in females. These girls are born with a single perineal orifice. The vagina, urethra, and rectum are fused together inside the pelvis, creating a single common channel that opens into a single orifice in the location where the urethra normally opens (Fig. 16.1). The length of the common channel varies from case to case, from 1 to about 10 cm with an average of approximately 3 cm.

Thirty percent of these patients suffer in addition from a very dilated vagina full of fluid and/or mucus, called hydrocolpos [1] (Fig. 16.2). The reason why these very dilated vaginas retain fluid remains a mystery, since they are never really atretic. We speculate that there must be some sort of valve mechanism that interferes with the emptying of the fluid. Most of the patients with hydrocolpos, in addition, have duplicate Müllerian systems (Fig. 16.3).

The hydrocolpos may produce two important complications:

(a) The first is the possibility of compressing the trigone of the bladder, producing an extrinsic ureterovesical obstruction, megaureter, and hydronephrosis.
(b) The second possibility is that the hydrocolpos, left undrained, may become infected; creating a pyocolpos that eventually may perforate, which is a catastrophic event with risk of death. In addition, the resulting inflammation may scar the vagina and impact the future reconstruction.

Approximately 60 % of the patients with cloacas also have a double Müllerian system consisting of the presence of two hemiuteri and two hemivaginas [1]. This septation disorder may be partial or total. In addition, it can be symmetric or asymmetric. In the asymmetric types, the double Müllerian system phenomenon is frequently associated with a unilateral atresia of the Müllerian structure. When this goes unrecognized, it may produce an accumulation of menstrual blood at the age of puberty, as well as retrograde menstruation into the peritoneal cavity (Fig. 16.4) which produces rather dramatic signs of an acute abdomen and requires an emergency laparotomy. The presence of double Müllerian systems also has important potential obstetric implications that will be discussed later in this chapter.

Cloacas represent a very wide spectrum of defects, but the common denominator is the presence of a single perineal orifice. On the very bad side of the spectrum, one may find patients with a

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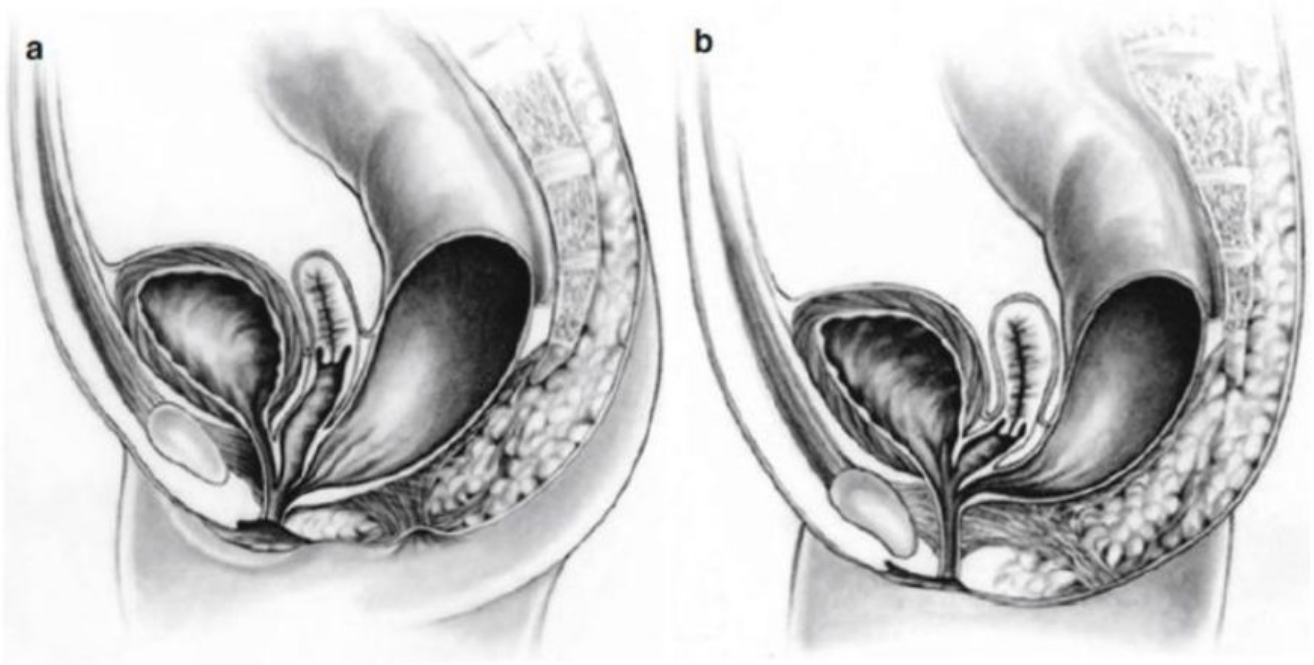


Fig. 16.1 Diagram of a cloaca. (a) Short common channel. (b) Long common channel

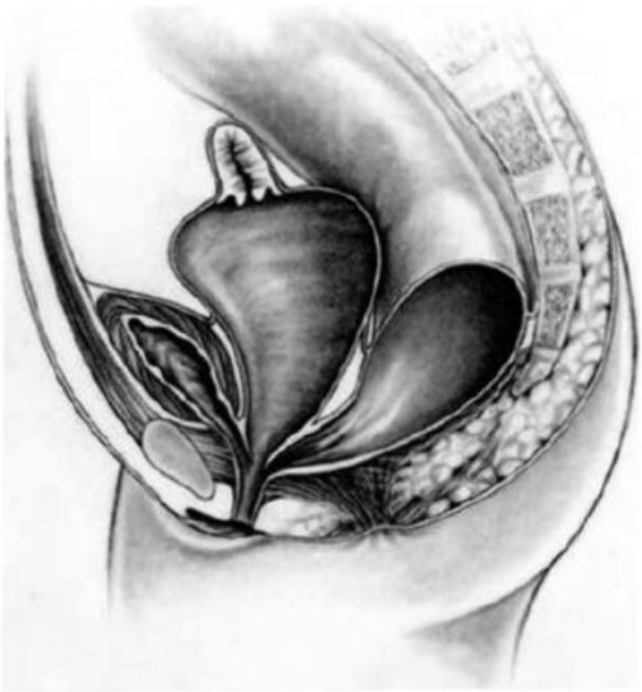

Fig. 16.2 Diagram of a cloaca with hydrocolpos

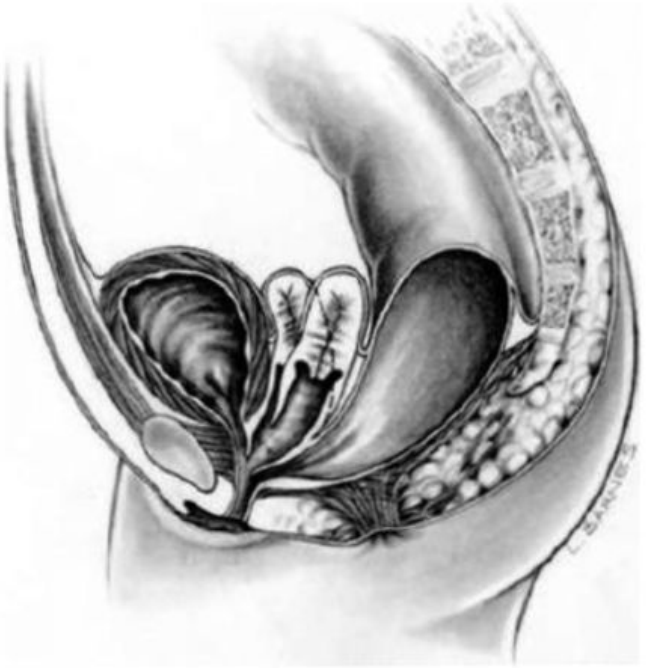

Fig. 16.3 Diagram showing a cloaca with two hemivaginas

common channel as long as 10 cm; in such cases, usually two little hemivaginas, as well as the rectum, connect to the urinary tract at the bladder neck or above the bladder neck (at the trigone) (Fig. 16.5).

We are far from knowing the genetic causes of this condition [2]. Yet, we have never seen two cases of cloaca in the same family.

The knowledge of the intrinsic anatomic characteristics of this malformation is relatively new. We were able to detect and read old publications that we think described patients suffering from cloacas, although were not recognized as such [3–6].

We were also very impressed by the fact that most publications prior to 1982 reported high numbers of rectovaginal fistula cases and very few cloacas [7]. In retrospect, we are convinced that the authors were reporting patients suffering

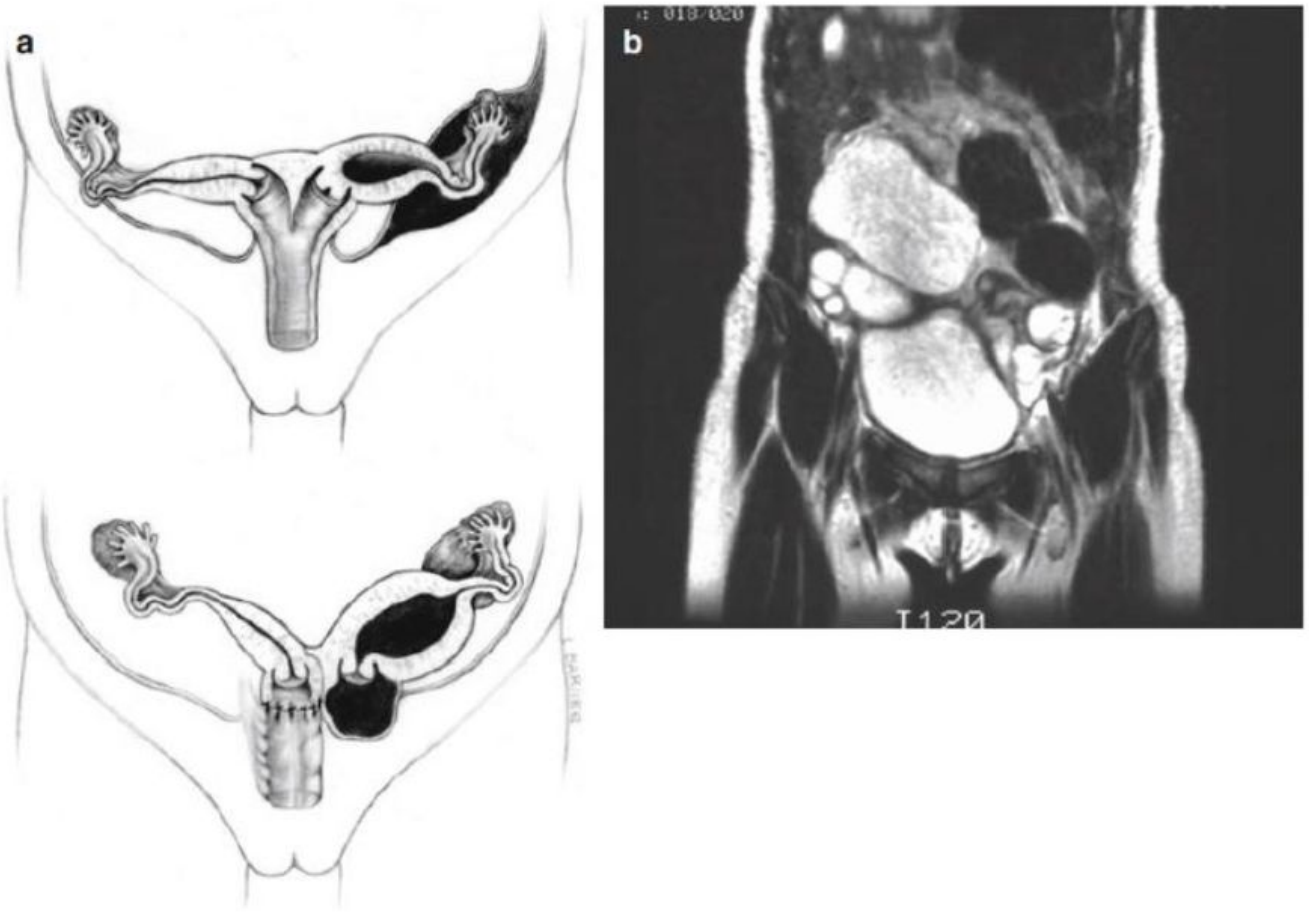


**Fig. 16.4** Accumulation of menstrual blood in a patient with obstruction of the Müllerian structures. (a) Diagram. (b) MRI

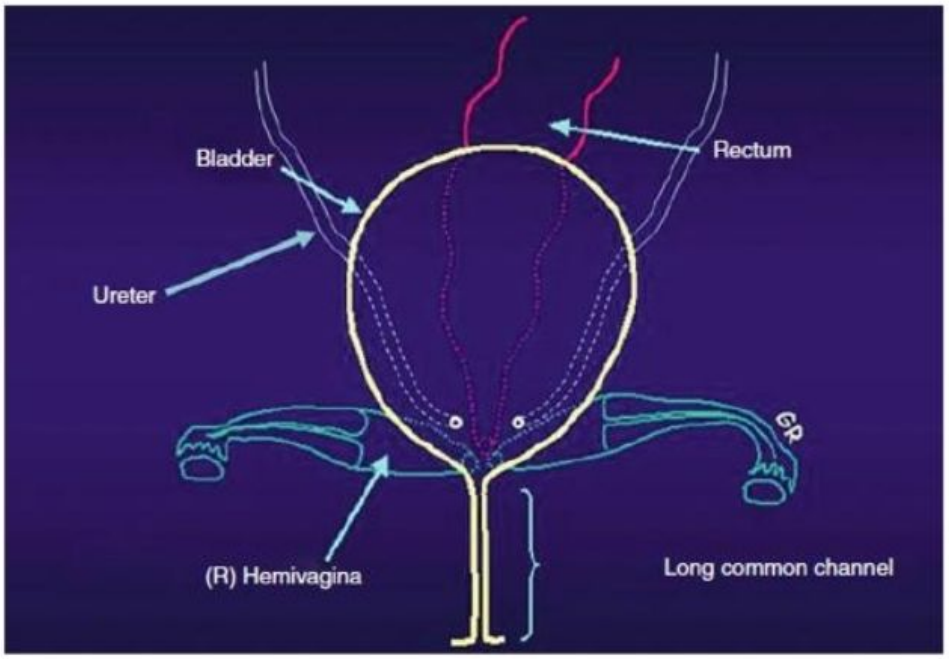


**Fig. 16.5** Diagram showing a cloaca with a very long common channel

from cloacas as "vaginal fistulas." We believe that because of the large number of cases of cloacas that we have seen, coming with a history of suffering from a "rectovaginal fistula," yet, when we examined them, we found an untouched persistent urogenital sinus and a pulled-down rectum; in the medical records of those cases, the word cloaca is not present.

Most publications prior to 1982 reported very few cases of cloacas; many of them were

autopsy findings. The cases that underwent an attempted repair suffered from a high mortality. The treatments used include a colostomy at birth, followed by a rectal pull-through, leaving the patient with a urogenital sinus to be repaired "later" [8–23].

The terminology used in those years was also confusing. The authors frequently published series that included cloacas and other different conditions such as "adrenal hyperplasia," "vaginal atresia," and "high anorectal malformation." One particular publication from 1973 [17] is the most prominent one because it proposes the full repair of the vagina. However, looking at the diagrams, it becomes clear that the technique used by Dr. Raffensperger, the author, may be applicable only in cases with a relatively large vagina, located very low.

All pediatric surgeons as well as hundreds of patients are in debt with Dr. Hardy Hendren for his contributions in the field of pediatric surgery and pediatric urology. His seminal work on the surgical management of cloacas is the most important one that we found in our literature review [24–32]. In his initial publications, Dr. Hendren referred to this malformation as "urogenital sinus and anorectal malformation" [25]. Dr. Hendren's contribution was particularly important in dealing with complex reoperations and repairing the challenging urologic-associated defects of these patients.

Some authors refer to cloacas with a clear embryologic bias, and therefore, they used rather confusing terms such as "urorectal septum malformation sequence" [33] or "urorectal septal defects" [34] including variants such as adrenal hyperplasia, as well as male cases [35]. Others include the cloacas as part of "Müllerian duct anomalies" [36].

### 16.1.1.1 Associated Defects

Twenty percent of those patients with a common channel longer than 3 cm had an absent kidney. When the common channel was shorter than 3 cm, 17 % of the patients suffered from this anomaly. Hydronephrosis occurred in 45 and 22 %, respectively, in patients with a common channel longer or shorter than 3 cm.

**Table 16.1** Correlation between sacral ratio and length common channel

| Length of common channel | Sacral ratio | | |
|---|---|---|---|
| | Lateral film | Anterior/ posterior film | Average |
| Less than 3 cm | 0.68 | 0.55 | 0.615 |
| More than 3 cm | 0.6 | 0.53 | 0.56 |

It is important to notice that in all other anorectal malformations, absent kidney is the most common anatomic-associated anomaly. The high incidence of hydronephrosis in this malformation is consistent with the fact that the most serious problems that these patients will suffer from (including death) are urologic.

Vesicoureteral reflux occurs in 40 and 21 %, respectively, in patients with common channel longer and shorter than 3 cm.

Most patients with hydronephrosis suffered from vesicoureteral reflux. At birth, however, some patients with hydronephrosis and megaureter seemed to suffer from a ureterovesical obstruction. In reality, the obstruction was an extrinsic one, caused by a tense hydrocolpos. When the hydrocolpos was drained, the vesicoureteral reflux becomes obvious.

Hemivertebra occurs in 13 % of cases (lumbar, thoracic, cervical, and sacral). The average sacral ratio in patients with cloacas is 0.52 AP and 0.64 lateral. Table 16.1 shows the correlation between sacral ratio and the length of the common channel.

Cardiovascular anomalies occur in 20 % of cases in cloacas. Patent ductus arteriosus occurs in 8 % of cases, atrial septum defect in 19 % of cases, ventricular septum defect in 5 % of cases, and tetralogy of Fallot in 2 % of cases. Tethered cord occurs in 36 % of the patients with cloacas.

Esophageal atresia was present in 11 % of our cloacas and duodenal atresia in 3 %.

### 16.1.1.2 Goals of Treatment

The treatment of cloacas represents a significant technical challenge. The final goals of treatment must result in a patient with urinary control, bowel control, sexual function, and capacity to procreate. These goals, of course, are sometimes achieved, sometimes partially achieved, and

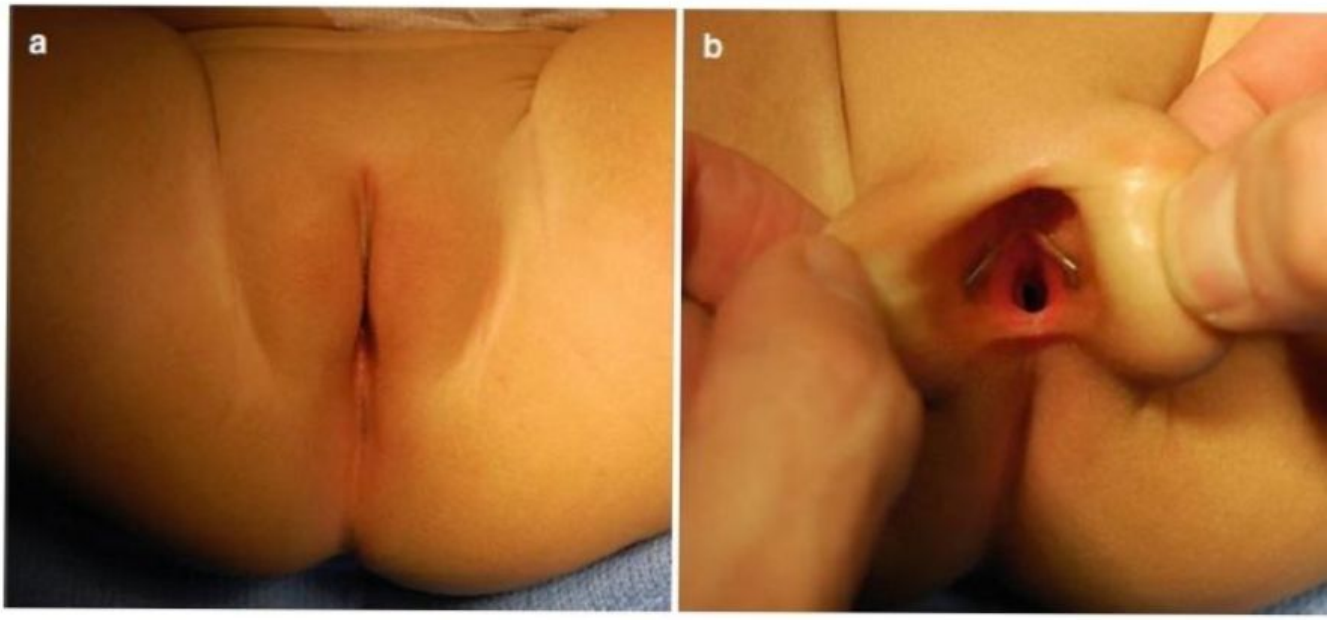


Fig. 16.6 Perineum of a patient with a cloaca. (a) Without separating the labia. (b) Separating the labia

sometimes not achieved at all. For the worst scenario, we are firm in our philosophy that all patients with anorectal and urogenital malformations should be clean of stool and dry of urine in the underwear after the age of three, either because they were born with a benign malformation that was adequately reconstructed or because even when they were born with a malformation with bad functional prognosis, the patient is maintained artificially clean of stool (subjected to a successful bowel management program) [see Chap. 20] and dry of urine (subjected to intermittent catheterization) through the native urethra or through a neourethra (continent diversion).

Since we are dealing with a spectrum of defects, one should expect a spectrum of results after the treatment.

### 16.1.1.3 Neonatal Management

The diagnosis of a cloaca is a clinical one. It is enough to look at the patient's perineum and make the correct diagnosis (Fig. 16.6). These patients have a single orifice, yet, the perineum has other important characteristics that help to predict the internal anatomy and the final functional prognosis. A "good-looking" perineum consists of the presence of a well-formed midline groove and a well-located and clear anal dimple, indicating that the patient has a good sphincter (Fig. 16.7). On the other hand, a "bad-looking"

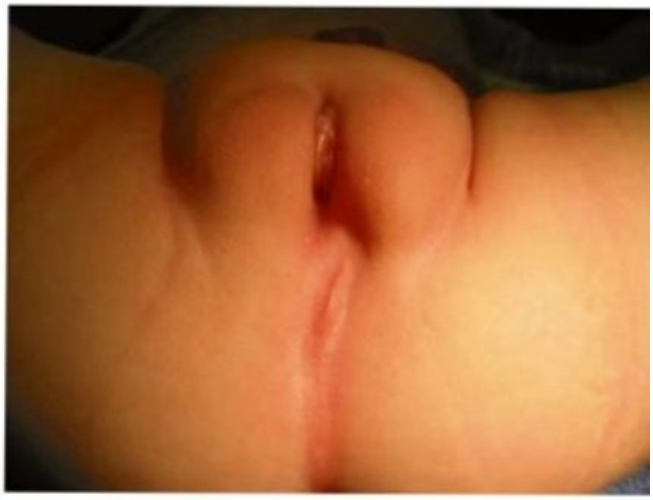

Fig. 16.7 "Good-looking perineum." Obvious midline groove and prominent anal dimple

perineum includes a single perineal orifice but, in addition, a completely "flat bottom" with no traces of sphincter mechanism (Fig. 16.8) and most likely, a poor functional prognosis. In between those extremes of the spectrum, one can find a variety of external appearances.

Occasionally, one can find a very large single perineal orifice, leaking urine, and with evidence of a mild separation of the pubic bones (Fig. 16.9). Those external signs correspond to a patient who has separated pubic bones and a malformation called covered cloacal exstrophy [37]. These patients have no bladder neck; their bladder is very small because it has never been

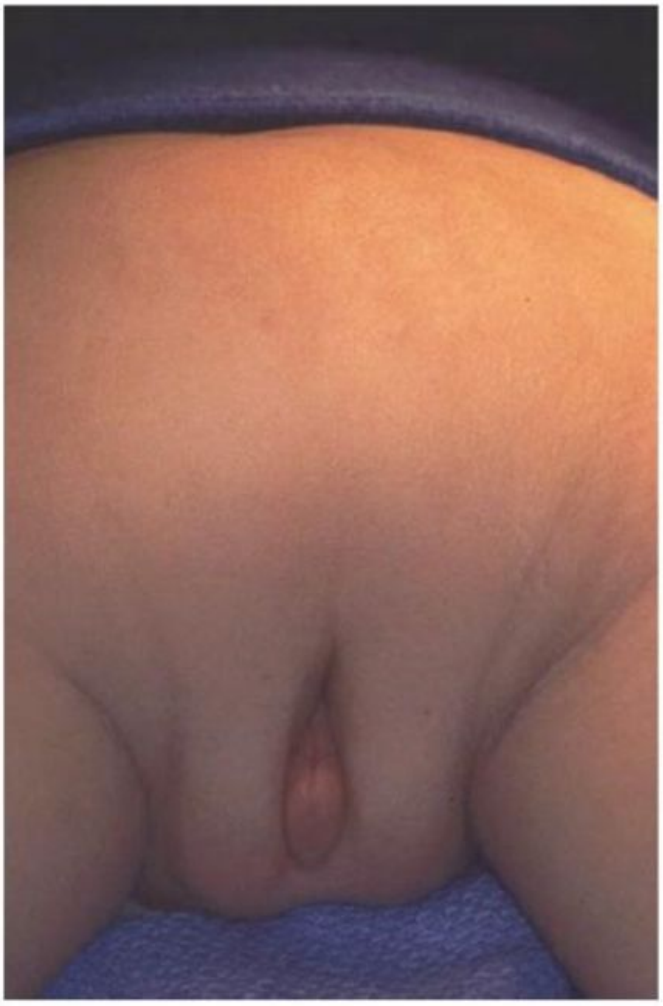

Fig. 16.8 "Bad-looking perineum." "Flat bottom," absence of midline groove, no evidence of anal dimple

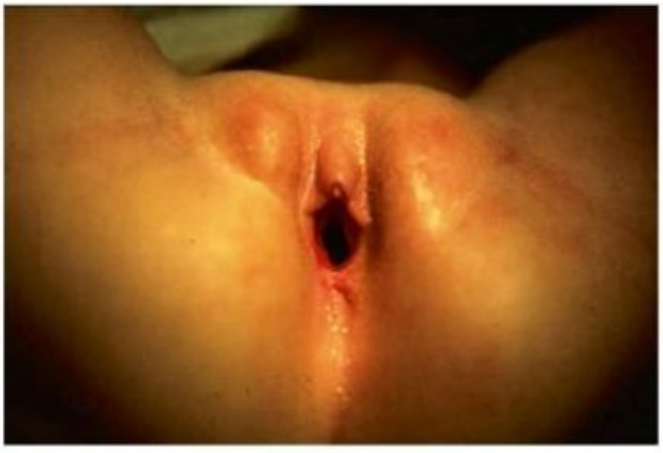

Fig. 16.9 Perineum of a patient with a large single perineal orifice. The pubic bones are separated

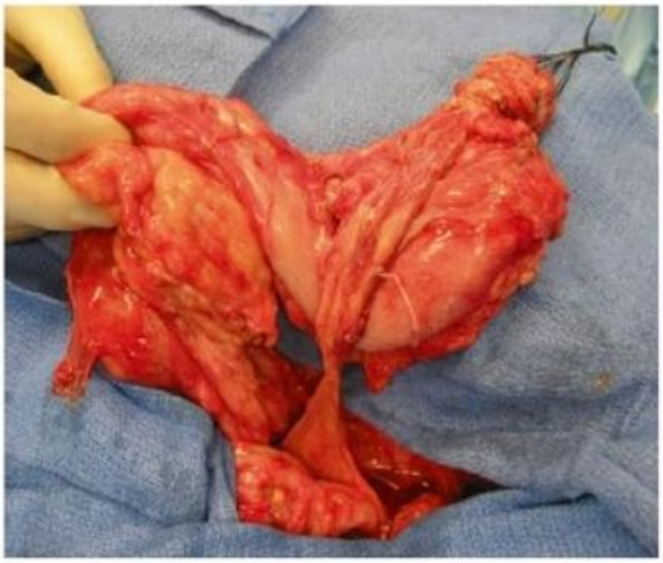

Fig. 16.10 Grotesque abnormal colon frequently seen in cases of cloacal exstrophies or in covered cloacal exstrophies

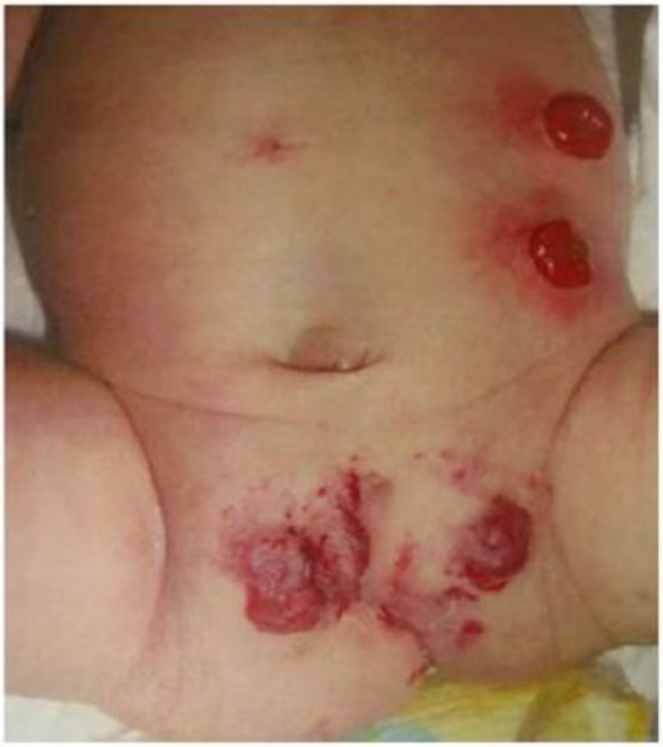

Fig. 16.11 External appearance of a patient with covered cloacal exstrophy. The umbilicus is frequently located lower than normal. Observe low implantation of the umbilicus and hemangiomas

full. Eventually, these girls will need a total urinary reconstruction. In addition, in these patients (covered exstrophies), it is very common to find inside the abdomen the same kind of anatomic abnormalities seen in cloacal exstrophies, mainly the presence of a very short colon with a very abnormal blood supply (Fig. 16.10). Yet, the abdominal wall is intact, which makes a difference with cloacal exstrophies (Fig. 16.11).

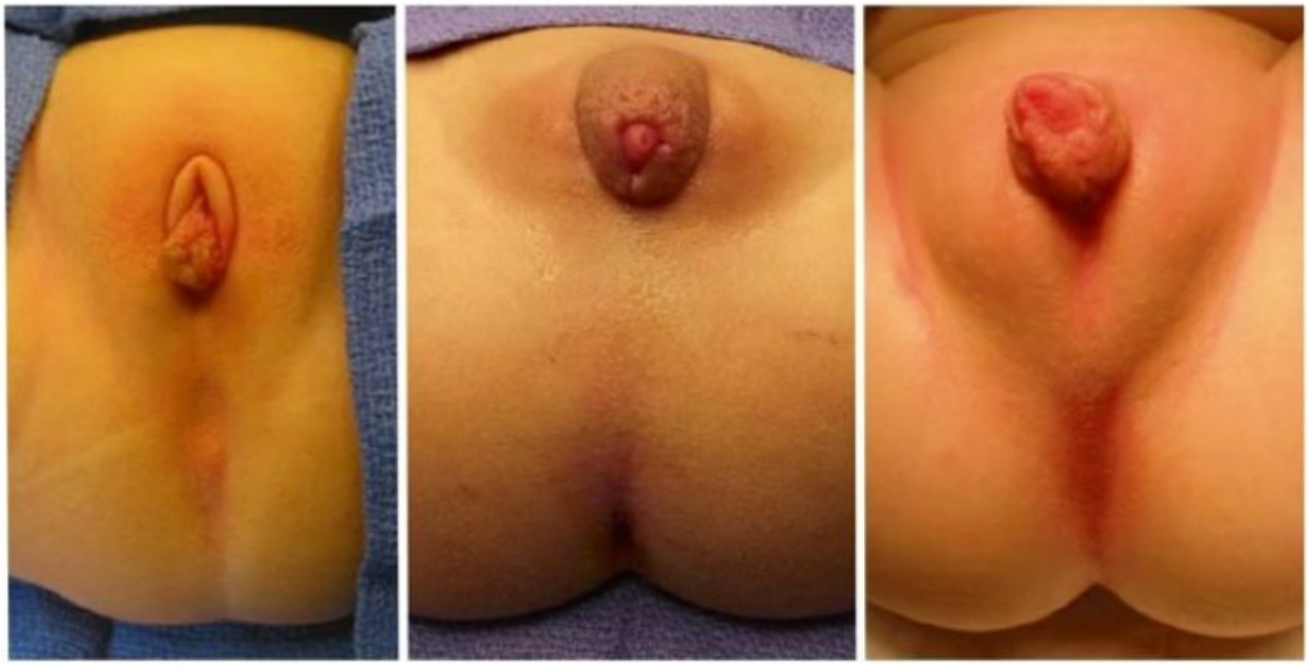

Fig. 16.12 Pictures of patients with a cloaca and a pseudophallus. Palpation of this structure allows to feel only folded skin and no real corpora

It is not unusual to find hypertrophic folds of skin in the area of the single perineal orifice, which gives a false impression of a phallus (Fig. 16.12) [38], and that is why 65 cases in our series came to our institution with the misdiagnosis of intersex made at other hospitals. In our experience of over 531 cases, we only had one case of gonadal dysplasia associated with a vestibular fistula, but never with a cloaca.

The patients that came to our institution with a cloaca and with a history of a suspected diagnosis of "intersex" described the unpleasant experience of being told that their baby had an undetermined gender. It usually took a couple of weeks, with consultation to urology, genetics, and many laboratory tests, to conclude that the patients actually were females suffering from a cloaca.

The key for the diagnosis of those cases with a pseudophallus resides in the palpation of that structure. One can feel that it is actually folded skin with no palpable corpora. Retrospectively, we found five publications referring to this condition as "pseudohermaphroditism" [39]. Some authors used the term "caudal anomalies" [40], "ambiguous genitalia with VATER" [41], or "caudal developmental field defect with female pseudohermaphroditism and VACTERL anomalies" [42]

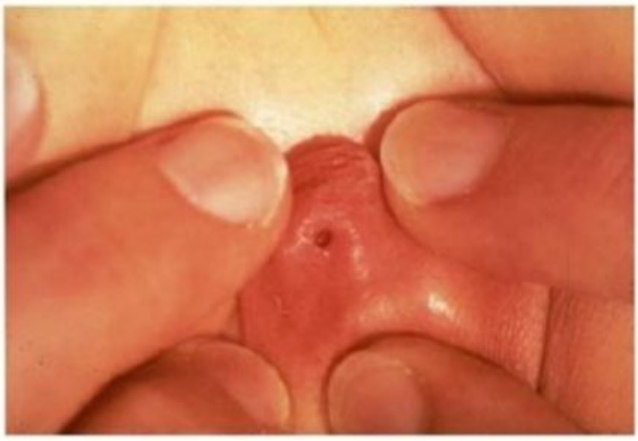

Fig. 16.13 Picture of a cloaca. The single perineal orifice is very small and is located at the tip of a pseudophallus. This kind of external anatomy is usually associated with severe urologic defects and a long common channel

and finally "penis-like clitorises with megalourethra in non-virilized female fetus" [43]. Looking at the pictures of all those cases presented, it was obvious that all those patients suffered from cloacas with normal female gonads and chromosomes.

Figure 16.13 shows a single, very small, very narrow perineal orifice located in the tip of a pseudophallus, which usually means that the patient has other important associated urologic malformations and a long common channel.

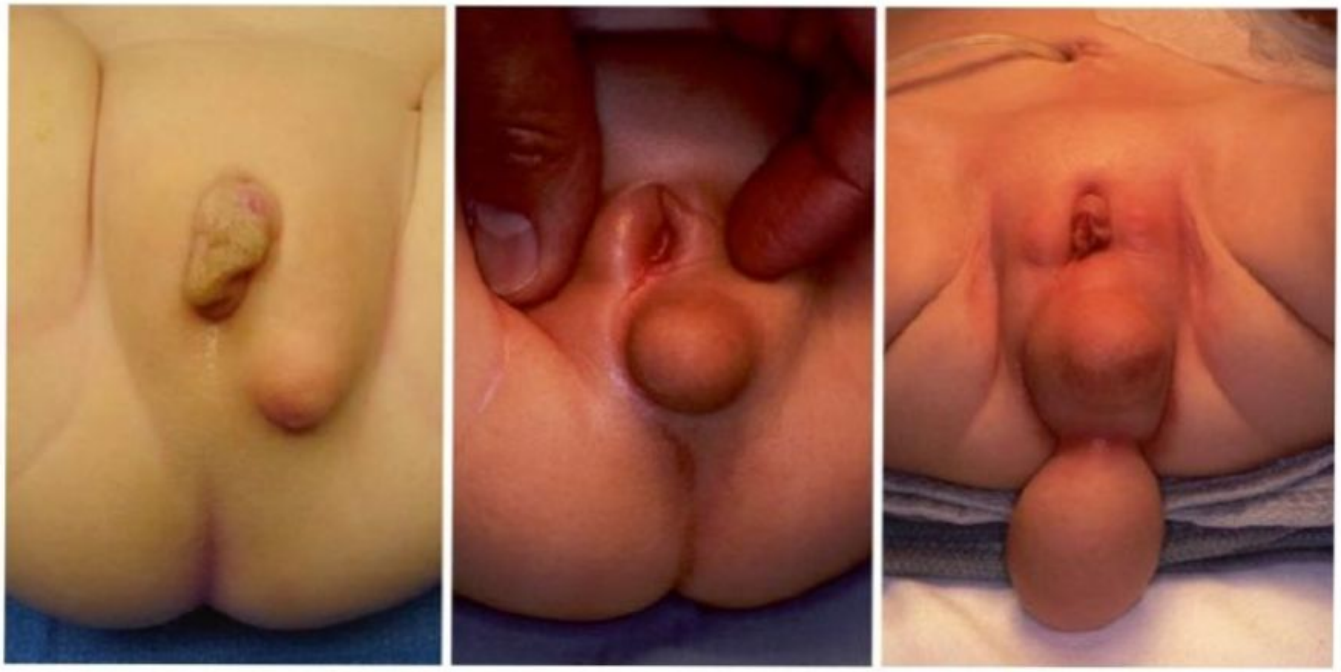

**Fig. 16.14** Lipomas in the perineum of patients with cloaca

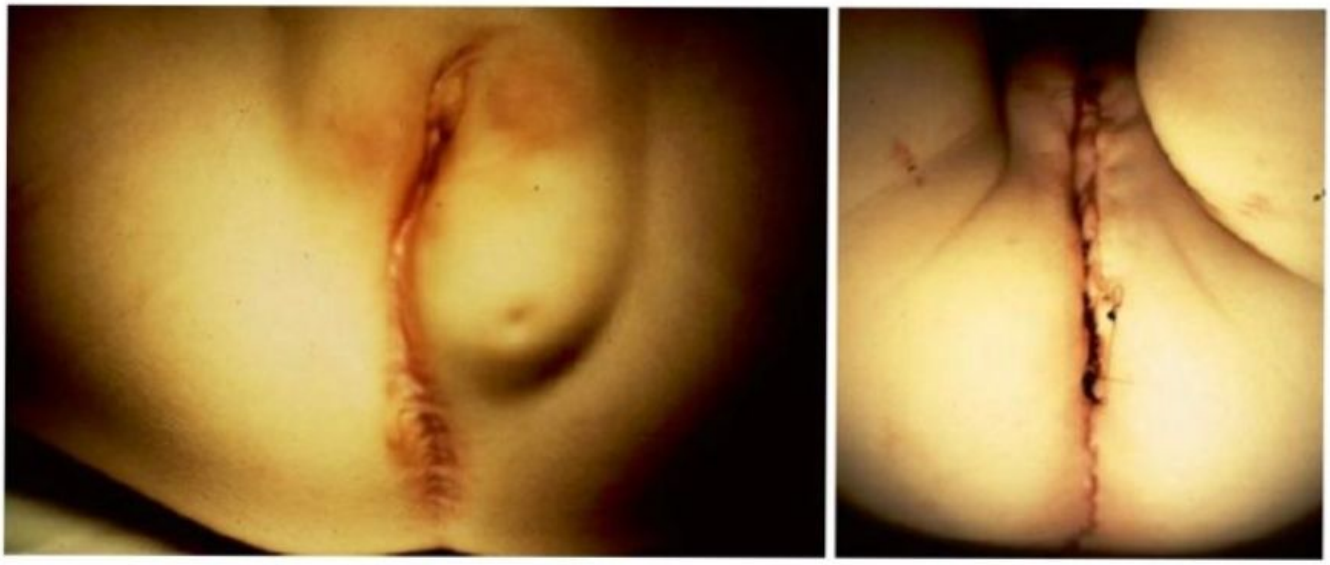

**Fig. 16.15** Preoperative and postoperative appearance after a cloaca repair, including the resection of lipomas

Figure 16.14 shows the perineum of patients with cloaca and lipomas. Lipomas are relatively common in the perineum of patients with cloacas and do not necessarily mean that they require a more complicated type of treatment. At the time of the main repair, the lipoma can be easily excised (Fig. 16.15).

As previously shown, a patient with a cloaca has a very high likelihood of suffering from a urologic condition. The first 24 h of life, like in all other babies with an anorectal malformation, should be used to rule out the presence of associated problems that may represent a risk for life. The most important one, in patients with a cloaca, of course, is urinary tract obstruction. The baby must have a kidney ultrasound to rule out the presence of hydronephrosis and also a pelvic ultrasound to rule out the presence of hydrocolpos and megaureter (Fig. 16.16). A plain abdominal film in a baby with a single perineal orifice may show an image of a pelvic mass, as shown in Fig. 16.17. This represents, most likely, hydrocolpos that must be drained soon. An ultrasound that shows hydronephrosis, megaureter, and a

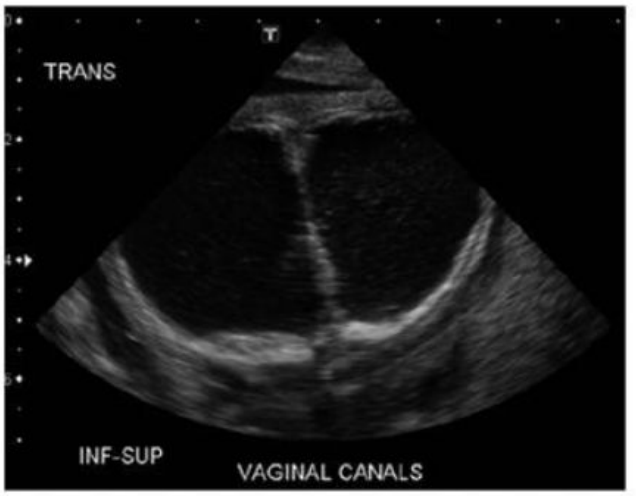


**Fig. 16.16** Pelvic ultrasound of a patient with bilateral hydrocolpos

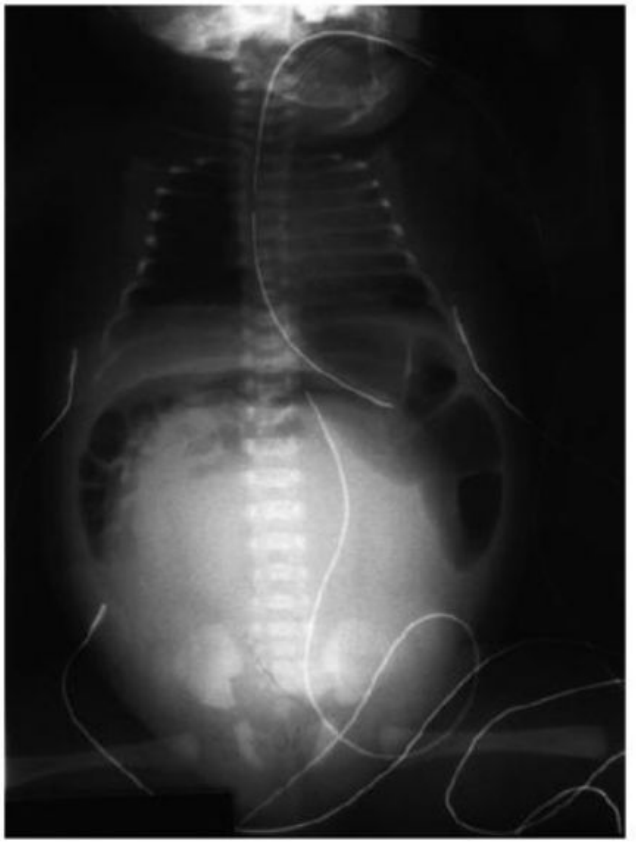

**Fig. 16.17** Abdominal film of a newborn baby with a cloaca and a large hydrocolpos

pelvic cystic mass most likely represents a hydrocolpos that is compressing the trigone and is the cause of the bilateral megaureters and the hydronephrosis.

Hydrocolpos as a cause of megaureters and hydronephrosis has been poorly recognized in the cases that we have received from other institutions. Many patients with hydronephrosis and megaureters were subjected to unnecessary, non-indicated ureterostomies, vesicostomies, and/or nephrostomies (Fig. 16.18). Most of the time, the simple drainage of the hydrocolpos takes care of the problem of megaureter and hydronephrosis, except in those patients who have, in addition, vesicoureteral reflux and difficulty emptying their bladder.

Vesicostomies are occasionally indicated in these babies when we demonstrate that the common channel is too narrow and interferes with the emptying of the bladder. Also, we have seen an indication in babies with massive reflux and megaureters. However, if the patient has a hydrocolpos, the first step should be the drainage of the hydrocolpos, prior to making decisions concerning other procedures. Almost always, drainage of the hydrocolpos is all that is needed to decompress the ureters.

Also, the baby must have an echocardiogram to rule out cardiac conditions. Esophageal atresia must be ruled out in the usual manner. A spinal ultrasound is indicated to evaluate for the presence of tethered cord, and an x-ray film of the abdomen will show the characteristics of the lumbar and thoracic spine, as well as the sacrum, and a sacral ratio can be calculated.

Between 18 and 24 h after the baby is born, a decision must be made concerning the surgical treatment. These babies need a colostomy. The primary treatment of a cloaca without a colostomy has not been attempted, as far as we know. In Chap. 5 in this book, we recommended the opening of a descending colostomy with a mucous fistula, completely separated from the proximal stoma and reduced in size to avoid prolapse, since we only need that orifice to perform irrigations and diagnostic tests (high-pressure distal colostogram). In patients with cloaca, we must put more emphasis in being sure that the patient is left with a piece of colon, distal to the mucous fistula, long enough to guarantee that the pull-through will be possible in the future.

If the patient has evidence of hydrocolpos, the surgeon must be prepared to drain the hydrocolpos at the same time. We specifically recommend a midline subumbilical incision that will give the surgeons access to the entire lower abdomen.

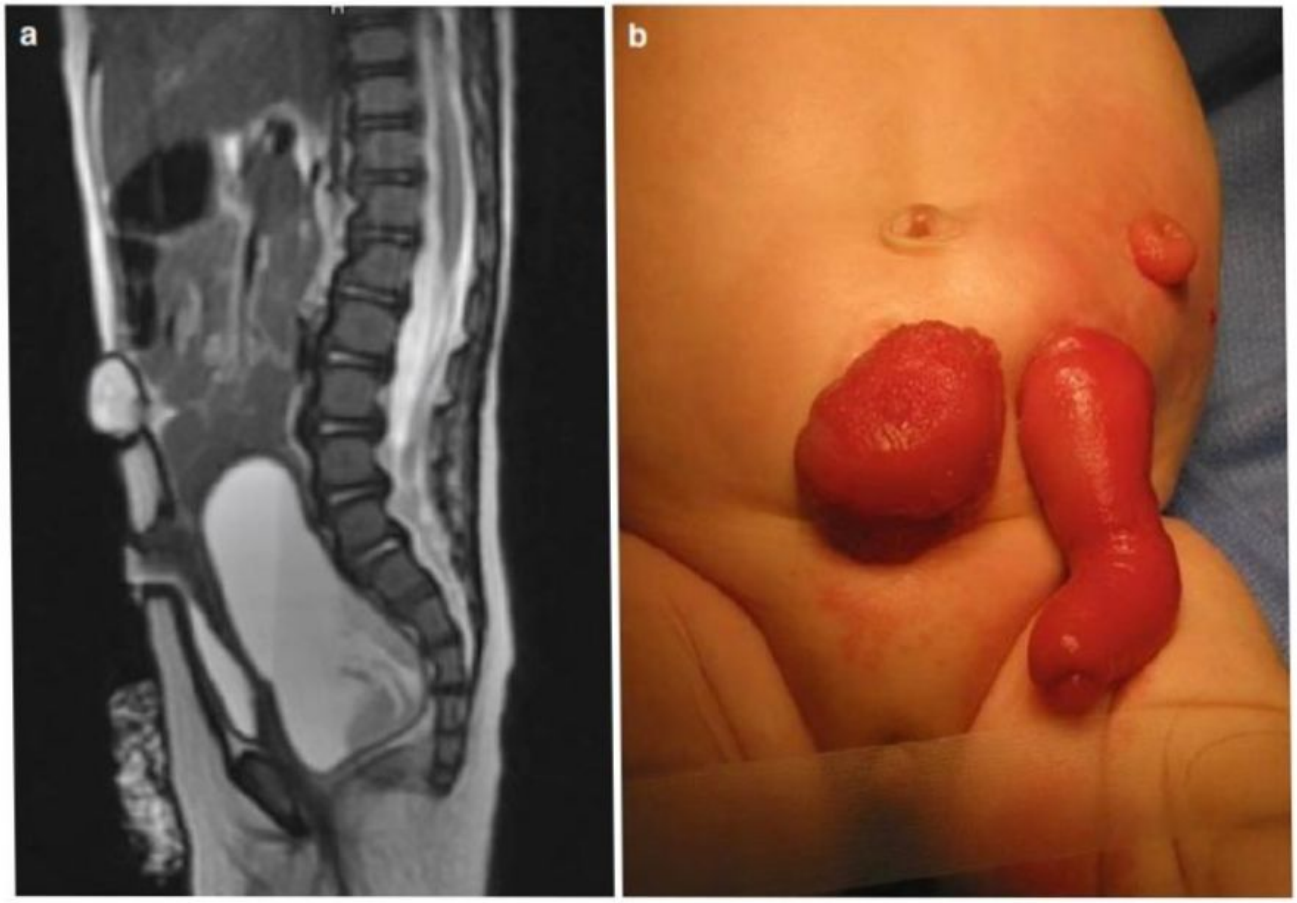


**Fig. 16.18** (a) Picture of a baby with a cloaca, who underwent a colostomy and a non-indicated vesicostomy. The vesicostomy is prolapsed because it was not done correctly and the hydrocolpos remained tense and undrained. (b) MRI showing a sagittal view, the vesicostomy and the undrained hydrocolpos

Through this incision, the surgeon can identify the junction between the descending and sigmoid colon. There, the colon is divided and the stomas are created, separated enough to be able to use a stoma bag without including the mucous fistula. The proximal stoma is created in the left flank and the mucous fistula in the left lower quadrant. At the same time, through this incision, the surgeon will be able to drain the hydrocolpos. We prefer to drain it with a tube. We have used different types of catheters for this drainage. We specifically recommend the use of a pigtail catheter that can be exteriorized through one of the lower quadrants. We like a curled catheter because it is less likely to fall out during the initial several months of life as the inflammation recedes and the vagina moves away from the abdominal wall. Since most of the patients with hydrocolpos have both hemivaginas distended, what we have done in such cases is to create a window in the septum between both hemivaginas in order to use a single tube to drain both hydrocolpos. Figures 16.19 and 16.20 show the aspect of the abdomen of one of these babies with a cloaca after the colostomy has been opened and the hydrocolpos has been drained.

Figure 16.21 shows the kidney ultrasound of a baby with a cloaca born with hydronephrosis and hydrocolpos: (a) before the drainage of the hydrocolpos and (b) shows the same patient's ultrasound after the hydrocolpos has been drained. We cannot overemphasize the importance of draining the hydrocolpos. We have received a series of patients that had a colostomy, vesicostomy, nephrostomy, or ureterostomy, but not drainage of the hydrocolpos. Those patients had multiple problems, including vesicostomy prolapse. One of the patients had a pyocolpos, and another one had a perforation of the infected vagina with severe peritonitis. Several presented with failure to thrive, acidosis, and urinary tract infections, all of which resolved once the hydrocolpos was drained.

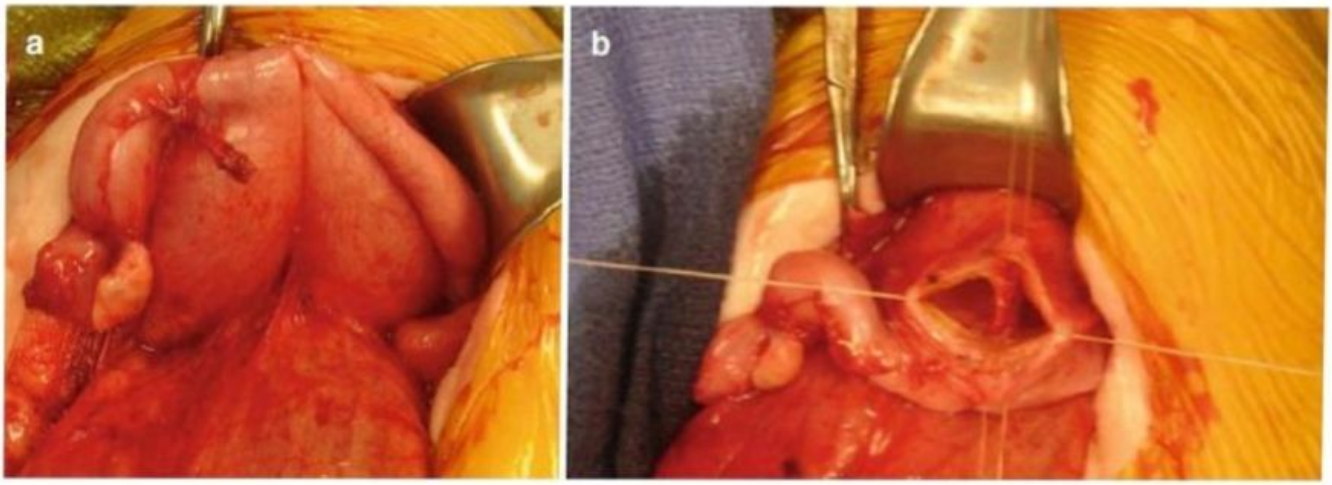


**Fig. 16.19** Intraoperative appearance of a case with two large vaginas (bilateral hydrocolpos). (a) Before drainage. (b) Creation of a "window" in the septum between both hemivaginas

If the bladder cannot empty due to the presence of a quasi-atresia of the common channel, then a vesicostomy would be indicated. Also, in the event of a patient who has a drained hydrocolpos, well-decompressed urinary tract but severe reflux, megaureter as demonstrated on a cystogram, and urinary tract infections, a vesicostomy could be indicated, with a plan for urologic reconstruction in the future. Early ureteral reimplantation of megaureters in a little baby with a bladder that most likely will have some degree of malfunction and kidneys with significant congenital damage is not recommended. We therefore prefer the opening of a temporary vesicostomy, which represents the best way to protect the kidneys.

Some of the hydrocolpos are giant and may even interfere with the respiratory function. Also, some babies with cloacas are extremely sick at birth; they have ascites, hydronephrosis, high creatinine, and severe renal damage.

The presence of calcified meconium in the abdominal film of a newborn baby with a cloaca [44] may represent a serious sign. Sometimes the meconium passes through the fallopian tubes into the peritoneal cavity producing severe peritonitis.

Once the colostomy and urogenital tract decompression has been done, the surgical emergency has been solved. Patients usually recover very well from this operation, and they start eating, growing, and developing normally. The exceptions, of course, are patients that are born with severe kidney damage and renal failure, requiring hemodialysis and consideration for a kidney transplant.

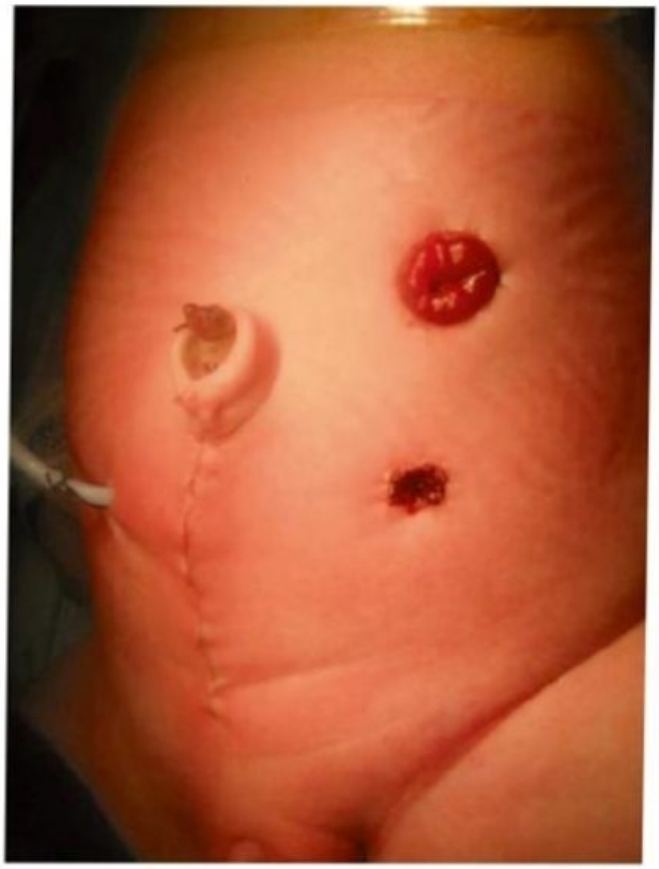

**Fig. 16.20** Abdomen of a patient with a cloaca showing the separated stomas and the vaginostomy tube

A female baby born with a cloaca that has a colostomy and is not doing well postoperatively usually has an undrained obstructed urinary tract with or without hydrocolpos. Therefore, the first study in such patients should be an ultrasound to rule out the presence of hydronephrosis, megaureter, large bladder, or hydrocolpos and act accordingly.

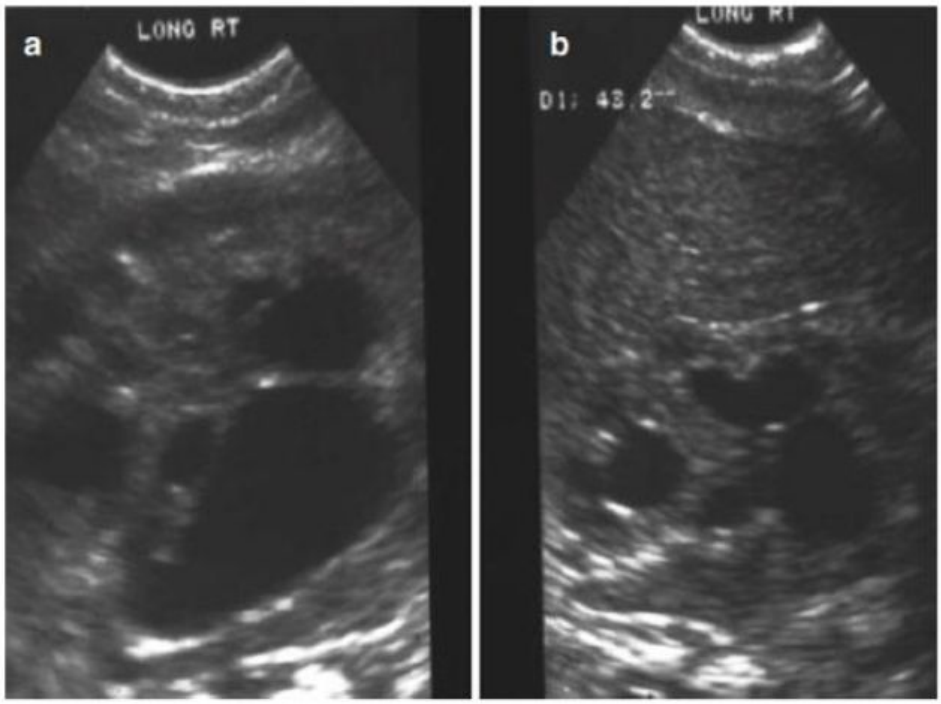


**Fig. 16.21** Kidney ultrasound of a patient born with a cloaca and hydronephrosis. (a) Before drainage of hydrocolpos. (b) After drainage

Another reason why these patients have sepsis and do not grow well sometimes is because the colostomy is inadequate. We are strongly opposed to the opening of loop colostomies in these babies, because that type of stoma frequently allows the passing of stool into distal bowel with direct fecal contamination of the urinary tract.

When the patients are well treated, their colostomy is adequate, and their urinary tract and hydrocolpos are well drained, they usually recover very rapidly and can go home. Within several months, they will be ready for the main repair.

### 16.1.1.4 Main Repair

In June 1982, for the first time, we had the opportunity to use the posterior sagittal approach to repair a cloaca under direct vision. Fortunately, that first cloaca was what we now consider a "benign type" of malformation, meaning that the common channel was relatively short (less than 3 cm), and therefore, we were able to repair the malformation successfully. That particular patient today has urinary control, bowel control, sexual function, and already has successfully delivered a baby by cesarean section.

During the first few years after 1982, our approach for the repair of cloacas consisted of separating the rectum from the urogenital tract like in all other malformations, followed by the separation of the vagina from the urethra and bladder, reconstruction of what used to be the common channel as a neourethra, mobilization and dissection of the vagina to be able to pull it down to be placed posterior to the urethra, and performing a pull-through of the rectum to be placed within the limits of the sphincter [45] (Fig. 16.22). Soon enough, we learned that that approach was highly successful in a certain type of malformations that now we call "benign," but was not successful in other more complex types. The main lesson learned during the last 32 years is that we are dealing with a wide spectrum of defects [1, 46]. It has been an eye-opening, constant learning experience. The more experience we develop, the more we understand that the spectrum seems to be wider and wider. As will be shown in this chapter, the learning process allowed us to design surgical maneuvers applicable to different anatomic variants of these defects. The posterior sagittal anorectovaginourethroplasty is the name that we gave to the repair when it was done in the way that was already described, meaning separation and mobilization of the three structures (rectum, vagina, and urethra), done posterior sagittally.

In many patients, the posterior approach was not enough to repair the malformation, and it was necessary to open the abdomen to complete the repair.

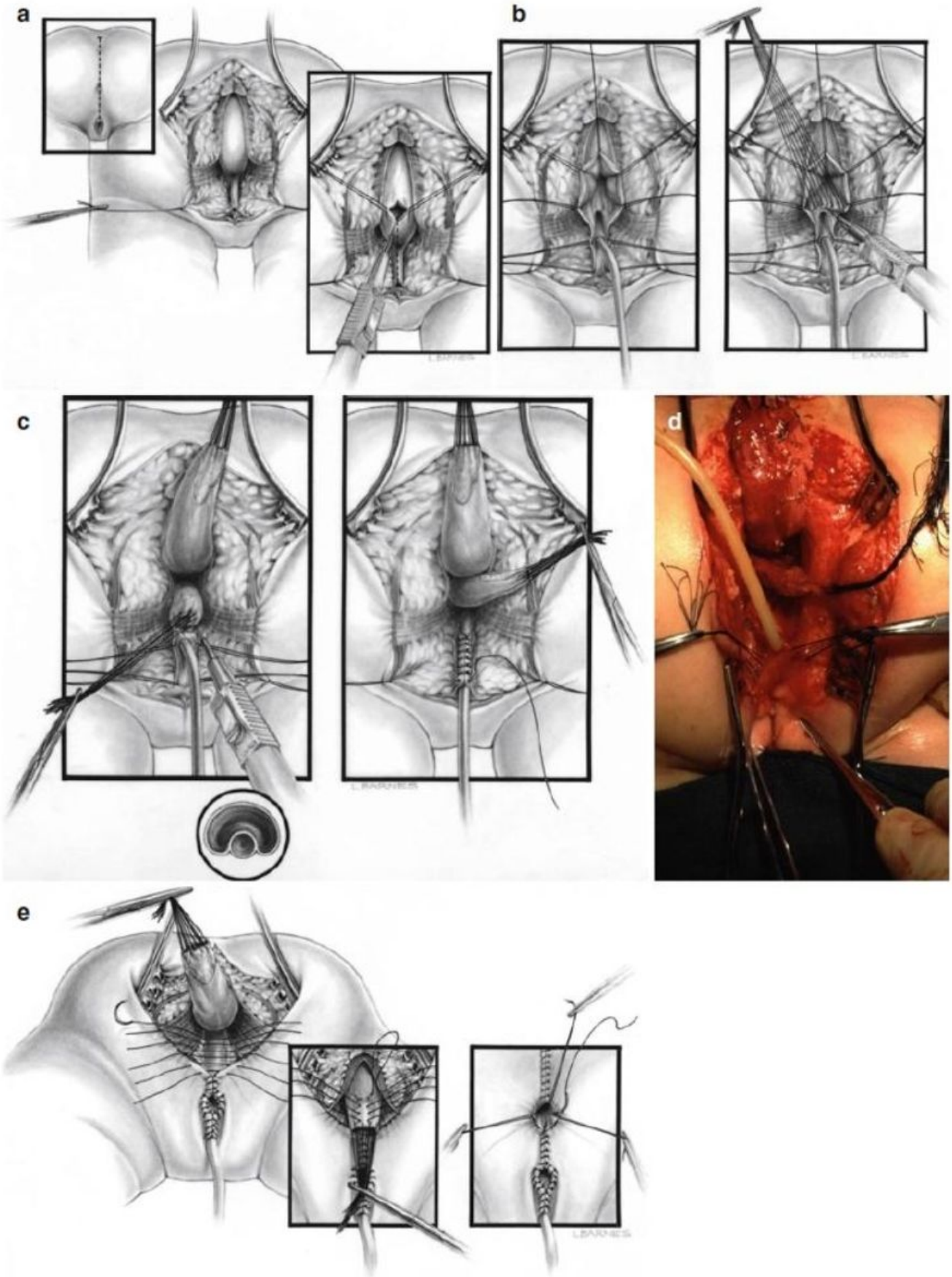


**Fig. 16.22** Diagrams and pictures showing the technique originally used by us, before the advent of the total urogenital mobilization. (**a**) Opening. (**b**) Diagram showing the separation of the rectum from the vagina. (**c**) Diagram showing the vagina being separated from the urethra. The old common channel is reconstructed as a neourethra. (**d**) Intraoperative picture showing rectum and vagina separated. (**e**) Diagram showing the reconstruction being completed

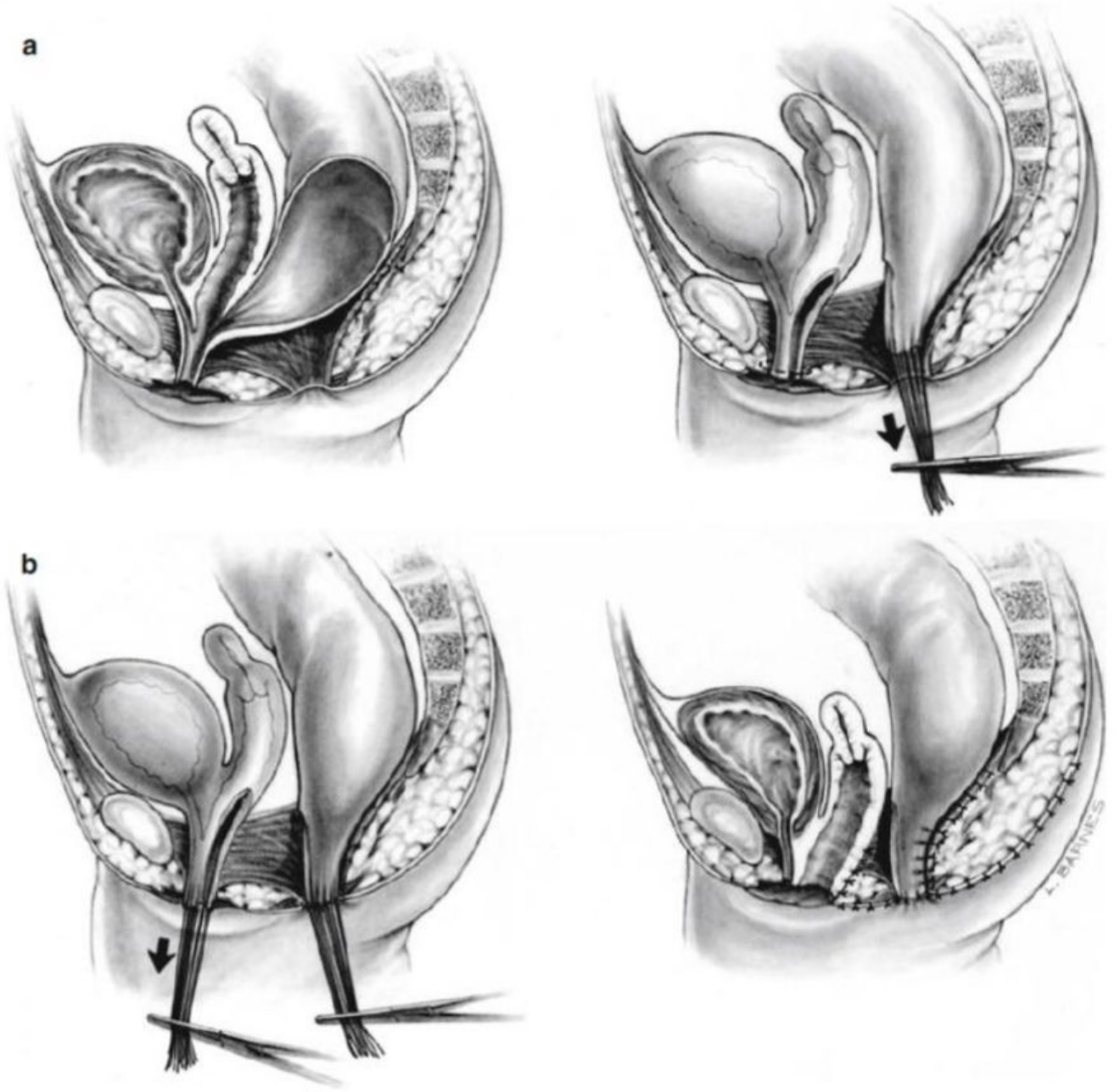


**Fig. 16.23** Total urogenital mobilization (diagrams showing the basic concept). (**a**) Separation of the rectum. (**b**) Total urogenital mobilization

In 1996, for the first time, we used an innovative surgical maneuver that we called "total urogenital mobilization," which allows us to reduce the operative time about 70 %, significantly reduces the blood loss, makes the operation more reproducible, and renders better cosmetic and functional results in the management of cloacas [47] (Fig. 16.23) (Animations 16.1 and 16.2). Subsequently, we found that the total urogenital mobilization was not enough to repair more complex types of defects, and, therefore, we designed the "transabdominal extended total urogenital mobilization," which allows us to repair cloacas with common channels between 3 and 5 cm. Yet, even with the use of an "extended transabdominal" approach, some cloacas required further technically demanding maneuvers, including the complete separation of bladder and urethra from the genital tract (Animation 16.3). In order to do that, we had to open the bladder and pass feeding tubes through the ureters to avoid their injury. In addition, in some patients, we perform a maneuver called "carving the pubic cartilage," in order to create a shorter trajectory for the urethra and vagina to be pulled down behind the pubis and to be sutured next to the clitoris. In some specific type of cases, we apply a

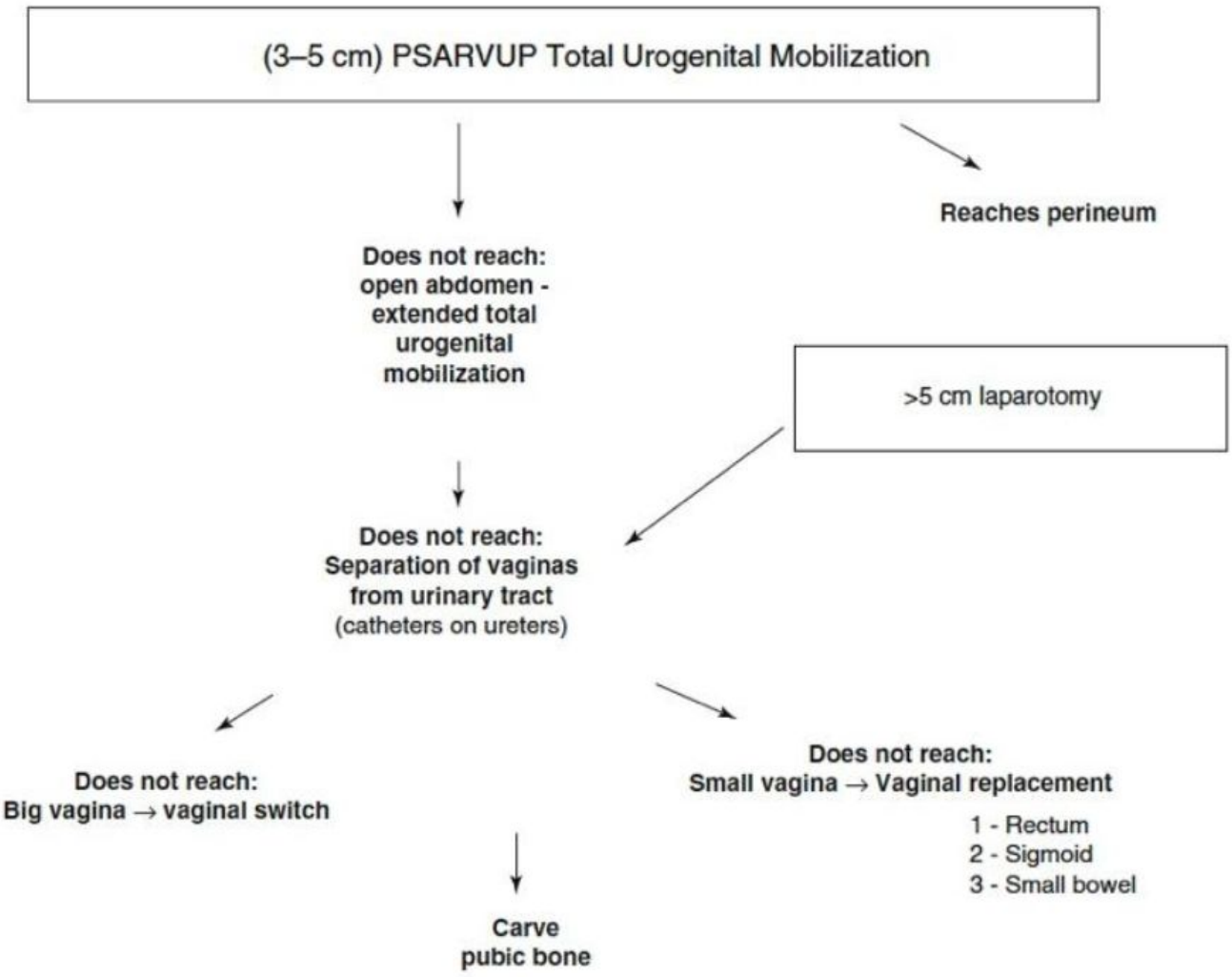


**Fig. 16.24** Decision-making algorithm to repair cloacas with a common channel longer than 3 cm

maneuver called "vaginal switch" that will be described below [48]. In another group of cases, we have to replace the vagina totally or partially, and we perform that with the rectum, colon, or small bowel. Finally, there is a group of cloacas with an extremely long common channel (more than 5 cm), in which we leave intact the common channel to be used eventually as a conduit for intermittent catheterization, and we go directly through the abdomen to separate the vagina(s), and the rectum, from the trigone or the bladder neck.

As we learned more about the complexity of cloacal malformations, we developed a serious concern about the reproducibility of some of the techniques used to repair complex cloacas. Fortunately, more than 50 % of the cloacas have a common channel shorter than 3 cm. This means that they can be repaired posterior sagittally, without opening the abdomen and using the maneuver called "total urogenital mobilization." We believe that the total urogenital mobilization is highly reproducible, and we believe that most pediatric surgeons can learn to do it well. On the other hand, we believe that those cases of cloacas with a common channel longer than 3 cm must be repaired by those surgeons specially dedicated and experienced in dealing with these malformations.

Figure 16.24 shows the different steps of the decision-making algorithm in the repair of cloacas. We will describe each one of them.

### Cloacas with a Common Channel of Less Than 1 cm

Figure 16.25 shows a cloaca with a short common channel. In these cases, we recommend a relatively simple procedure that we call a posterior sagittal anorectovaginoplasty. The urethra is left untouched. Basically, what we do in these cases is to separate the rectum from the vagina the same way that we do in cases of

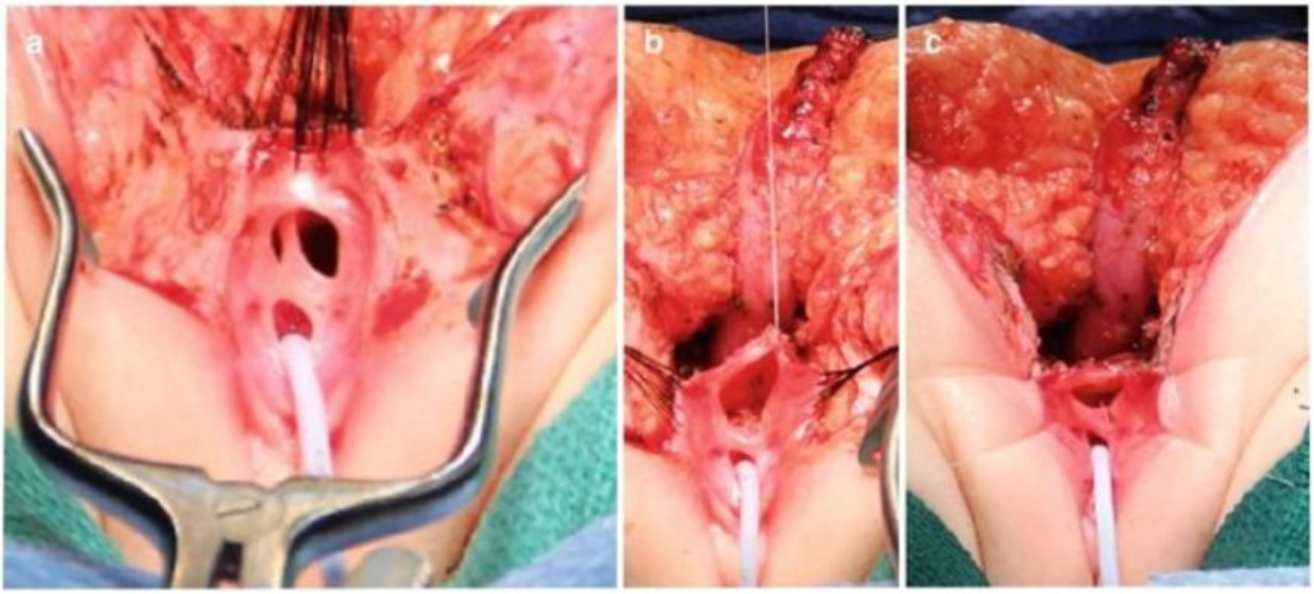


**Fig. 16.25** Intraoperative picture of a cloaca with 1 cm common channel. (**a**) Exposure – multiple silk sutures in the rectum. Observe the vaginal septum. (**b**) Sutures placed in lateral vaginal walls. The rectum has been already separated, and the vaginal septum has been resected. (**c**) Repaired introitus – the lateral walls of the vagina are sutured to the labia. The introitus has been enlarged

vestibular fistulas (see Chap. 15). Next to that, rather than separating the vagina from the urinary tract or performing a total urogenital mobilization, we mobilize only the lateral and posterior walls of the vagina, enough as to be able to suture the edges of the vagina to the skin of the neolabia (Fig. 16.25b). By doing that, we do not disturb the urethra or the common wall between the vagina and urethra, which is a high-morbidity type of maneuver. The cosmetic effect of this operation is excellent. The patients look and behave basically like a patient operated on for a rectovestibular fistula. We call this type of cloaca "cloaca type 1." The results in terms of bowel and urinary control are not different from those of patients with rectovestibular fistulas when they have a normal sacrum. Figure 16.25c shows the final result after one of these introitoplasties. These patients may have mild female hypospadias, which is irrelevant because they do not need intermittent catheterization and because the urethral meatus is readily visible.

### Cloacas with a 1–3 cm Common Channel

Fortunately, 66 % of our patients with cloacas belong to this type. These patients have, in general, a good prognosis. Twenty-eight percent of them will require intermittent catheterization after the reconstructive operation, and the bowel function depends very much on the quality of the sacrum and spine [1, 46].

The procedure to repair these malformations was performed by us any time from 1 to 12 months of age. If a baby happens to be born in our institution and is growing and developing normally, we do it between 1 and 3 months of life. Most of our patients, however, come from other institutions, and, therefore, we have experience doing this procedure at different ages.

We start the operation by performing vaginoscopy and cystoscopy. We strongly recommend for the general pediatric surgeon to do the vaginoscopy and cystoscopy as a separate setting. By doing that, he or she will be able to measure the length of the common channel and based on that to:

- Determine whether or not he is capable of doing that operation or if the patient should rather be referred to another center.
- Determine whether or not it will be necessary to open the abdomen for the reconstruction. This represents important information for the anesthesiologist as well as the entire operating team. It helps with equipment needs, predicting operating time, etc.
- Determine the final functional prognosis.
- Determine whether or not the patient needs a total bowel preparation, in case some form of vaginal replacement with bowel is necessary.

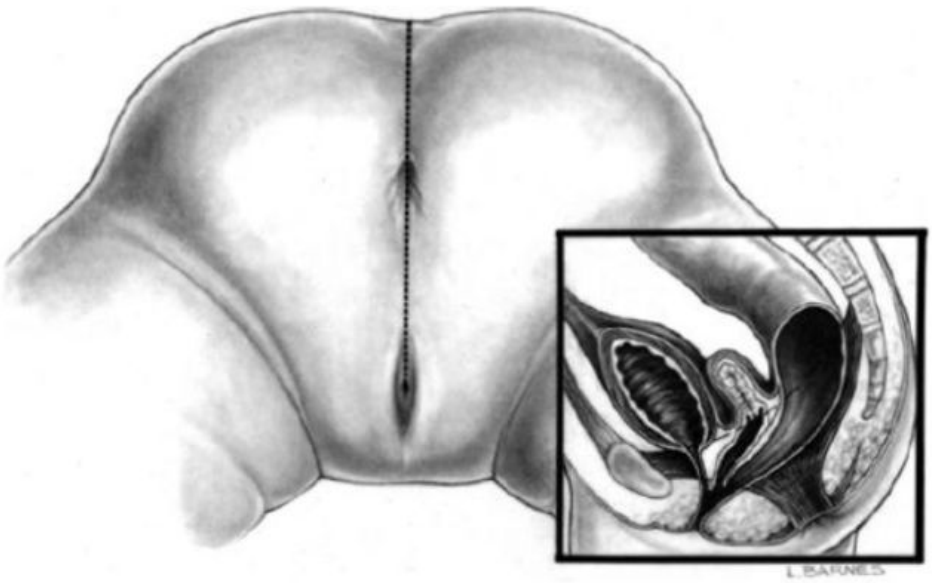

**Fig. 16.26** Diagram of a posterior sagittal incision to repair a cloaca

When the vaginoscopy and cystoscopy shows that the patient has a common channel of 1–3 cm, we can be confident that we can repair that malformation using only the posterior sagittal approach and total urogenital mobilization, without opening the abdomen. The operation will take us approximately 3 h. The cosmetic result is excellent, and the function, in general, is very good.

The patient is placed in prone position with the pelvis elevated and is washed, prepped, and draped in the usual manner. A posterior sagittal incision is used, running from the middle portion of the sacrum down to the single perineal orifice. We divide the skin, subcutaneous tissue, parasagittal fibers, and the entire sphincter mechanism precisely in the midline (Fig. 16.26). The common channel is opened exactly in the midline, including the vagina and the rectum (when it is found), exposing the internal anatomy of the malformation (Fig. 16.27). This step is facilitated by placing a mosquito clamp in the single perineal orifice, to help guide the midline incision of the posterior aspect of the common channel.

The first step, as in all cloacas, consists of separating the rectum from the vagina. When the patient has two Müllerian systems, the rectum is found in the middle of both hemivaginas. Usually, it opens in a little orifice located in the posterior aspect of the vaginal septum. Multiple 5-0 silk stitches are placed around the rectal opening in order to apply uniform traction (Fig. 16.28). Special emphasis is placed on

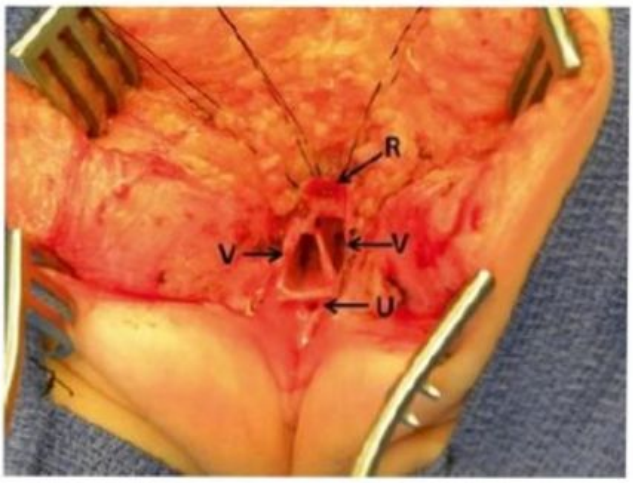


**Fig. 16.27** Picture showing the anatomy of the most common type of cloaca. *R* rectum, *V* vagina, *U* urethra

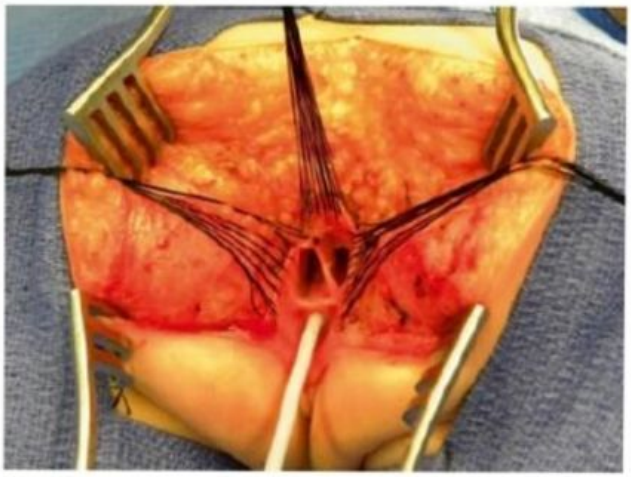

**Fig. 16.28** Multiple fine sutures are placed on the edges of the rectal wall, the lateral walls of the vagina, and the common channel

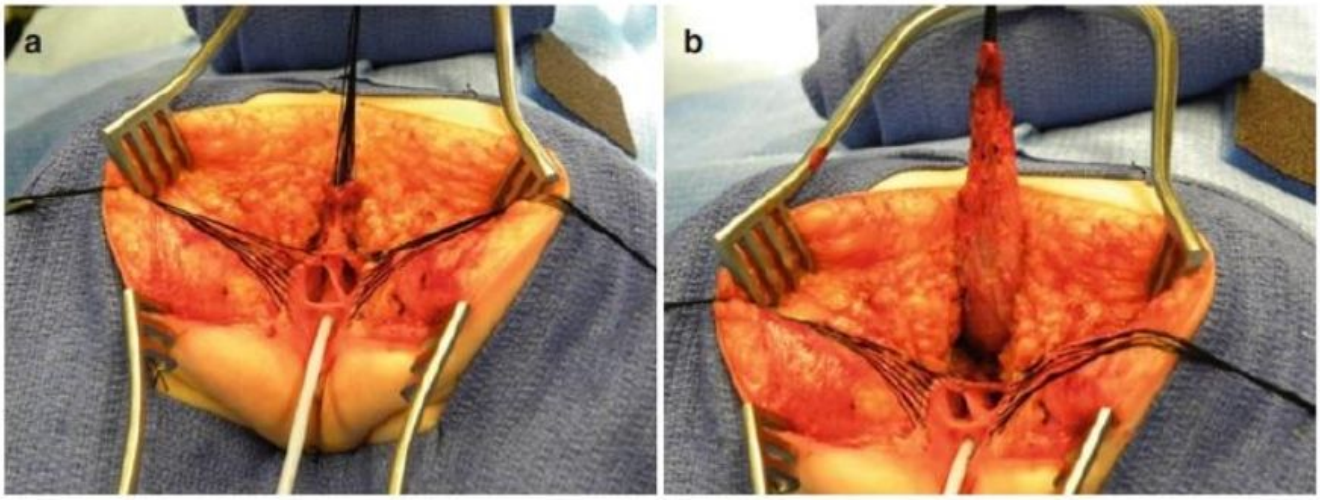


**Fig. 16.29** Rectal dissection. (**a**) The beginning of the separation of the rectum from the vagina. (**b**) Rectum fully separated

creating a plane of dissection in the common wall existing between the rectum and the vagina(s). The use of uniform traction is highly recommended in order to achieve this. We must keep in mind that these structures (rectum and vagina(s)) share a common wall without a natural plane of dissection. Once the rectum and vagina(s) are fully separated, a circumferential dissection is performed, applying uniform traction on the rectum, dividing the bands and vessels that hold the rectum in the pelvis. As we progress with this dissection, we keep gaining length until we have enough rectum to comfortably reach the perineum within the limits of the sphincter (Animation 16.2)

After we finish that part, in the past (before 1996) [1, 45, 46], we used to separate the vagina from the urinary tract, which was a technically demanding maneuver that we do not do anymore in this type of malformation. It took many hours to do this, and over 10 % of our patients suffered from vaginal strictures and/or urethrovaginal fistulas as a consequence of that separation [47]. Because of that, in 1996, we switched to the total urogenital mobilization. For this we place multiple 5-0 silk sutures in the edges of the common channel and the lateral walls of the vagina to apply a uniform traction (Figs. 16.28 and 16.29). Another set of sutures is placed in a horizontal, transverse fashion, about 5 mm from the clitoris (Fig. 16.30). The common channel is divided distal to the transverse line of sutures between the clitoris and the sutures using the needle-tip cautery. The incision includes the full thickness of the common channel. A plane of dissection exists between the pubis and the common channel (Animation 16.2). The separation of the common channel from the posterior aspect of the pubis is a very easy maneuver because there is an obvious plane, and within a couple of minutes, we can reach the upper part of the pubis. Once there, it is relatively easy to identify white, avascular bands that represent the suspensory mechanism of the bladder, vagina, and urethra (Fig. 16.31). These are divided with the cautery as well as their lateral attachments on both sides of the vagina. When we divide these suspensory ligaments of the vagina and urethra, one can see a characteristic fat herniating through the fascia. This is a characteristic retropubic fat pad that indicates that we are in the right plane (Fig. 16.32). The suspensory ligaments of urethra and vagina extend onto both lateral walls of the vagina and must be divided, trying to preserve the blood supply of the vagina. By doing this division of the suspensory ligaments, we gain approximately 2 cm of length in the common channel. We then go to the dorsal part of the vagina(s) and divide the bands, holding them posteriorly and laterally. By doing that, we usually gain another centimeter. As a consequence, in most instances, the total urogenital mobilization allows the mobilization of the vagina and urethra with a common channel of 3 cm comfortably (Fig. 16.33). Occasionally, we were able to totally repair cloacas with up to a 4.5-cm

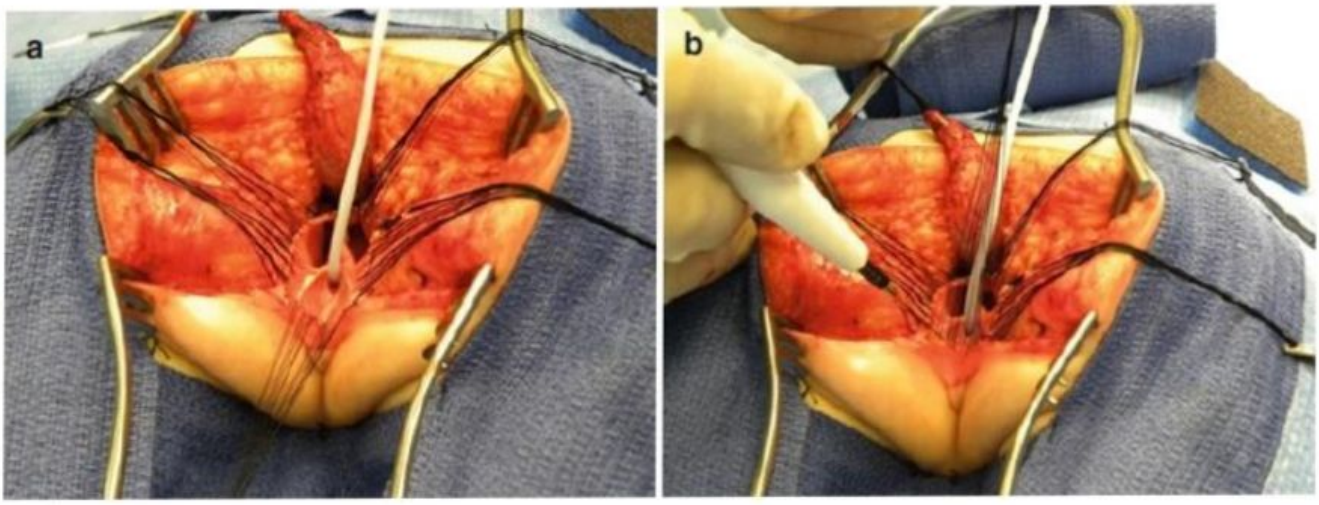


**Fig. 16.30** Picture showing another set of sutures places horizontally, approximately 5 mm proximal to the clitoris. (a) Sutures in place. (b) The urogenital sinus is divided between the clitoris and the sutures

common channel only posterior sagittally, using this maneuver. Other times, we do not know why, we can only gain 2 cm of length, due to lack of elasticity and an inflammatory process that we find in some of these patients.

Once we mobilize the urogenital sinus, we then split in the midline of what used to be the common channel, into two lateral flaps (Fig. 16.34). By doing that, we can suture the urethral meatus to the tissue behind the clitoris with interrupted 6-0 Vicryl sutures (Fig. 16.35). The two flaps that we develop from what used to be the common channel now become part of the neolabia that is sutured to the skin with interrupted 6-0 Vicryl sutures (Animation 16.2). The lateral walls of the vagina(s) are sutured to the neolabia until we create a nice-looking introitus (Fig. 16.36). The electrical stimulator is then used to determine the limits of the anal sphincter, which are marked with temporary silk stitches. The perineal body is reconstructed between the posterior limit of the vagina and the anterior limit of the sphincter. We use 4-0 Vicryl or 5-0 Vicryl sutures to bring together the tissue of the perineal body. These stitches are very important because they represent the main supporting mechanism to avoid dehiscence of the perineum. The skin of the perineal body is sutured usually with interrupted 6-0 Vicryl or 5-0 Vicryl sutures. By doing this, we bring together the anterior limits of the anal sphincter (Animation 16.2). The rectum then is placed within the limits of the sphincter and in front of the levator mechanism. The posterior edge of the levator muscle is sutured with interrupted 5-0 Vicryl sutures in the same way that it is done in all other malformations. The posterior edge of the muscle complex on each side is sutured together in the midline, taking with the same stitches a bite of the posterior rectal wall. The ischiorectal fossa and the subcutaneous tissue are both closed with interrupted 5-0 Vicryl sutures, and the skin is closed with subcuticular 5-0 monofilament, absorbable sutures. The anoplasty is performed with 16 circumferential stitches of 6-0 long-term absorbable sutures after we resect the excessive rectal tissue, usually between 5 and 10 mm (Fig. 16.37).

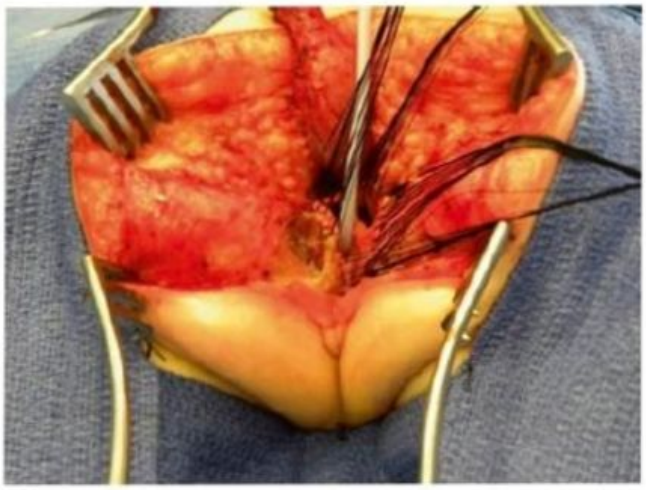

**Fig. 16.31** Picture showing the lateral dissection of the common channel and vagina

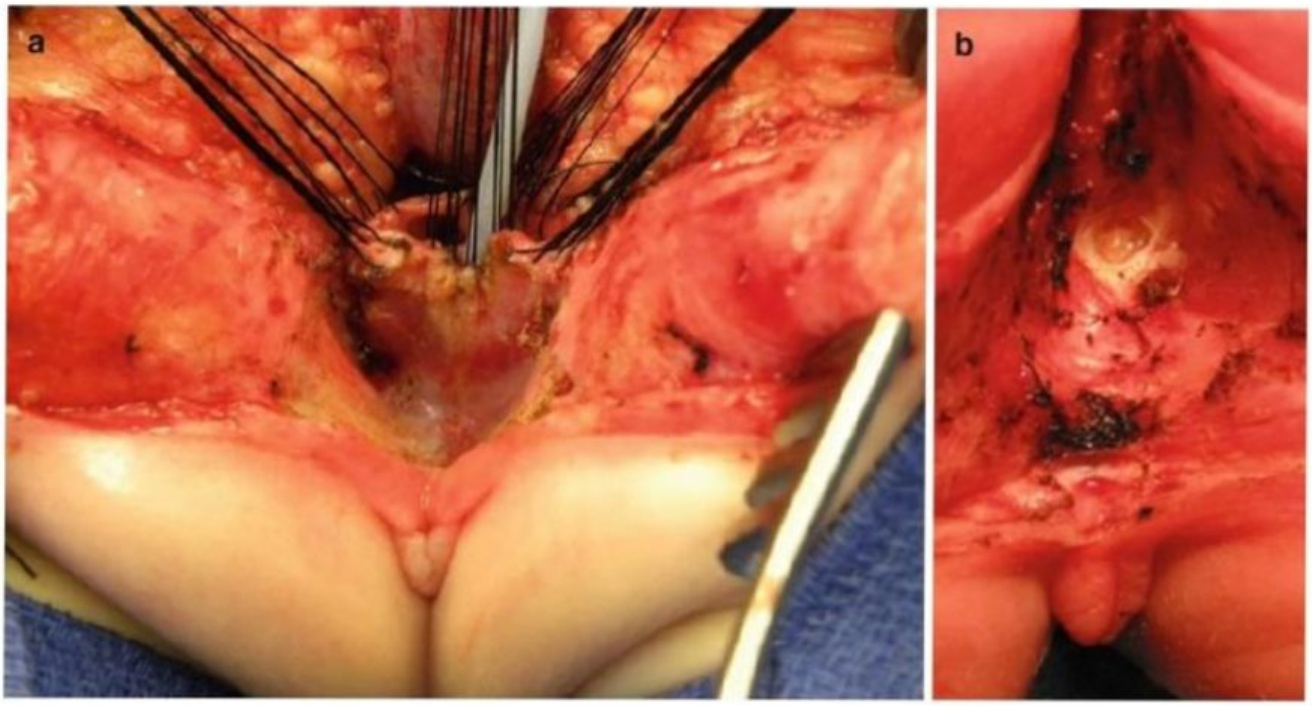


**Fig. 16.32** The suspensory ligaments of urethra and vagina. (**a**) Exposure – observe whitish fascia. (**b**) Divided suspensory ligaments (observe retropubic fat)

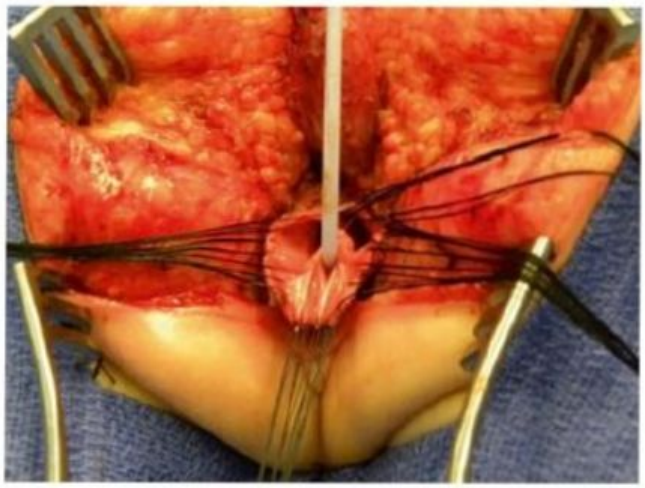

**Fig. 16.33** Picture showing a fully mobilized urogenital sinus

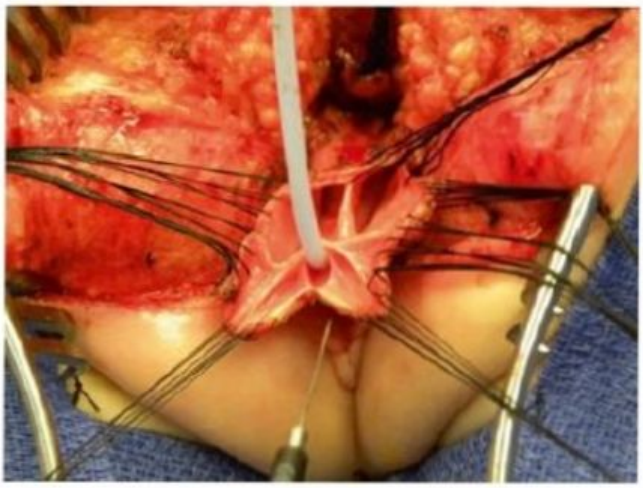

**Fig. 16.34** The urogenital sinus (original common channel) is divided in the midline

These patients do very well and can eat the same day of surgery. The patients stay in the hospital approximately 48 h. A Foley catheter remains in place for approximately 2 or 3 weeks. We must keep in mind that about 20 % of these patients may eventually require intermittent catheterization, and, therefore, we leave the Foley catheter until the postoperative inflammatory process allows us to see where the urethral meatus is located, in case the patient needs intermittent catheterization, before we pull the catheter out. Two weeks after surgery, the parents come to the clinic, we teach them how to dilate the anal orifice, and they do it following our protocol of anal dilatations as previously described.

Prior to the colostomy closure and under the same anesthesia, a vaginoscopy and cystoscopy are performed to confirm that the urethra and vagina are patent and healthy. In the event of finding problems with these, they have to be taken care of, prior to the

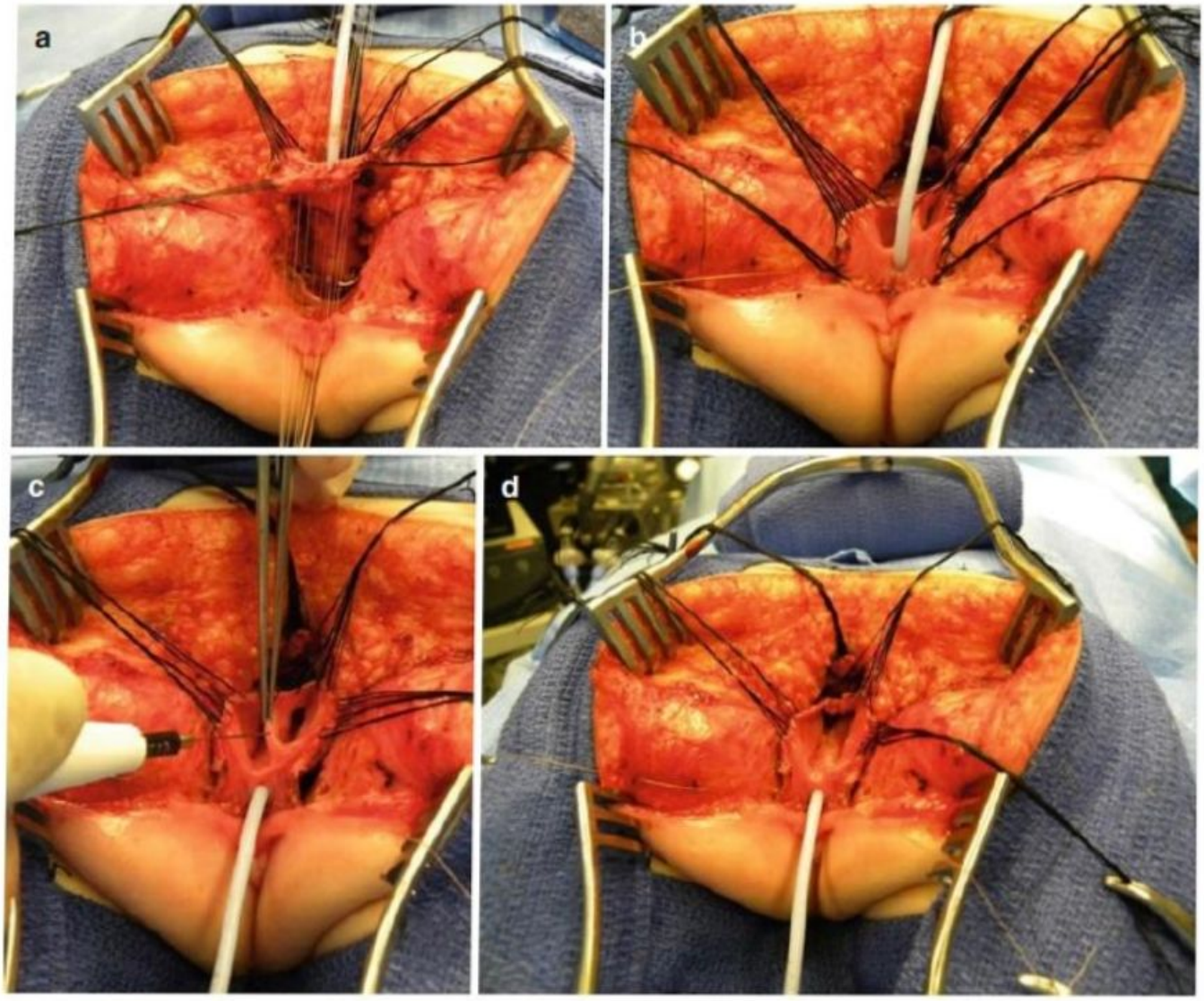


**Fig. 16.35** Urethral opening repositioning and resection of the vaginal septum. (**a**) Fine long-term absorbable sutures are used to anastomose the urethral opening immediately behind the clitoris. (**b**) Sutures are tied. (**c**) Resecting the vaginal septum. (**d**) Vaginal septum resected

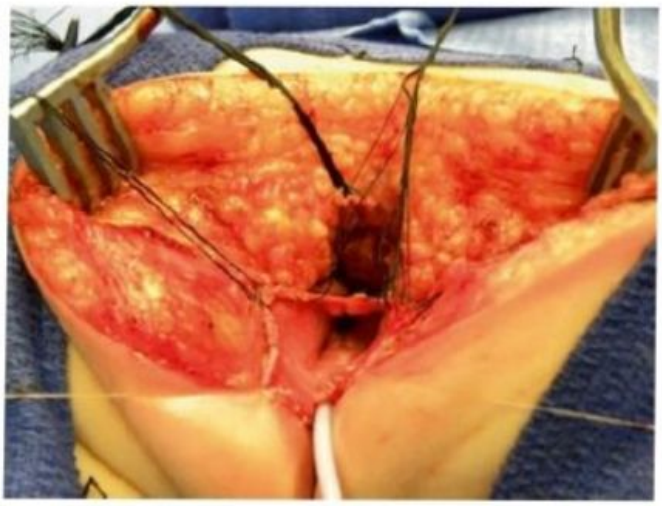

**Fig. 16.36** Suturing vaginal walls to the neolabia

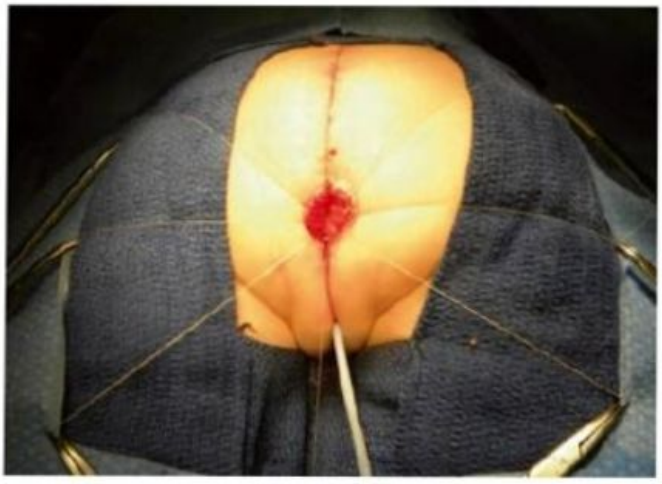

**Fig. 16.37** Final external aspect of a repaired cloaca

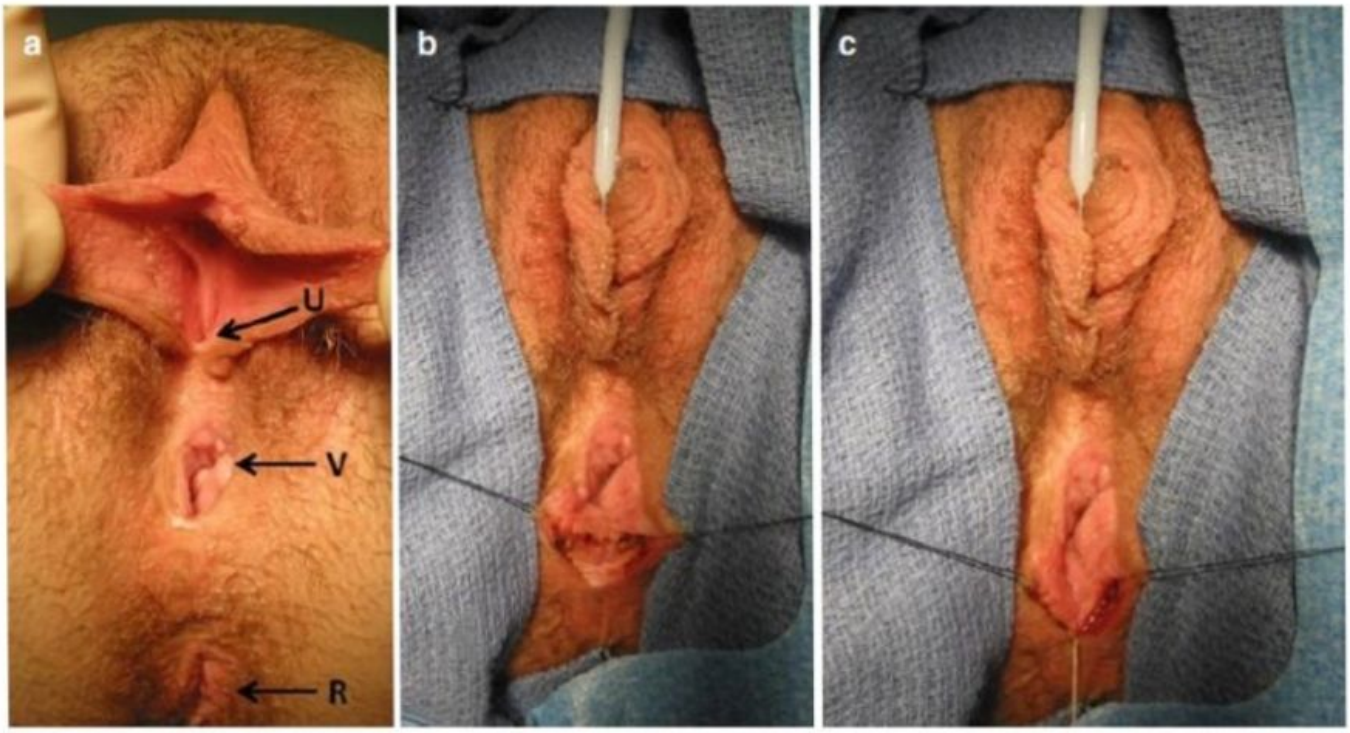


**Fig. 16.38** External vaginoplasty to enlarge a strictured vaginal orifice. (**a**) Narrow vaginal orifice *R* rectum, *U* urethra, *V* vagina. (**b**) Longitudinal incision of the posterior aspect of the anal opening. (**c**) Horizontal suture

colostomy closure. If, for instance, at the time of colostomy closure, we find a narrow ring-like, vaginal opening with a wide, deep compliant vagina, we may do nothing at this age. On the other hand, if the orifice is too narrow and we believe it is at risk of closing completely, then we perform an external vaginoplasty to make the orifice larger and postpone the colostomy closure for a month (Figs. 16.38 and 16.39). On the other hand, if the patient has a long, narrow vaginal stricture, she may need a complete reoperation. We frequently saw this kind of complication prior to the advent of the total urogenital mobilization. The total urogenital mobilization prevents this complication from happening most of the time since it preserves a very good blood supply for the vagina and the urethra.

After the total urogenital mobilization, some patients leak urine for a period of several weeks. A voiding cystourethrogram at this point may show an image consistent with an absent bladder neck, due to the pulling of the whole urogenital sinus from below. This happens mainly when the repair required a significant traction of the urogenital tract. However, most of these patients recover normal urinary function after a few weeks. The colostomy can be closed following the same principles that we mentioned in the chapter related to colostomy closure.

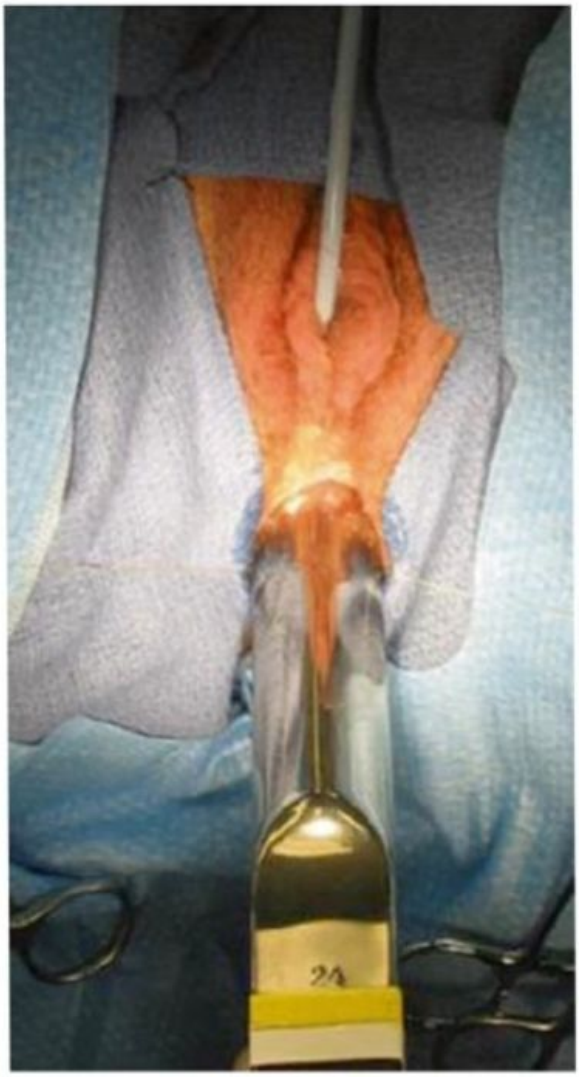

**Fig. 16.39** Enlarged vaginal orifice. Large Hegar dilator inserted

### Cloacas with a 3- to 5-cm Common Channel (Animation 16.3)

When the endoscopy allows us to determine that the patient has a common channel length of 3–5 cm, we perform a total body preparation as described in Chap. 11 because we know that most likely it will be necessary to open the abdomen, in addition to the posterior sagittal approach, to repair the malformation. We tell the anesthesiologist about these findings and the possibility that the operation will go for a longer period of time than one that could be done posterior sagittally only. After we perform a total body preparation, we put the patient in the prone position with the pelvis elevated and perform the same posterior sagittal incision as previously described. The internal anatomy of the malformation is exposed; the rectum is separated from the urogenital tract in the same way as previously described. A total urogenital mobilization is performed as previously described. Occasionally, we are happily surprised to find out that it is possible to reconstruct the urethra, vagina, and rectum without opening the abdomen. However, most of the time, the total mobilization is not enough to repair the malformation. In such a case, the patient is turned to the supine position and the abdomen is opened with a midline infraumbilical incision.

The next step is to perform what we call "extended transabdominal urogenital mobilization." Traction is applied to the dome of the bladder; the lateral attachments of the bladder are divided to obtain a better exposure (Fig. 16.40). The midline incision is extended all the way down to the pubis. Between the bladder and posterior aspect of the pubis, one can see the space that we created from below with the total urogenital mobilization. The urogenital sinus is then brought up through this space between the bladder and pubis (Fig. 16.41). At this point, we divide all the pelvic avascular attachments of the bladder and urethra. Through the laparotomy, these attachments are easily seen and divided. Usually this maneuver allows us to gain extra length on the urogenital mobilization. If that is enough to complete our repair, we then go ahead and pull through back down the urogenital complex and repair the urethra and vagina as previously described. If that is not enough to achieve a tension-free anastomosis between the urethra and clitoris and vagina and neolabia, a maneuver called "carving of the pubic cartilage" is indicated.

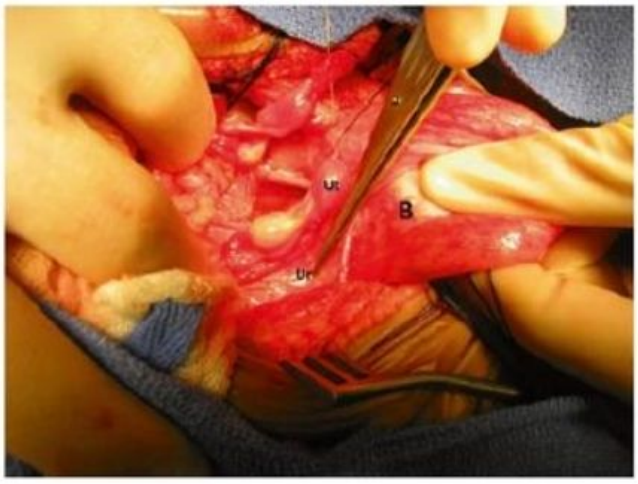


**Fig. 16.40** Intraoperative picture showing the bladder pulled caudally to have a better access to the pelvic floor. *B* bladder, *Ut* uterus, *Ur* ureter

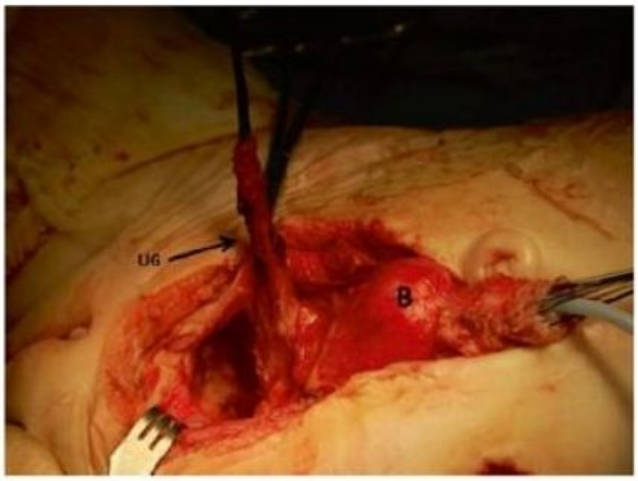


**Fig. 16.41** Intraoperative picture showing bladder and urogenital sinus out of the pelvis. *Us* urogenital sinus, *B* bladder

#### Carving of the Pubic Cartilage Maneuver

Under normal circumstances, the urogenital sinus is located behind the pubic cartilage; it runs below the cartilage and up, anterior to the cartilage to connect to the clitoris. Resecting approximately 50 % of the lower portion of the pubic cartilage does not compromise the pelvis stability, and yet, it allows a straighter trajectory

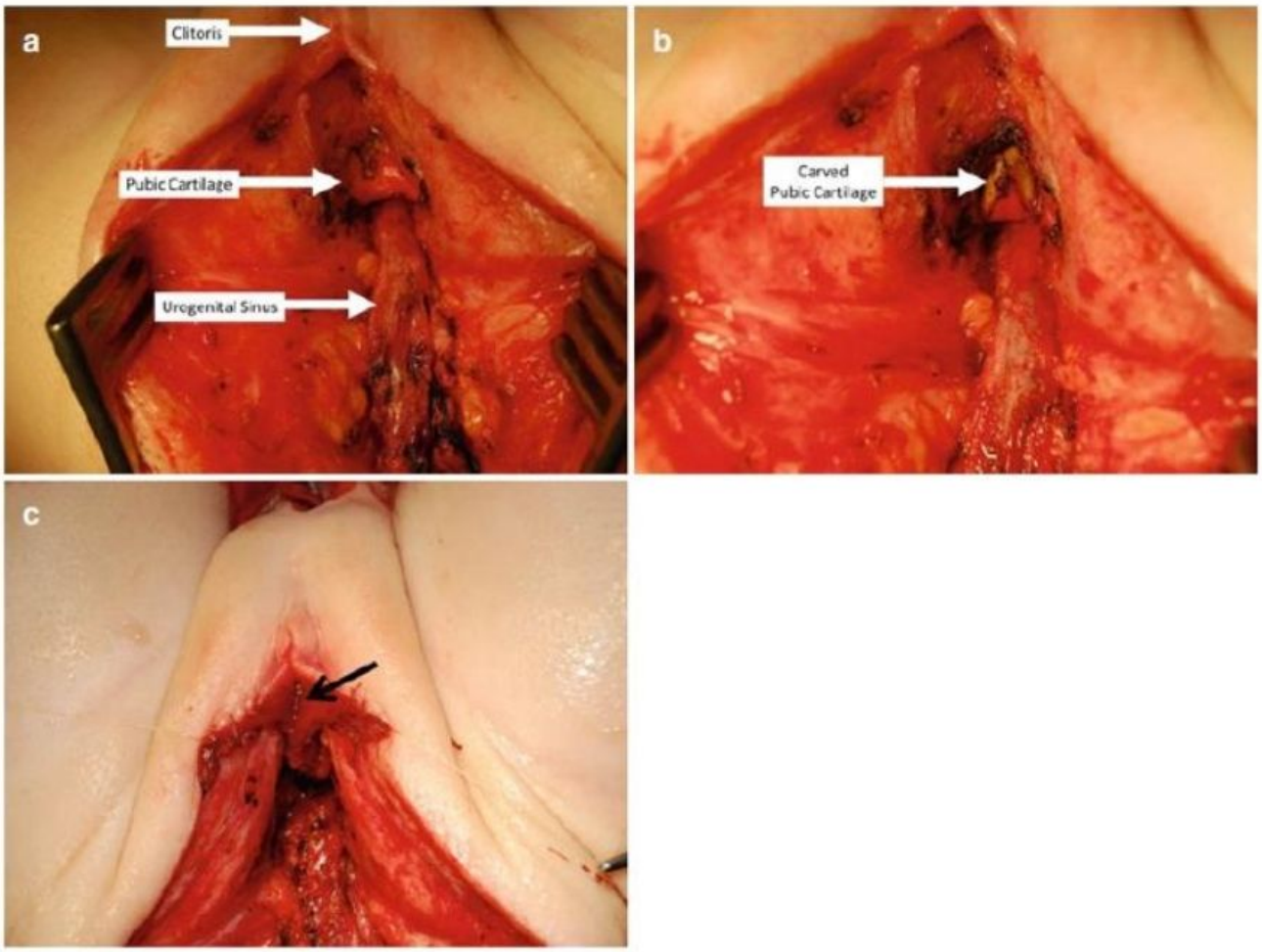


**Fig. 16.42** Carving the lower part of the pubic cartilage to create a shorter trajectory of urethra and vagina. (a) Before carving. (b) After carving. (c) Urethra sutured – *arrow* in urethral opening

of the urogenital sinus. The resection of the cartilage can be done easily, with the needle-tip cautery on "cutting" mode in babies. In older patients, this can be done with a "rongeur" type of instrument. This maneuver may allow for a tension-free anastomosis between the urethra and vagina with the clitoris and neolabia (Fig. 16.42). This maneuver may work in cases that need 0.5–1 cm of extra urethra and vaginal length to achieve a tension-free anastomosis. If that is not enough, then the next step must be the separation of the vagina from the urinary tract.

### Separations of Vagina(s) from the Urinary Tract (Animation 16.3)

This is the most technically demanding maneuver of the entire repair of cloacas. This procedure, in the past, before the total urogenital mobilization, was attempted from below, but it was very difficult to do. Now that we do the total urogenital mobilization, we do not have to separate the vagina from the urinary tract in cases with common channel shorter than 3 cm. Yet, in cases with longer common channel, we must separate both structures through the abdomen. The separation is done through a laparotomy but with bladder and vagina(s) fully mobilized and out of the abdomen. The bladder is opened in the midline, and feeding tubes are introduced through each one of the ureters (Fig. 16.43a). We must keep in mind that in cloaca patients, both ureters pass through the common wall between the vagina and the bladder. The separation of these two structures may include the skeletonizing and dissection of both ureters. If the patient suffers from reflux, this is a golden opportunity to perform a ureteral reimplantation or, if appropriate, a cutaneous ureterostomy; otherwise, to do it later would represent a technically more demanding

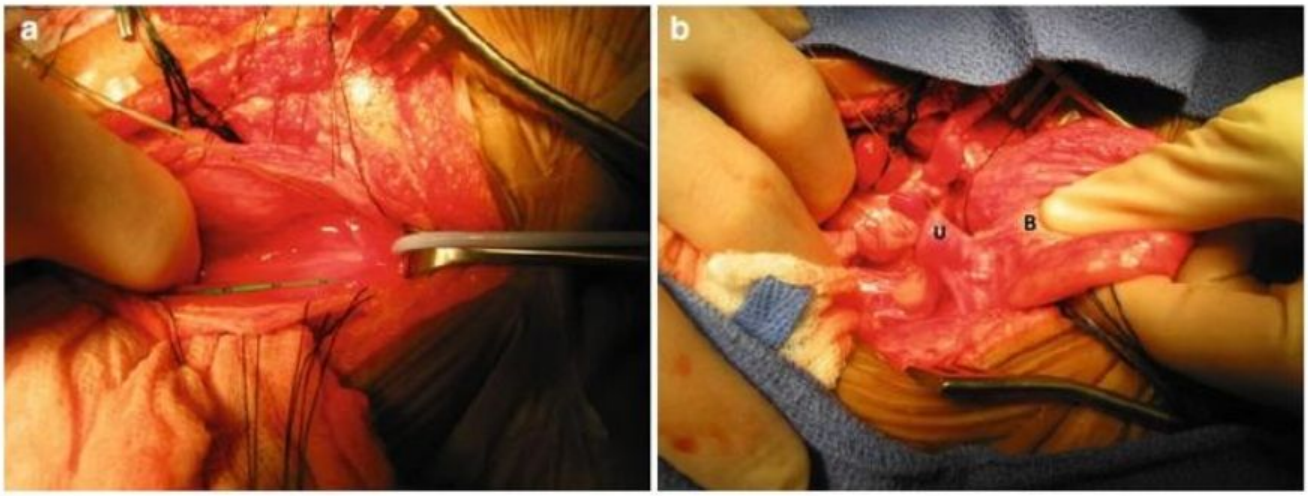


**Fig. 16.43** Intraoperative pictures taken during the separation of vagina from the urinary tract. (**a**) Bladder open and catheters placed in the ureters. (**b**) The assistant puts fingers into the bladder and thumb outside the bladder, pulling it caudally. Traction sutures are placed in the uterus to apply traction and facilitate the dissection. *B* bladder, *Ut* uterus

procedure. The assistant puts two fingers inside the bladder and applies traction caudally into the bladder (Fig. 16.43b). Vicryl sutures are used to pull the uterus or hemiuterus in the opposite direction. A plane is created in the middle of the wide common wall that exists between the vagina(s) and the urinary tract. This common wall extends from the urethra and includes the bladder neck, trigone, and part of the bladder. In general, the most technically demanding steps of the operations designed to repair anorectal malformations is actually the separation of the structures (rectum, genitalia, and urinary tract). We are supposed to separate them without damaging them. This is difficult to achieve for several reasons:

(a) Those structures are congenitally fused without a plane of separation.
(b) The common wall has a very rich blood supply.
(c) The exposure is difficult because these structures are located in a place difficult to reach from below or from above.
(d) The ureters run through this common wall.

The separation of structures usually takes about 70 % of the total operative time. To achieve a good repair, it is necessary to achieve a good separation of these structures with minimal or no damage. The previously described "extended transabdominal total urogenital mobilization" allowed us to perform the separation of structures basically outside the abdomen. The surgeon works from above between the bladder and the vagina(s). We perform the entire dissection with a fine needle-tip cautery. During the dissection, the surgeon must stop frequently to verify that the thicknesses of the vaginal wall, as well as that of the bladder, are equal. In other words, the surgeon does this to be sure that neither of those walls are becoming too thin. Intermittently, this dissection is interrupted to palpate the location of the ureters (previously catheterized). Also, it is convenient to perform part of the dissection backwards, meaning from caudal to cephalad, since the common channel is exposed and brought up through the incision. We keep dissecting a little bit from below and then from above, until both dissection planes meet. At that point, the only structures that join the genitourinary structures to the patient's body are the ureters, the ovarian vessels, and the internal iliac vessels. These vessels must be kept in mind and carefully respected.

Figure 16.44 shows the posterior aspect of the bladder and trigone. Both ureters can be seen intact.

Once the separation has been achieved, the surgeon can plan the type of reconstruction that is best for the patient's specific anatomic variant. The first possibility is that after the separation, one becomes happily surprised to find that the vagina(s) actually reaches the perineum. This is

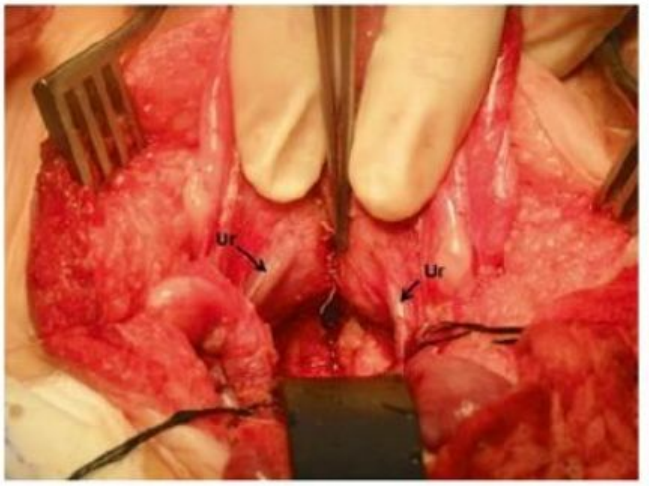


Fig. 16.44 Picture taken after the bladder and vagina have been separated. The ureters can be seen intact. *Arrows* on ureters. Black retractor pushing down uterus and vagina

Fig. 16.45 Anatomic characteristics of a case that will benefit from a vaginal switch maneuver

the ideal time to remove the vaginal septum that separates two hemivaginas (if present) to trim off damaged vaginal tissue and tubularize the available vaginal tissue in preparation for the pull-through.

The total separation of vagina(s) and urinary tract runs with the implicit risk of devascularization of the distal urethra. It is necessary to be sure that the urethra that is sutured immediately behind the clitoris has a good blood supply. At least ten of our patients suffered from an ischemic-acquired urethral atresia after one of these operations performed by us.

If the urethra's blood supply is severely deficient, the surgeon must make a decision about the possibility of permanently closing the bladder neck. In that case, the patient needs a vesicostomy; subsequently, an artificial conduit for bladder catheterization will be done (Mitrofanoff principle) [49]. Later in the patient's life (3–4 years), the functional and anatomic characteristics of the bladder can be studied to determine its capacity, detrusor activity, compliance, as well as the presence of vesicoureteral reflux, and a final reconstruction can be planned, which may include a bladder augmentation and a Mitrofanoff type of procedure.

Planning the reconstruction of the vagina, urethra, and rectum, after the total separation of the structures, the surgeon will face one of several scenarios, and based on those, he/she will make a decision. The first possibility will be that the patient has anatomic characteristics that make her suitable for a surgical maneuver called "vaginal switch."

### Vaginal Switch

This maneuver is applicable only when the patient has a specific type of anatomy (Fig. 16.45). These patients have two hemivaginas very separated, as well as the hemiuteri with a vaginal septum and originally two large hydrocolpi. If we can estimate that the distance between one hemiuterus and the other is longer than the vertical length of both hemivaginas, then the patient may be a candidate for the vaginal switch maneuver [48]. As can be seen in Fig. 16.46, the maneuver consists in sacrificing one of the hemiuteri, being careful enough to preserve the ovary and its blood supply. The vaginal septum is removed. Both dilated hemivaginas are tubularized into a single vagina, and what used to be the dome of one side, the place where we resected the hemiuterus, becomes the lowest part of the new switched-down vagina (Fig. 16.46) (Animation 16.4). This maneuver works, and we have several patients menstruating through this type of repair, but it is only applicable if the patient has the specific type of anatomy already described. If, early in the operation, one estimates that the anatomy of the patient belongs to this category, one has to separate completely only one hemivagina from the urinary tract and try to preserve the blood supply of the opposite side. In this type of maneuver, the blood supply of the entire switched

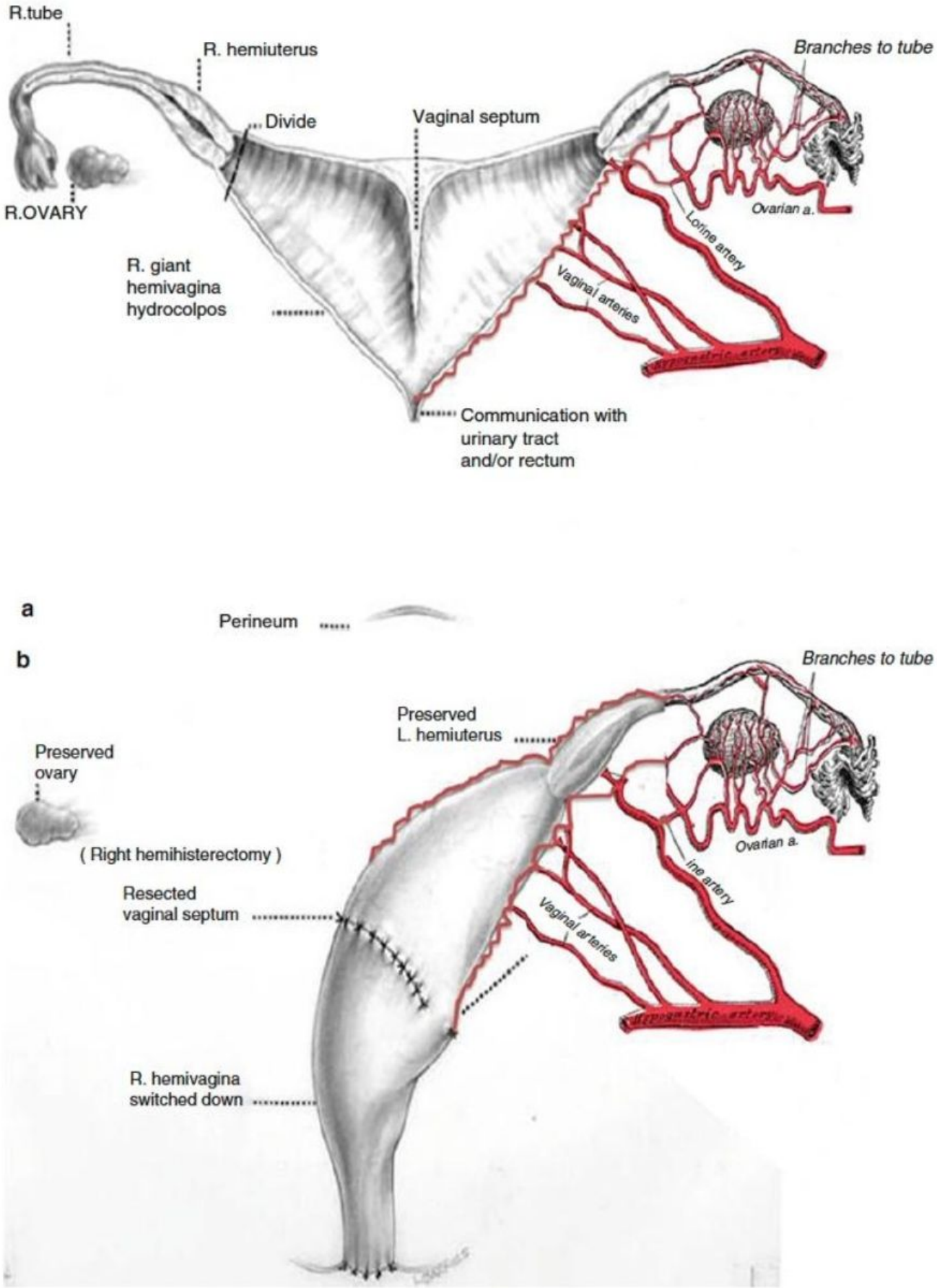


**Fig. 16.46** Schematic representation of the basic principles of a vaginal switch maneuver. (**a**) One hemiuterus will be amputated. The blood supply of that hemivagina is sacrificed being careful to preserve the blood supply of the other hemivagina and ovaries. The vaginal septum is resected and both large hemivaginas are tubularized together. (**b**) What used to be the dome of one hemivagina is pulled down to create the introitus

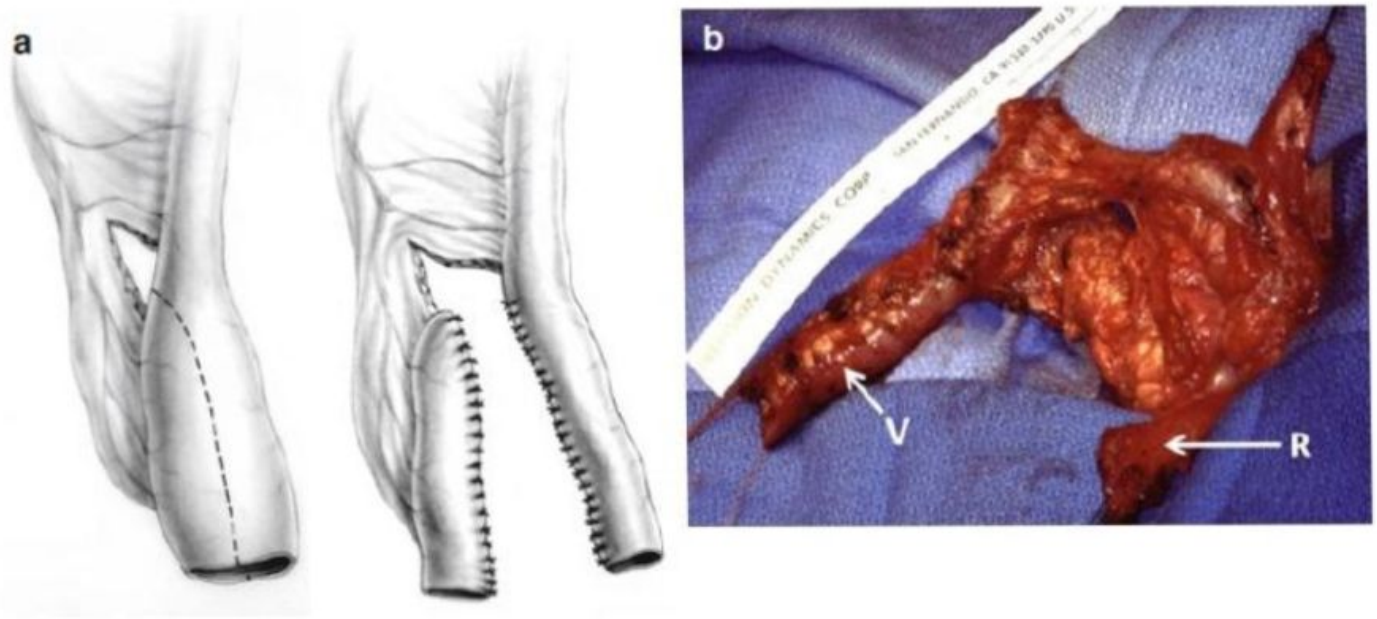


**Fig. 16.47** Vaginal replacement with part of a dilated rectum. (a) Diagram. (b) Intraoperative picture. *V* vagina, *R* rectum

vagina will depend on the preservation of the blood supply of the opposite hemivagina. If one can see that the distance between both hemiuteri is not long enough, then we have to separate both hemivaginas completely from the trigone and urinary tract as previously discussed.

The next possible scenario could be the case of a patient in whom we have gone through all the steps previously described, and her anatomy does not make her suitable to be repaired using a vaginal switch maneuver. The vagina(s) is too short and/or located too high in the pelvis. Under those circumstances, the patient will need a vaginal replacement.

### Vaginal Replacement

Vaginal replacement has been done by many authors through many years. Most authors including us prefer to use the colon [50–59]. Others used local tissues expanded with different methods [60–62].The amniotic membrane has also been used [63].

We have developed a significant experience in 130 cases suffering from cloaca that required vaginal replacement. Our order of preference in terms of tissue to be used for the replacements are rectum 50 cases, colon 44 cases, and finally, small bowel 36 cases.

### Vaginal Replacement with Rectum

There are several ways to replace the vagina with rectum. It all depends on the anatomic characteristics of the patient. If the patient has a very dilated rectum, conceivably, we can divide the rectum longitudinally, preserving the blood supply of both portions, the one that is going to be the neovagina and the other one that will remain as rectum (Fig. 16.47a). The vagina and the rectum are both tubularized. Both structures are rotated 90° in opposite directions to avoid the overlap of two suture lines, which is an important predisposing factor for the formation of a fistula (Fig. 16.47b). If the patient has no internal genitalia (rarely occurs), then the vagina is created only for sexual purposes and is not anastomosed to any internal structures. On the other hand, if the patient has an internal genitalia, then we have to perform an anastomosis between the upper portion of the neovagina (old rectum) and the original native short vagina.

The longitudinal incision of the rectum to create two separated tubular structures (vagina and rectum), preserving a good blood supply of both of them, is an interesting technical maneuver worth describing in detail.

Through all these years, we have learned that the rectum has an excellent intramural blood

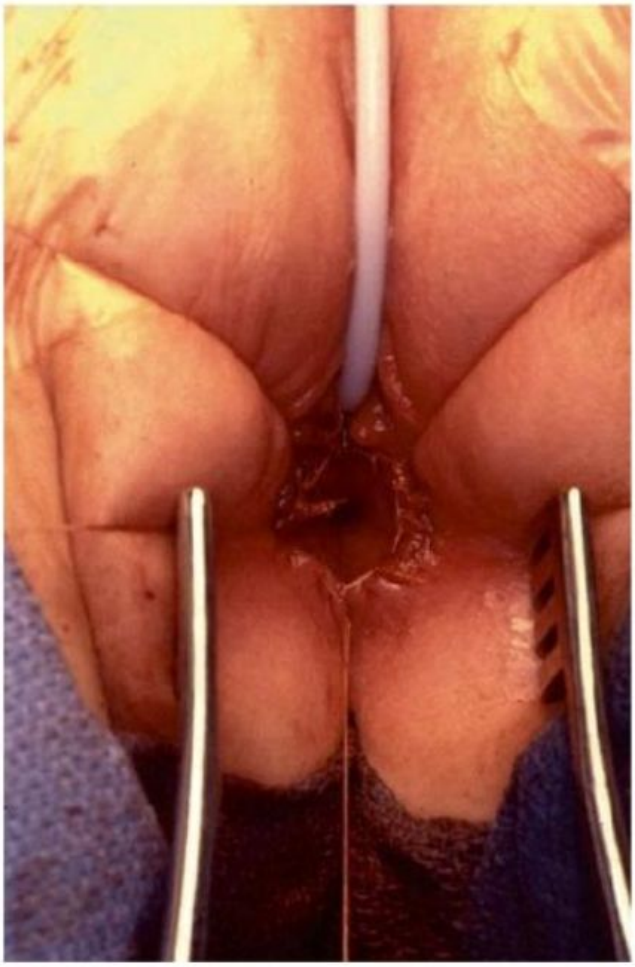

Fig. 16.48 Finished vaginal replacement

supply. It is perhaps the only hollow viscus of the human body that can be deprived, to a limit, of its extrinsic blood supply and still survives due to an excellent intramural blood supply, provided it maintains its continuity with a piece of colon with good blood supply. This is only possible if the surgeon manages to maintain intact the bowel wall. In other words, we can dissect and mobilize a significant length of rectum, burning and dividing the vessels that represent its extrinsic blood supply and still have a good blood supply, provided we perform the dissection as close as possible to the bowel wall but without injuring it and provided the upper rectum receives a good blood supply from inferior mesenteric vessels. Figure 16.47 shows the longitudinal division of the rectum and the tubularization. As can be seen, the blood supply of the rectum after this maneuver will depend entirely on the intramural blood supply. Obviously, the piece of rectum designated to replace the vagina must receive its blood supply from at least one or two of the inferior mesenteric vessels. The lower anastomosis is performed between the neovagina (rectum) and the neolabia. The perineal body is reconstructed in the usual manner as well as the rectum (Fig. 16.48).

In the case of a patient with a non-dilated rectum but with plenty of available length, we can plan on using the most distal part of the rectum as a neovagina and mobilize the upper rectum or sigmoid as a neorectum (Fig. 16.49a). In order to do that, it is imperative to learn to preserve the blood supply of the distal rectum. One must keep in mind that the mesentery of the rectum is completely different from the mesentery of the rest of the gastrointestinal tract. In the small bowel and colon, the mesentery reaches the bowel on the so-called mesenteric side only. It is very obvious that the rest of the bowel has no mesentery and no fat, just the seromuscular layer. The rectum, on the other hand, is surrounded by fatty tissue with vessels, which has received the name of mesorectum, and surrounds basically the entire circumference of the rectum. In order to preserve the blood supply of the distal rectum, it is necessary to create a plane of separation between the bowel wall itself and the fat tissue with vessels (Fig. 16.49b). The dissection between the mesorectum and bowel wall is carried out all around the rectal wall at the place where we previously planned that the rectum would be divided, leaving the distal part as the neovagina and the upper part as the neorectum. Prior to making the decision to divide the rectum, we must be absolutely sure that we have enough bowel length proximally to reach the perineum with no tension. We must keep in mind that occasionally the presence of a sigmoid colostomy created too distal may interfere with our plan.

Like previously mentioned, if the patient has no internal genitalia, the neovagina (original rectum) is closed blind on its upper end and will be used only for sexual purposes.

There are other ways to replace the vagina with rectum using techniques of combined longitudinal section with the use of the distal bowel (Fig. 16.50), depending on the specific anatomic circumstances.

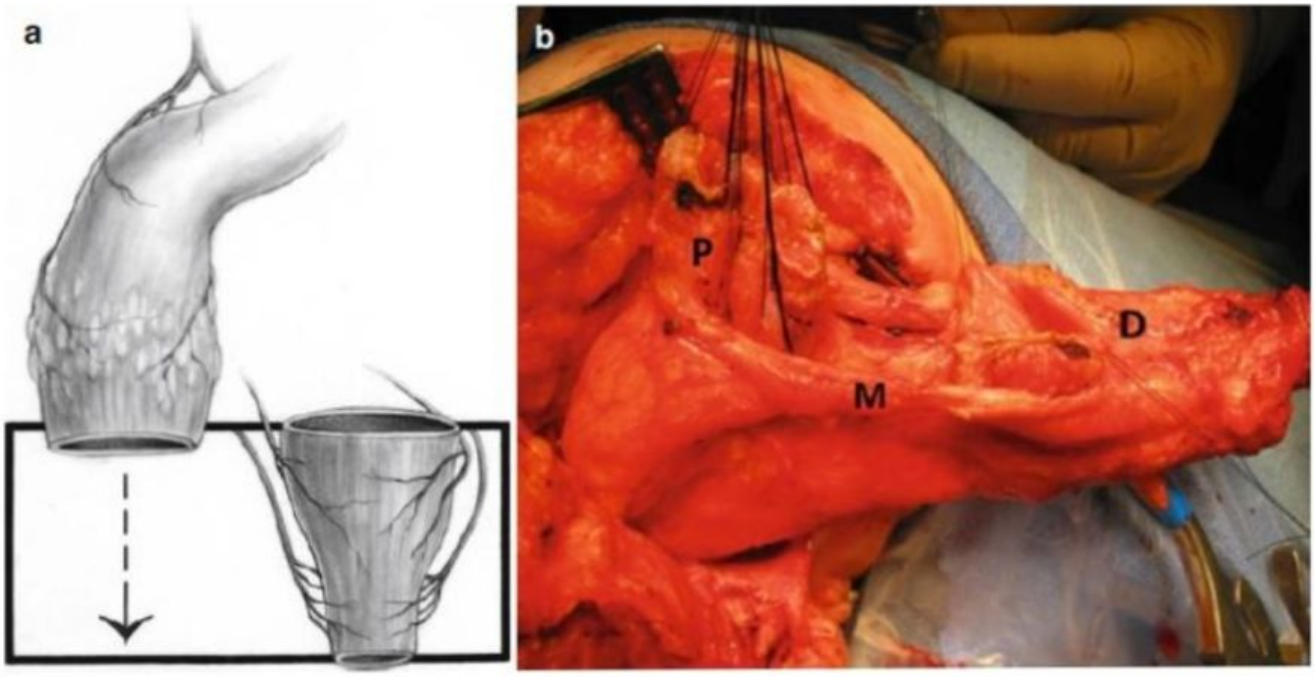


**Fig. 16.49** Vaginal replacement with the distal part of rectum, in patients who have plenty of rectosigmoid length. (**a**) Diagram. (**b**) Intraoperative picture. Dissecting the mesorectum from the rectal wall. *D* distal rectum, *M* mesorectum, *P* proximal rectum

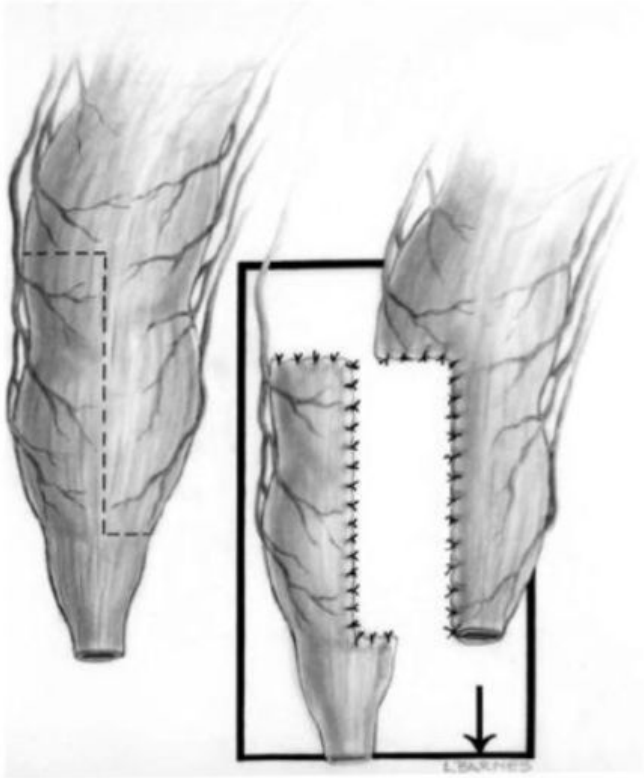

**Fig. 16.50** Vaginal replacement with rectum. Other technical alternative

If the rectum is not adequate to be used for vaginal replacement, then our next choice is the colon.

### Vaginal Replacement with Colon

In the past, we have typically used the sigmoid colon when available (Fig. 16.51). Frequently, the colostomy interferes with this, and that is why, in patients with cloacas who may require vaginal replacement, a higher type of colostomy has been recommended by Hendren [31]. Lately, however, in spite of the patients having a colostomy located in the left lower quadrant, we have found that the left transverse colon and the descending colon have a nice vascular arcade of vessels that make that part of the colon ideal for vaginal replacement (Fig. 16.52) (Animation 16.5). Therefore, we have chosen to take the colostomy down and use that part that used to be the colostomy to replace the vagina. The pedicle of the left colon graft often reaches the perineum more easily than the sigmoid would.

### Vaginal Replacement with Small Bowel

This is our last choice, because it is our impression that the blood supply of the small bowel is more delicate and more prone to suffer from occlusion with a mild twist of the pedicle of the bowel that has been pulled down. When we decide to use the small bowel, we prefer to use

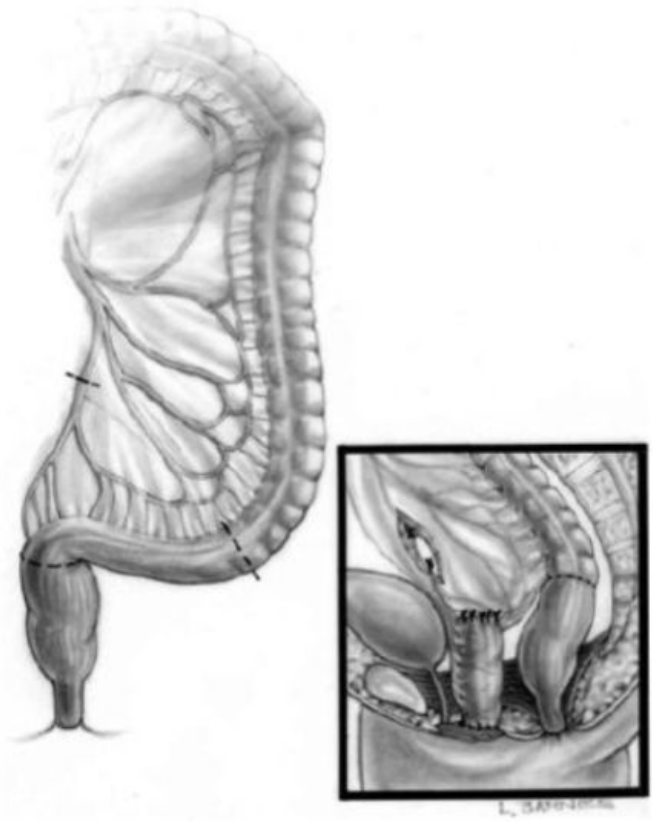

**Fig. 16.51** Diagram of vaginal replacement with sigmoid colon. *Dashed lines* show the limits of bowel resection and vessel ligation

terminal ileum. We made an interesting observation; the length of the mesentery at the cecum represents the length of the superior mesenteric axis (Fig. 16.53). If one takes the terminal ileum about 10 cm proximal to the ileocecal valve, it is easy to see that the mesentery is longer than at the cecum, and if we go even more proximal, the mesentery is even longer. The longest mesentery of the small bowel seems to be located about 15 cm proximal to the ileocecal valve, and because of that, we thought that this part of the bowel would be ideal to be used for vaginal replacement. Accordingly, we observe the blood supply to decide which vessels divide and which ones to preserve to be sure that the bowel reaches the perineum (Fig. 16.54). Again, one must be extremely meticulous in the way we pull the bowel, because a little twist will produce ischemia and we may lose the graft. If the patient has internal genitalia, then, as previously mentioned, the small bowel (neovagina) is to be anastomosed to the internal genitalia and to the new labia. On the other hand, if the patient has no internal genitalia, the bowel in the upper part must be closed blind, and we have only to do an anastomosis to the neolabia (Fig. 16.55).

### Cloacas with Extremely Long Common Channels

When the endoscopy allows us to see that the patient has an extremely long common channel (more than 5 cm), it is conceivable that the separation of the structures (rectum, vagina, and urinary tract) could be done easier through a laparotomy rather than through the perineum or posterior sagittally (Animation 16.5). In these cases, the anatomy is like the one illustrated in Fig. 16.5. The rectum opens either in the bladder neck or in the trigone, and two little hemivaginas open also in the trigone or in the bladder neck in a perpendicular fashion (with no common wall). The ureters also open in that particular area. These patients therefore have about five important structures opening in the same area of the urinary tract. The separation of these structures usually leaves the patient with no bladder neck or with a damaged one. Good clinical judgment and experience are required to make a decision about reconstructing the bladder neck or closing it on a permanent basis. In the last circumstance, the patient will need a vesicostomy, and eventually, she will require a bladder augmentation and a Mitrofanoff type of procedure. On the other hand, frequently we separate these structures carefully and we are left with a bladder neck that looks like it can be reconstructed successfully, and we have cases in which this reconstruction has been successful.

The rationale that supports the idea of approaching these patients through a laparotomy first is that we can leave intact what used to be the common channel as a conduit for intermittent catheterization; that will work beautifully, provided the bladder neck is reconstructed adequately.

It has been our impression that, in general, patients with cloacas have a good bladder neck. If this bladder neck is preserved or reconstructed adequately, these patients have no problems holding urine in the bladder. The main type of bladder malfunction that we see in these patients is incapacity to empty. Most cloaca patients operated on

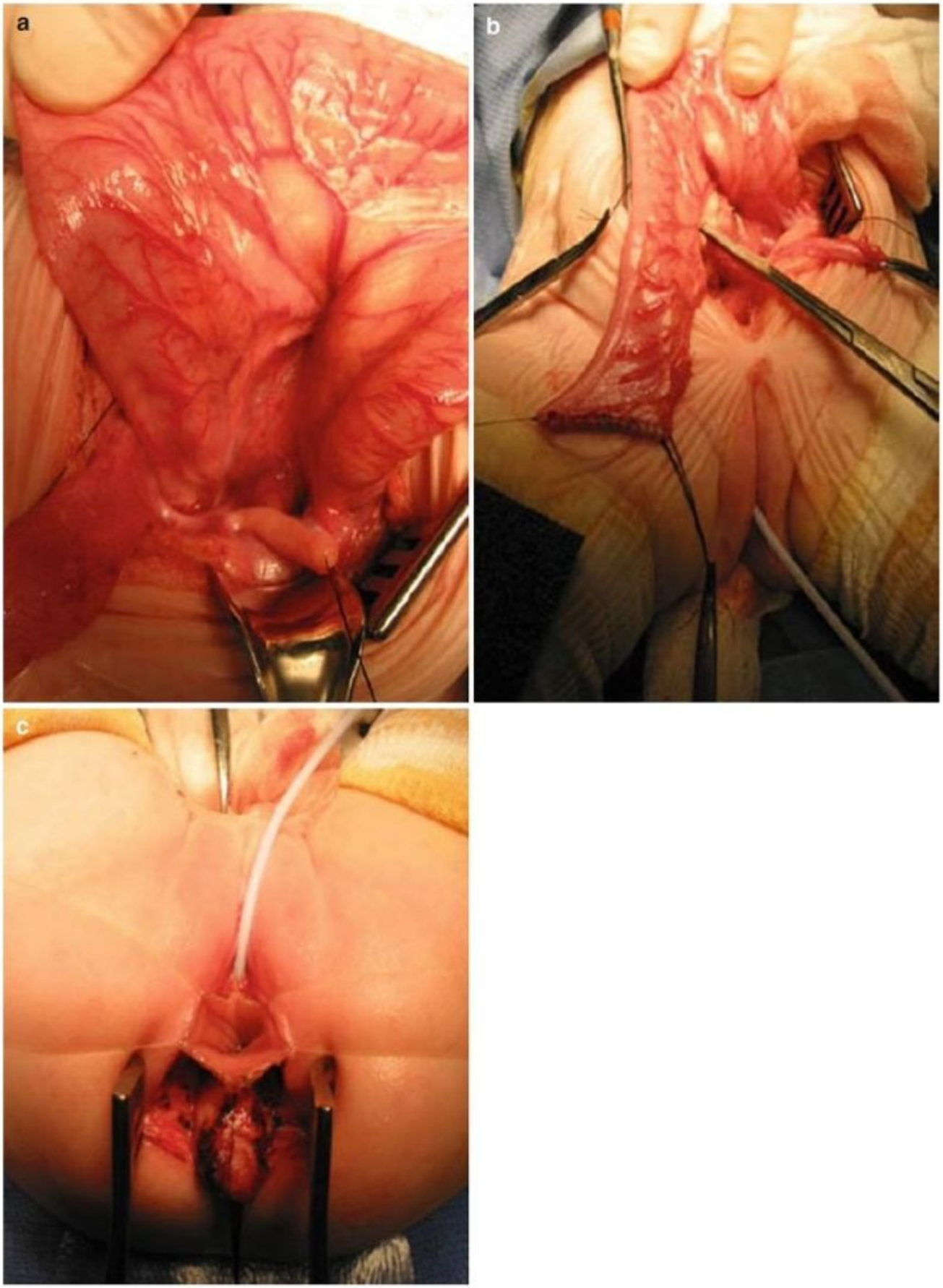


**Fig. 16.52** Intraoperative pictures of a vaginal replacement with descending colon. (**a**) Studying and selecting the vessels that must be preserved. (**b**) Measuring the length of colon. (**c**) Creation of the introitus

**Fig. 16.53** Vaginal replacement with small bowel. The mesentery of the small bowel is longest in the terminal ileum, about 15 cm proximal to the ileocecal valve. *Arrows* show the direction of the traction

have a smooth, large, floppy bladder with a good bladder neck. They cannot empty the bladder; therefore, during the period of filling up of the bladder, the patient may remain completely dry in the underwear. Once the bladder is completely full, they start dribbling urine as an overflow type of phenomenon. This makes these patients ideal candidates for intermittent catheterization. However, in most types of cloacas, the common channel is surgically manipulated and, therefore, sometimes is not regular and smooth enough to allow a successful and easy catheterization.

In these extremely high types of cloacas, with a very long common channel, we insist on leaving the common channel untouched and to approach the patient initially directly through the abdomen, preserving intact the common channel, to be used as a conduit for catheterization. The

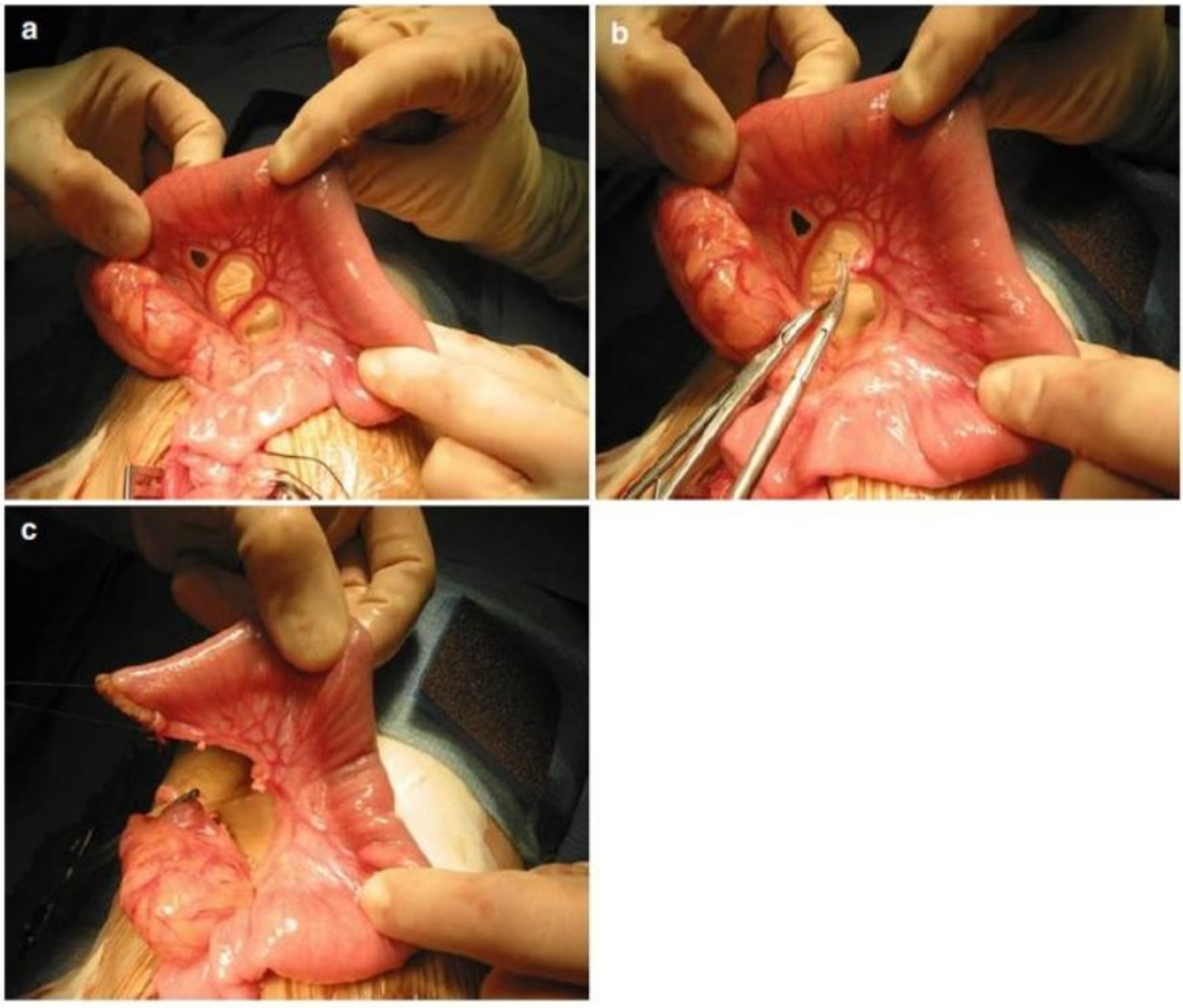


**Fig. 16.54** Intraoperative picture. Vaginal replacement with small bowel. Selecting the vessels to be preserved. (**a**) Selecting the vessels. (**b**) Dividing the vessels preserving the arcade. (**c**) Dividing the bowel

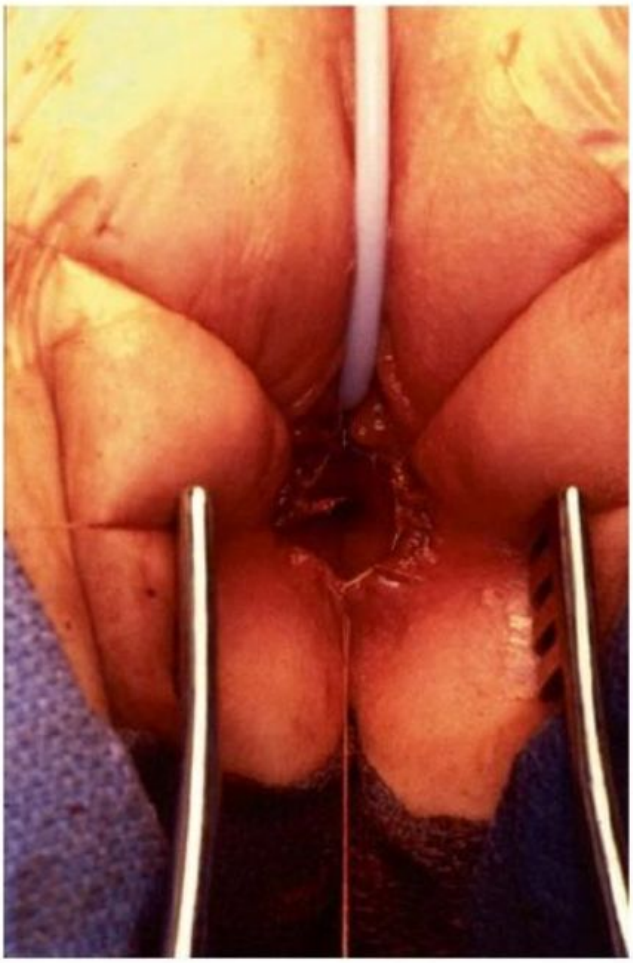

Fig. 16.55 New introitus made with small bowel

higher the malformation, the shorter the common wall between the bladder and vaginas, which is why it is relatively easy to separate vaginas and urinary tract through a laparotomy.

Occasionally, the laparotomy is indicated because of a very high rectum and not so much because of the urogenital tract. Sometimes, the rectum opens very high in between both hemivaginas and cannot be reached posterior sagittally, even after the total urogenital mobilization has been performed, and, therefore, one has to go into the abdomen just to look for the rectum and pull it down.

The posterior sagittal approach, as well as the total urogenital mobilization, has been adopted by many pediatric surgeons, with variable results [64–73]. Other types of repair have been also advocated, although with very few cases [74–80]. An interesting report from China was recently published [81]; the authors claimed that they were able to use serial mechanical dilatations of the common channel and eventually they were able to create a urethra and vagina from the dilated common channel!!

At the end of the cloaca repair, we must decide to leave our patient with a Foley catheter through her native urethra, with a suprapubic cystostomy or with a vesicostomy. In general, we make that decision based on the following principles:

- Foley catheter: Patients with a common channel shorter than 3 cm, repaired via posterior sagittal approach, with a good sacrum, and no tethered cord. We assume that after 2 weeks of having the catheter, the patient will be able to void satisfactorily.
- Suprapubic cystostomy: Patients who required a laparotomy and we assumed that will need the catheter for a period of time shorter than 3 months. It is very useful because it allows us to evaluate the bladder function and make decisions, during the following 3 months post operation.
- Vesicostomy: Patients with poor renal function and or tethered cord, poor sacrum, vesicoureteral reflux, megaureter, hydronephrosis, and single kidney.

#### 16.1.1.5 Postoperative Care

Since we are dealing with a spectrum of defects, the postoperative care represents also a wide spectrum. A patient who has a cloaca type 1 can eat the same day and go home the next day after surgery. A patient who has a common channel shorter than 3 cm also can eat the same day of surgery and stay in the hospital 48 h. A patient who requires an 8–12-h operation may remain intubated overnight in the intensive care unit. We have found that little babies subjected to long surgical procedures (more than 6 h) may need to remain intubated and recover in the intensive care unit. It is our experience that these patients retain a lot of fluid in their bodies and may have respiratory problems. It takes several days for them to get rid of the excessive third-space sequestered fluid.

Little babies subjected to long operations should not be treated in institutions without an adequate, third-level intensive care unit, with experts in the management of these types of critically ill patients.

Some of these patients already have significant kidney damage and elevated creatinine at birth that

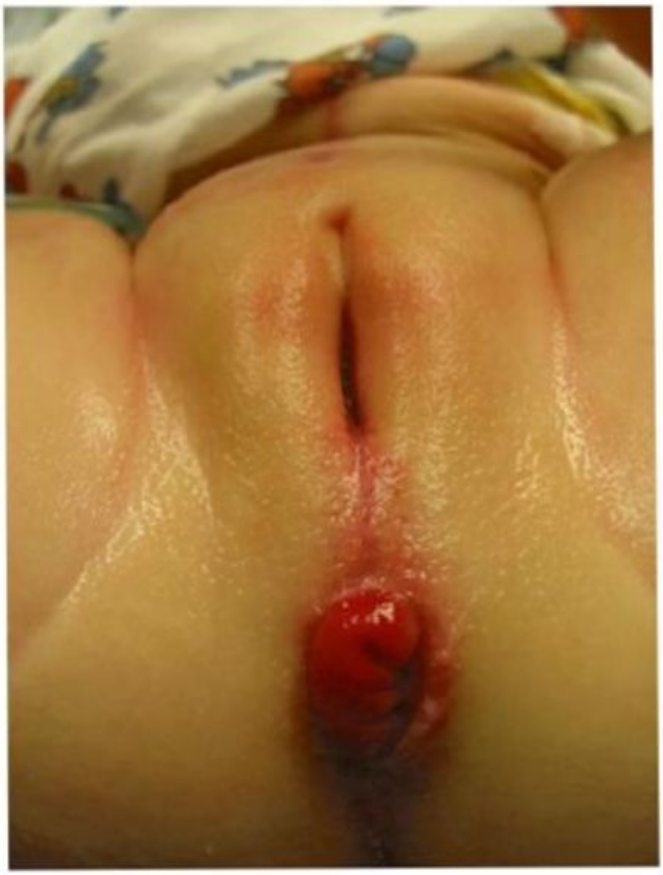

Fig. 16.56 Picture showing a significant rectal prolapse in a case with a repaired cloaca

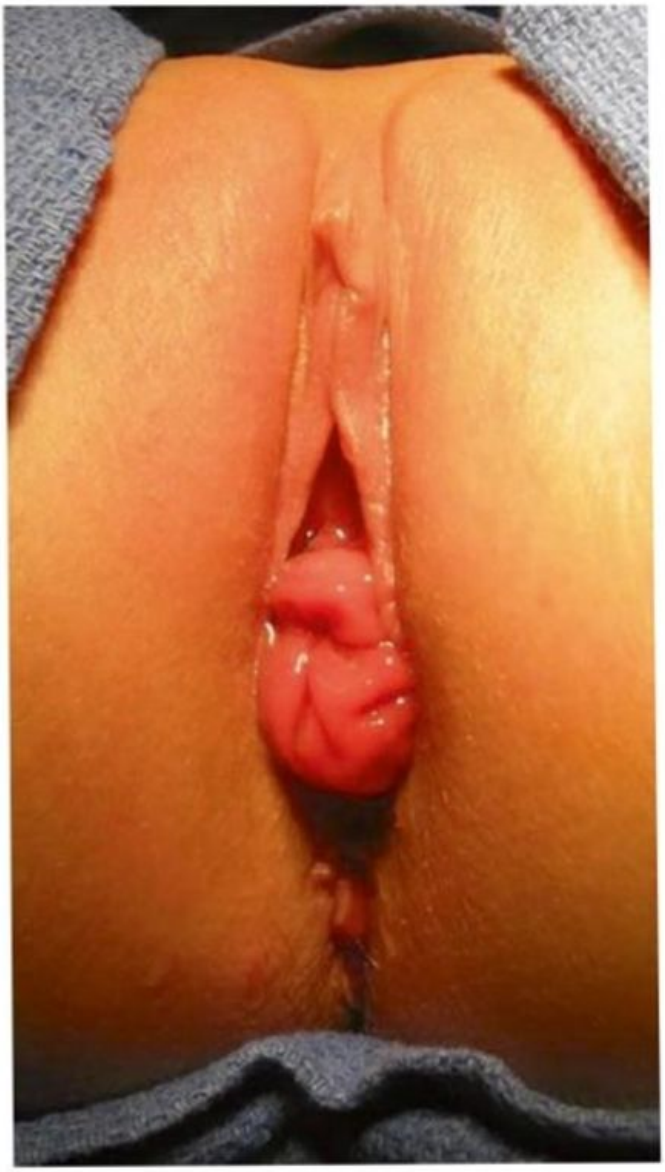

Fig. 16.57 Prolapse of neovagina created with bowel

requires special care by an expert team of nephrologists. We have seen that after these operations, the creatinine becomes elevated even without any evidence of urinary tract obstruction.

Sometimes, we repair these malformations and pull the colostomy down at the same time. In those cases, the patients remain with nothing by mouth, receiving parenteral nutrition for 7–10 days. The Foley catheter remains in for 2–3 weeks until the urethral orifice is visible, and a catheter could be reinserted if necessary.

Two weeks after surgery, the protocol of anal dilatations is started like we do in all the other types of malformations. At the time of colostomy closure, the patients need a vaginoscopy and cystoscopy to be sure that the repair was successful. In the event of finding any problems with the repair, the colostomy closure should be canceled, and we should focus on repairing the vagina or urethra as necessary.

Sometimes at the time of colostomy closure, we find that the patient has a significant rectal prolapse (Fig. 16.56). If that is the case, we repair the prolapse and postpone the colostomy closure at least for 1 month. Occasionally, when the vagina has been replaced with colon, rectum, or small bowel, one may also find some degree of bowel mucosa prolapse as part of the neovagina (Fig. 16.57). Again, this should be treated before the colostomy is closed because it can be done in a single-day admission and because the perineum is completely clean. A long, narrow stricture of the vagina requires a full reoperation. Fortunately, that is unusual in our cases.

Sometimes, we find that the urethra is difficult to catheterize. In such a case, it might be appropriate to open a vesicostomy and delay the need for intermittent catheterization until the baby is older.

At the time when we were doing repairs without the total urogenital mobilization, 10 % of our patients suffered from urethrovaginal fistulas.

Usually, these fistulas were located near the bladder neck and produced urinary incontinence, and therefore, the patient required a total reoperation. The advent of the total urogenital mobilization allowed us to have no more of these complications [47].

### 16.1.2 Urologic Concerns

The main long-term problems of patients with cloacas are usually urologic. These patients do not die from the cloaca malformation, but rather from major cardiac anomalies early in life or from kidney failure later on. The urologic concerns of patients with cloaca have been recognized for a long time [82]. There are four wonderful publications related with the long-term urologic and renal outcome of patients with cloacas [83–86]. The authors presented series of 12–64 cases that allowed them to reach valid conclusions. From this experience, as well as ours, it becomes clear that treatment of cloacas must be done by experienced pediatric surgeons and pediatric urologists. Protecting the kidneys must be the top priority from the first day of life. In addition, it is clear that these patients must be followed to be sure that the bladder function does not deteriorate and affect the kidneys.

In addition, two publications alerted us about the possibility that the total urogenital mobilization could provoke nerve damage that could result in poor bladder function [87, 88]. Based on our experience, we believe that patients with normal sacrum, no tethered cord, and with a common channel shorter than 3 cm should not have serious urinary problems, provided the total urogenital mobilization has been performed in a meticulous way. A similar experience was published by others [89–93].

We have learned that when a patient is born with hydronephrosis, we must anticipate that the patient already has a significant degree of kidney damage. Even if they have a normal creatinine, one must expect that at some point later in life, the patient may develop kidney failure. This is particularly true when the patient has a single kidney and hydronephrosis at birth. Therefore, one must be rather obsessive about the protection of the kidneys, particularly if it is a single one. We must take care of the reflux, obstruction, and/or infection. Frequently, we are confronted with the problem of a little baby with megaureters, an "inadequate bladder," and severe vesicoureteral reflux. For that kind of patient, we prefer to open a vesicostomy and wait until the ureters decrease in size, the bladder becomes more compliant, and at that point, the reimplantation of the ureter may have more chances of success. The reimplantation should be done after the bladder has been urodynamically studied to determine its functional characteristics. We also must determine the mechanism of emptying of the bladder, either because the bladder empties spontaneously or needs intermittent catheterization.

Real ureterovesical obstructions without hydrocolpos rarely occur in patients with cloacas. In the presence of a true ureterovesical obstruction, during the repair of a complex cloaca in a baby, with megaureter and a bladder with questionable function, we take care of the obstruction, creating a wide refluxing ureterovesical anastomosis and a vesicostomy. That represents the best protection for the kidney. We then wait until the ureter decreases in size, the patient grows up, and the bladder becomes more compliant. An evaluation of the bladder function then will allow one to determine the best course of action for the patient. We do not see a reason to do bladder augmentations and Mitrofanoff procedures in babies before 3 years of age, particularly, if the patients already have a certain degree of kidney damage. So far, 46 patients already underwent a bladder augmentation and a Mitrofanoff type of operation. Twenty-seven of those were done by the senior author and 19 by our pediatric urology colleagues.

These are patients for life and require a team of urologists, nephrologists, gynecologists, and colorectal pediatric surgeons to follow them.

The patient's bowel function will depend very much on the characteristic of the sacrum and spine. At the age of three, if the patient has no bowel control, the patient is offered our bowel management program (see Chap. 20). We insist that all patients after 3 years of age should be clean of stool in the underwear, as well as dry of urine.

### 16.1.3 Gynecologic Concerns

The association of anorectal malformations with defects of the internal and external genitalia has been described before [6, 94–96].

The negative implications of the hydrocolpos as well as the serious consequences of not draining it have been reported [97–103]. We cannot overemphasize the importance of suspecting, diagnosing, and draining a hydrocolpos as early as possible.

For the first few years of life (until puberty), the patients usually have no gynecologic problems. However, when they reach puberty, one must be very careful in following these patients. If the patient develops pubic and axillary hair and the breasts start to develop but no menstruation, one must suspect that there is some sort of obstruction or absence of one or both of the Müllerian structures. This is particularly true when the patient has no menstruation and episodes of abdominal pain with monthly exacerbations that reflect the presence of trapped menstrual blood. At that point, the patient needs an ultrasound of the pelvis and/or an MRI.

From the first 27 patients with cloacas from our series, who reached puberty, 7 of them required an emergency laparotomy, due to trapped menstrual blood in the peritoneum which formed pseudocysts [104]. After that experience, our routine now is to check the patency of the Müllerian structures early in life, whenever we have the opportunity to be in the abdomen of these patients. We do this by putting a no. 3 feeding tube through the fimbriae of the Fallopian tube and injecting a saline solution to confirm that it comes out through the vagina (Fig. 16.58). If one of the sides is obstructed and the other is patent, we prefer to resect the obstructed part, since the patient will do well with only one Müllerian structure. When both are obstructed, we decide to preserve both of them but tell the parents about this and follow the patient closely, particularly when they reach puberty.

Assuming that the patient starts menstruating normally in puberty, the next step would be to think about sexual function. When the patient is ready to develop sexual activity, it is important to perform an examination under anesthesia. The purpose of this is to determine whether or not the introitus is large enough for the patient to have normal sexual activity. Sometimes we find a fibrous ring in the area of our anastomosis of the vagina to the neolabia (Fig. 16.59). If the vagina has not been injured and does not have a long, narrow stricture, usually the repair of this ring is an easy, ambulatory

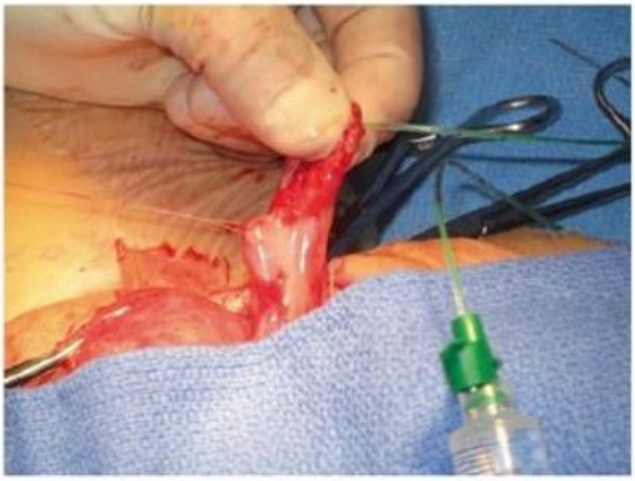

**Fig. 16.58** Picture showing the irrigation of fallopian tube to confirm its patency

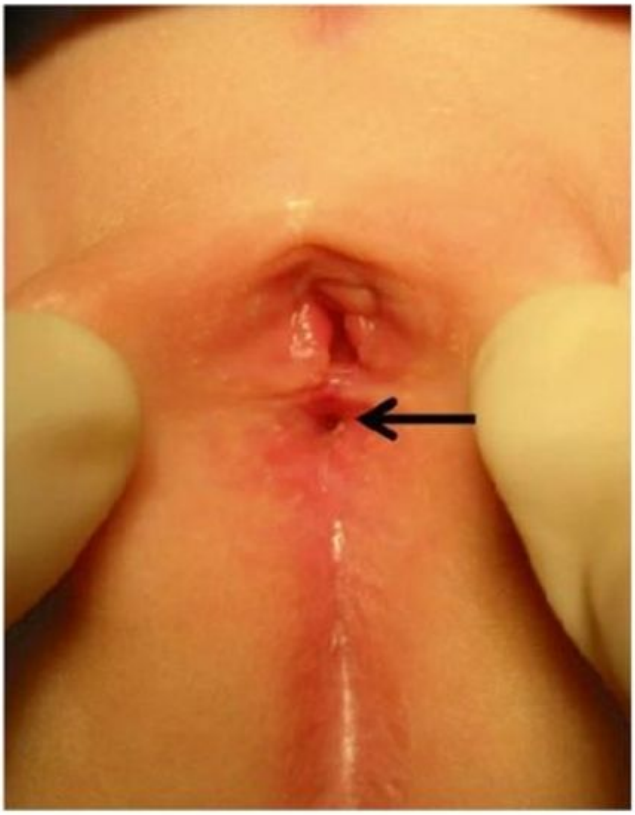

**Fig. 16.59** Picture showing a strictured vaginal orifice – *arrow* on the vaginal orifice

operation that should be done prior to sexual activity to avoid unpleasant experiences.

Two hundred and fourteen of our cloaca patients are older than 18 years. From those, 48 patients are all having normal sexual activity; 45 of them claim to have orgasms; another 3 patients tried sexual activity and have been unsuccessful; and 7 patients became pregnant and delivered babies by cesarean section.

The long-term follow-up of all cases suffering from congenital anomalies is extremely valuable. A new chapter on the gynecologic problems of patients born with cloacas is in the process of being integrated. Although, with small numbers, several important papers have been published concerning the long-term psychosexual outcome of patients born with cloacas and urogenital sinus [105–110]. Other unexpected problems may appear later in life in these patients, including tubal pregnancy [111] and tubo-ovarian abscess [112]. We have no experience with tubal pregnancy, but we have seen several cases of adolescents born with cloacas, repaired by us, who suffered from recurrent tubo-ovarian abscess that eventually resulted in a resection of the tube.

Vaginal septum with two hemiuterus occurred in 2 % of our cases with perineal fistula, 6 % of cases with vestibular fistula, and 60 % of our cloacas. It is extremely important to alert the parents of these babies about the future obstetric implications of those anomalies including premature labor and miscarriage [113–115]. Patients born with a cloaca must be followed for life by gynecologists with special interest and experience in this type of problem. The pregnancies in these patients are by definition "high risk."

Finally, malignancy of a cloacal remnant [116] has been reported, as well as originating from a neovagina [117]. In addition, the occurrence of ulcerative colitis in the neovagina has been reported [118, 119]. In our series, we have seen two cases of cancer and two of inflammatory bowel disease, at the colon used to replace the vagina.

### 16.1.4 Reoperations

Secondary procedures in cloacas are very common in our experience. This is perhaps a reflection of the nature of our center, receiving patients from all over the country and from other countries as well. From our experience of 531 cases of cloacas, 97 of them are actually secondary procedures. This high frequency of complicated cases previously operated also represents the fact that the surgical maneuvers described here to repair cloacas, perhaps, are not highly reproducible. Cloacas are not an exception when we affirm that children with congenital malformations "have a single opportunity" to be repaired, meaning that if that first attempt is not successful, the functional prognosis changes negatively after a secondary procedure.

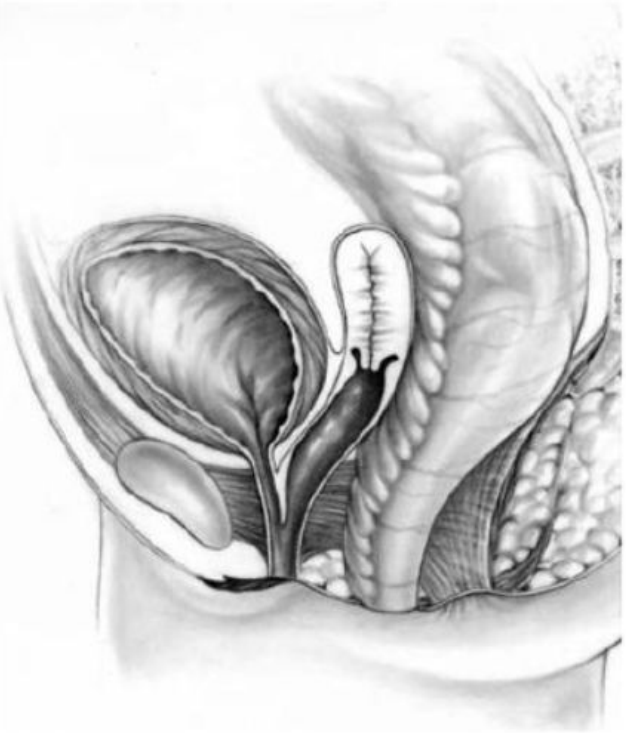

**Fig. 16.60** Diagram of a persistent urogenital sinus. The patient was born with a cloaca and underwent a repair of the rectal component of the malformation. The urogenital sinus was left untouched

The most common reoperation that we perform in patients with cloacas consists in repairing persistent urogenital sinuses (Fig. 16.60). These patients were born with a cloaca and underwent an operation at another institution to repair only the rectal component of the malformation. The patients were then left with a persistent urogenital sinus. Interestingly, from all the persistent urogenital sinuses operated by us, only one had come to us with a previous diagnosis of a cloaca. In other words, the surgeons were unaware of the fact that the

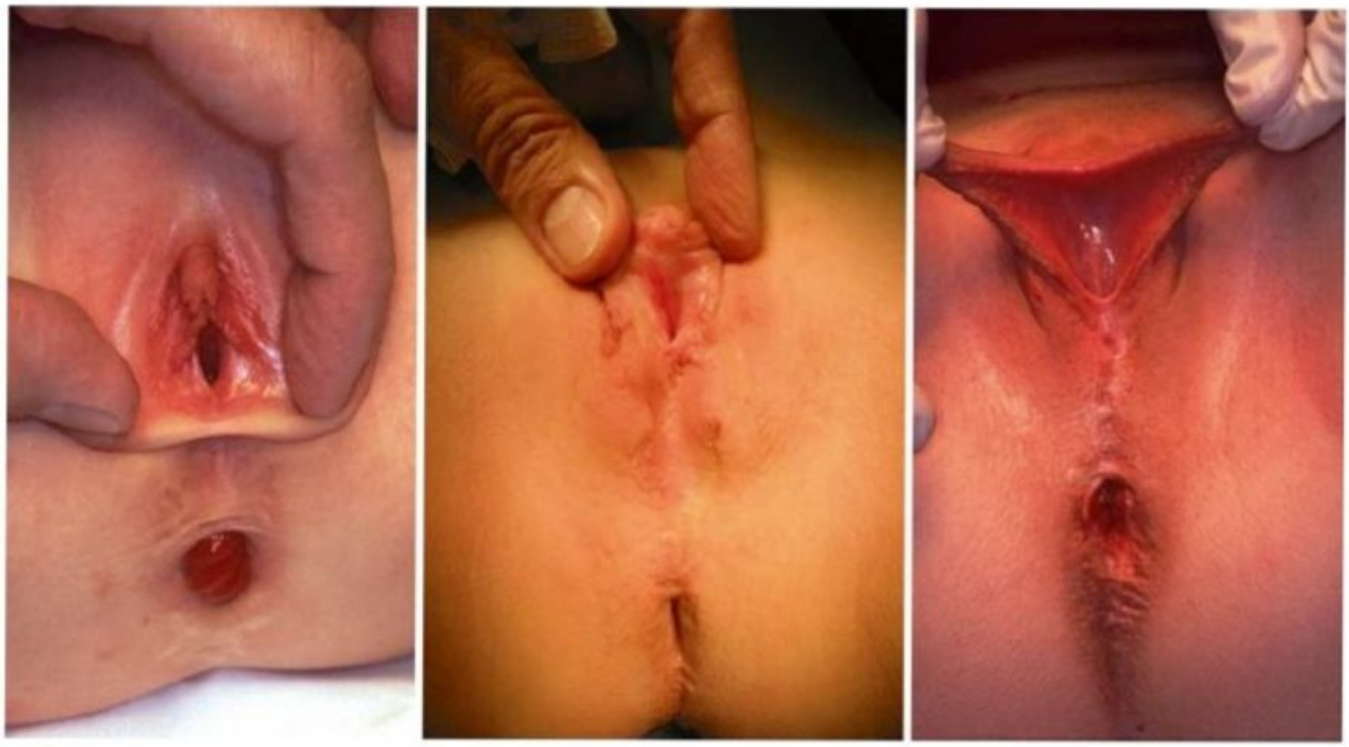

Fig. 16.61 Pictures of four patients with persistent urogenital sinus

patient had a cloaca; they tried to repair the malformation, but they only ended up working with the rectum and left the urogenital sinus untouched. On the other hand, all other cases came to us with the diagnosis of "rectovaginal fistula." In other words, the surgeons did not make the diagnosis of a cloaca and operated on the patient thinking that the patient had a rectovaginal fistula. The word cloaca was never mentioned in the medical records of the patients, and consequently, the surgeons repaired only the rectal component of the malformation. They did not perform a vaginoscopy or cystoscopy, pulled the rectum down, and left the urogenital sinus untouched. Amazingly, these patients continued their life, recovered from those operations, and become aware of their persistent urogenital sinus later in their lives. We actually had several adult patients with a persistent urogenital sinus. One of them had been married for 2 years. She came to us because of incapacity to perform sex satisfactorily. Our examination disclosed a very narrow persistent urogenital sinus (Fig. 16.61).

The cases of a persistent urogenital sinus labeled as "rectovaginal" fistula were more common at the beginning of our practice, which reflected (from our point of view) the fact that surgeons were thinking of "rectovaginal" fistulas and not about a cloaca. Looking at the literature, one can find a very obvious shift in the apparent frequency of "rectovaginal" fistulas and cloacas. Prior to 1982, the literature had many reports of "rectovaginal" fistulas and almost no mention of cloacas. In retrospect, we know that most of those "rectovaginal" fistulas operated and reported in the literature in the past were actually cloacas or rectovestibular fistulas. We know that for sure because of the number of patients that we received with persistent urogenital sinus and also because in many of those patients with "rectovaginal" fistulas, when we examined them, we found the original pocket, or sinus, remnant of the original vestibular fistula (Fig. 16.62).

As time went by, the number of persistent urogenital sinus that we saw decreased, fortunately, and we interpreted that as a manifestation of the surgeons becoming more knowledgeable about the existence of this condition. In other words, their index of suspicion that they were dealing with a cloaca increased through time.

Now we know that real, congenital, rectovaginal fistulas are extremely rare; we have seen only 7 cases in over 1,000 female patients with

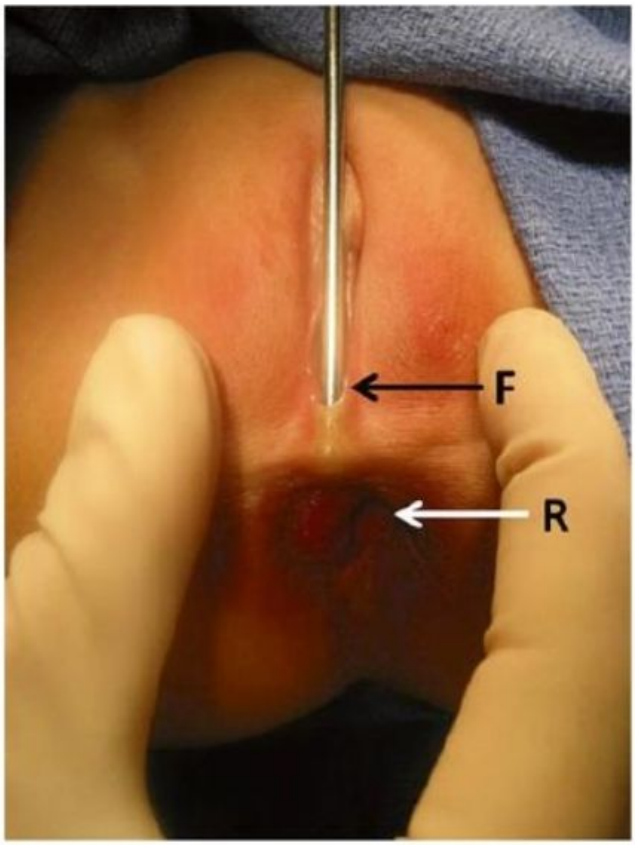


**Fig. 16.62** Picture of the genitalia of a patient born with a vestibular fistula who underwent an abdominoperineal operation under the misdiagnosis of "rectovaginal fistula." The original location of the fistula in the vestibule is clearly seen. *F* fistula, *R* rectum

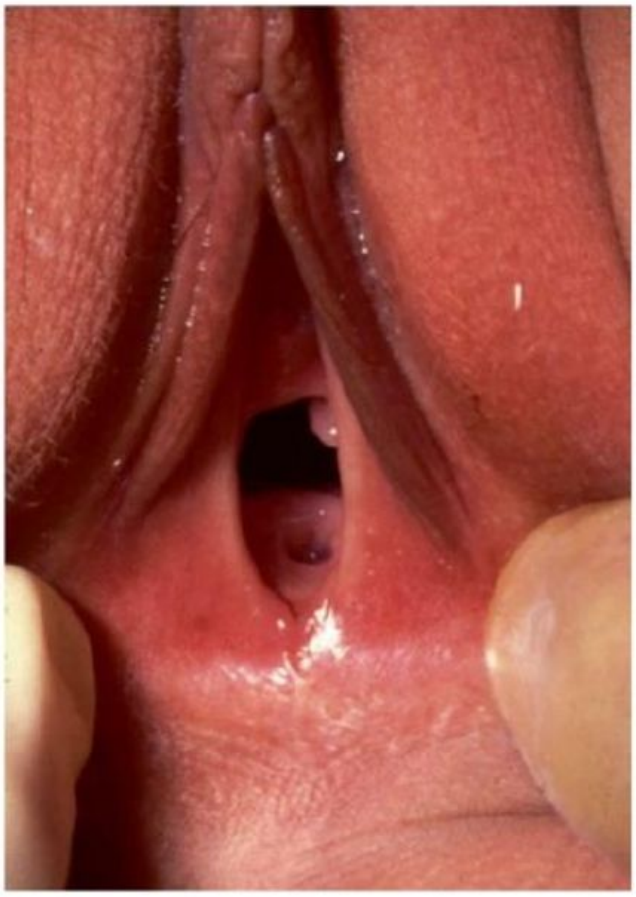

**Fig. 16.63** Picture of a case with a real congenital rectovaginal fistula. The rectal orifice is located inside the vagina (above the hymen)

anorectal malformations. In cases of rectovaginal fistulas, the rectal orifice must be located above the hymen (Fig. 16.63).

#### 16.1.4.1 Persistent Urogenital Sinus

Figure 16.60 shows a diagram of a persistent urogenital sinus. The rectum has been pulled down; frequently it is mislocated as shown in the figure, and the urogenital sinus persists untouched. As the patient reaches adolescence, her first manifestation is menstruation "through the urethra." Sometimes, the surgeons, during the repair of the rectal component of the malformation, damage the vagina, producing an acquired vaginal atresia. In that case, when the patient reaches puberty, she will accumulate menstrual blood (hemato-metrocolpos) and suffer from severe abdominal pain with monthly exacerbations. Figure 16.61a–c show the external appearance of the perineum of several patients with persistent urogenital sinuses.

The diagnosis is a simple one and is done by inspection. Vaginoscopy and cystoscopy will allow us to recognize the length of the common channel and the characteristics of the vagina(s), which, as previously mentioned, will allow us to plan on the magnitude of the operation and the possibilities of requiring a vaginal replacement. We have been repairing these malformations without a protective colostomy. However, as mentioned in Chap. 7 about bowel preparation, we are very strict in the cleaning of the entire gastrointestinal tract with GoLYTELY.

The patient is taken to the operating room with a central line already inserted and with the bowel completely clean. Most of the time, the persistent urogenital sinus can be approached posterior sagittally first, because of the length of the common channel. The patient is placed in the prone position with the pelvis elevated, and the skin of the genitalia, perianal area, and perineum is washed, prepped, and draped in the usual

manor. Multiple 5-0 silk stitches are placed at the mucocutaneous junction of the anal orifice. Frequently, this is strictured and/or mislocated. Applying uniform traction on those multiple 5-0 silk stitches, a posterior sagittal incision is created, running from the middle portion of the sacrum to the anal verge. The incision continues as a "hockey stick type" of incision around the anus, using the needle-tip cautery, changing from cutting to coagulation to provide meticulous hemostasis. The posterior sagittal incision continues deeper, going through the skin, subcutaneous tissue, parasagittal fibers, muscle complex, and the levator muscle. Applying uniform traction to the multiple silk sutures allows us to recognize the posterior rectal wall easily. The lateral walls of the rectum are also dissected applying uniform traction; all this is done with the needle-tip cautery. Finally, the anterior rectal wall is also dissected. Depending on the type of procedure that the patient previously had, one may find real rectum or colon. It is not uncommon to find that the previous surgeons resected the original rectum of the patient and pulled colon from inside the abdomen, down to the perineum. The reason for doing that is either because they used to perform an endorectal type of operation (Soave) which, by definition, sacrifices the rectosigmoid and then they pulled the colon from inside the abdomen (through the seromuscular cuff of the rectosigmoid left in situ) down to the perineum. Other times, perhaps more frequently, the surgeons open a colostomy too distally, and when they tried to repair the malformation, they found that the available length of rectosigmoid from the mucous fistula to the end of the rectum, where it connects to the urogenital tract, was too short. We have learned to preserve that piece of rectum, but many surgeons simply resect that part of the rectum and take the colostomy down, to save time and to make the procedure easier.

One can recognize whether or not we are dealing with colon or rectum. In the case of colon, one can identify the mesentery surrounding the colon. We must be careful in preserving the mesentery to avoid devascularization of the bowel, because the preservation of the blood supply is very important, particularly if we decide to use colon for vaginal replacement, when indicated. Other times, one finds the original rectum characterized by the absence of mesentery. As previously described, the rectum is surrounded by fat with vessels, which is the so-called mesorectum that does not have the characteristics of the mesentery of the colon. Most of the time, the dissection of the rectum in a reoperation is simpler than what we have anticipated. It may become very difficult in those patients who previously suffered from catastrophic complications such as abscesses, dehiscences, or retractions. In such cases, the amount of fibrosis surrounding the rectum can be extraordinary. When those complications did not occur in previous operations, the dissection is straightforward and relatively simple. We cannot overemphasize the importance of applying uniform traction while dissecting delicate structures, particularly in a reoperation. The dissection of the rectum must continue all the way up to the supralevator space. Frequently, we open the peritoneum during this dissection. Once the rectum has been fully dissected, we are ready to continue our incision more anteriorly. The perineal body is divided with needle-tip cautery, and the incision goes deeper until we identify the posterior wall of the vagina. Most of the patients that we have repaired with persistent urogenital sinus have a reachable vagina through the posterior sagittal approach. Once the rectum has been dissected and moved out of the way, the posterior vaginal wall is perfectly visible. Two 5-0 silk stitches are placed, taking the posterior vaginal wall. An incision is created in between both sutures to open the vagina and to see the characteristics of the urogenital sinus. The incision done at the posterior wall of the vagina is extended distally through the common channel to expose the entire anatomy. Depending on the characteristics of the urogenital sinus, mainly the length of the common channel, we will follow the strategies already mentioned during the repair of cloacas. Most of the time, the urogenital sinus is untouched and therefore, the surgical manipulation of the structure is not more difficult than in primary cases.

As previously described, if the common channel is shorter than 3 cm, we will proceed with the

total urogenital mobilization exactly the way we do in a primary case. Most of the times, that maneuver has been enough to repair these malformations. Dealing with adult patients or teenagers, the operation, of course, involves bigger structures, which represents a technical advantage. The vessels are bigger, and therefore, sometimes the needle-tip cautery used in coagulation mode is not enough to maintain good hemostasis and it is necessary to use suture ligatures. The venous plexus, located behind the pubis, must be preserved as much as possible. However, sometimes it is impossible; we hit those veins and, in teenagers or adults, it is necessary to use suture ligatures because the cautery is not enough to obtain good hemostasis. The decision-making algorithm mentioned in the treatment of the urogenital sinus of cloacas in primary procedures is applied in these cases the same as previously described.

Vaginal replacement with rectum, dividing the rectum longitudinally as previously described, is more commonly done in these cases because these patients have been using the rectum for a long time and are frequently suffering from constipation, and therefore, they have a dilated rectum. The fact that they have a clear mesentery, if they have colon in the pelvis, makes the preservation of the blood supply even easier.

Postoperatively, the patients remain 10 days with nothing by mouth, receiving parenteral nutrition. The rectum is reconstructed following the same principles described in the primary repair of a cloaca. If the rectum happens to be mislocated, now is the opportunity to put it in the right place.

#### 16.1.4.2 Acquired Vaginal Atresia or Stricture

Another common reason why patients are referred to us after an attempted failed repair of a cloaca is because they suffer from an acquired vaginal atresia or stricture. In these cases, the surgeon usually knew that he was dealing with a cloaca and actually tried to repair the vaginal component of the malformation. Unfortunately, for different reasons, the vagina was not mobilized properly and suffered from ischemia and acquired atresia or stricture (Fig. 16.64). As a consequence, the patient comes to us with a patent urethra and rectum, but without a vaginal orifice. The vagina may or may not have a communication with the urethra, but the distal end that the surgeon originally attached and sutured to the perineum disappeared. The distance between the blind end of the vagina and the perineum varies from patient to patient and, depending on that length, the magnitude and type of operation changes. In order to repair these complex cases, it is necessary, again, to mobilize the rectum. The mobilization of the rectum, in the way described in the paragraph related to persistent urogenital sinus, is the same in these cases. We have to mobilize the rectum in order to have access to the vagina. The exception could be a case in which the blind vagina is located so high that we estimate with a CT scan and/or MRI study that the blind vagina will be easier to reach through the abdomen rather than from below. Most cases, however, can be reached from below, at least to initiate the dissection. The rectum is dissected and mobilized in the same way as previously described. If the patient already has bowel control, it is our experience that a reoperation done correctly does not change the functional prognosis that the patient had from the fecal continence point of view.

Once the rectum has been mobilized, we find the posterior vaginal wall, open it, identify the blind distal end, and use multiple stitches to initiate our dissection. If the patient never underwent a total urogenital mobilization, we do it as a first step. On the other hand, sometimes the previous operation done by another surgeon included an attempt to separate the vagina from the urethra and they left the urethra intact. If that is the case, we can try to mobilize the vagina again. The decision-making algorithm described in the primary repair of a cloaca is applied here. In other words, if the total urogenital mobilization is not enough to make the vagina reach the perineum, we have to go into the abdomen to continue with the extended transabdominal urogenital mobilization. If that is not enough, we have to continue with the separation of the vagina from the urinary tract, and if that is not enough, we have to evaluate

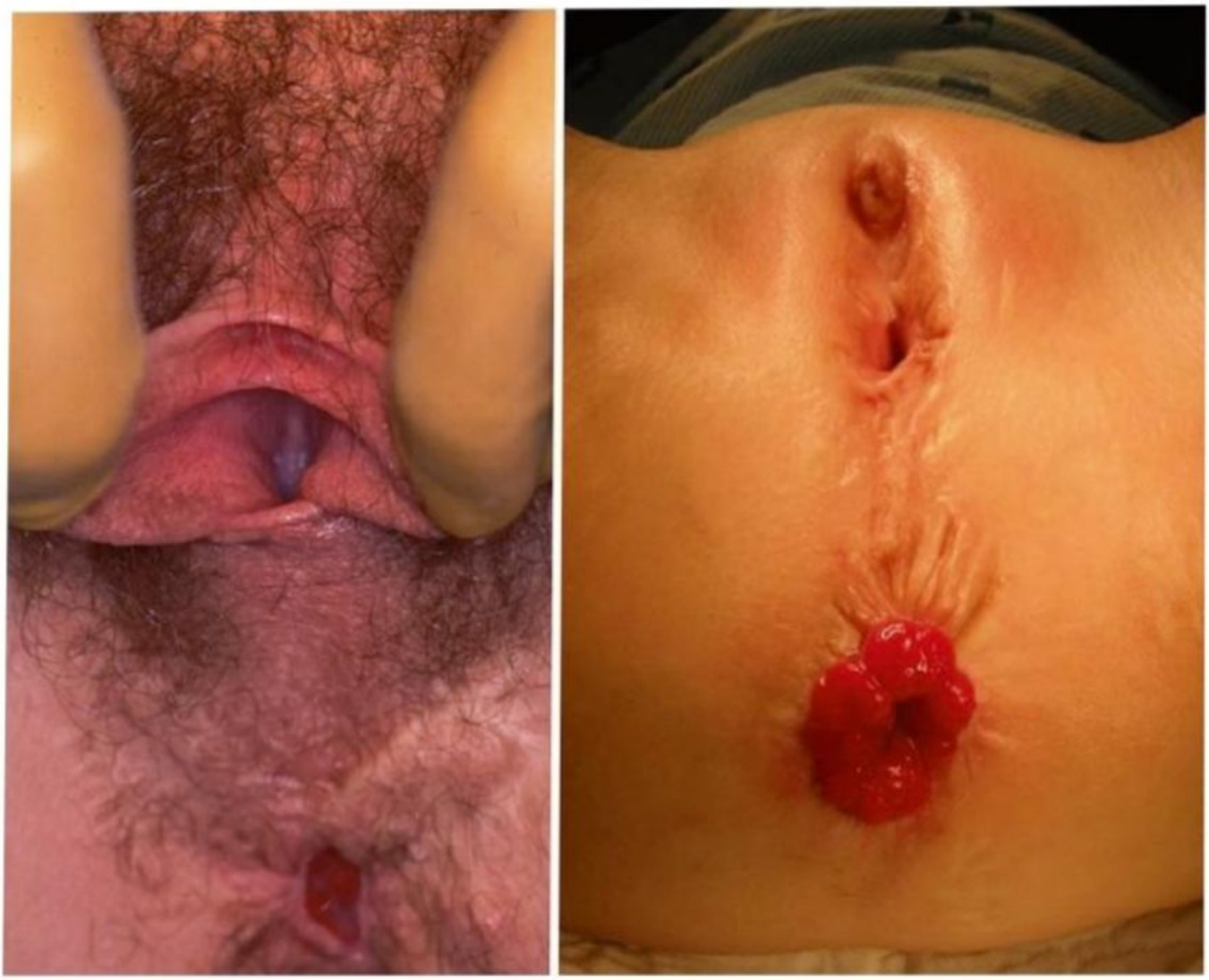

Fig. 16.64 Picture of the perineum of several patients who underwent a previous failed attempted repair of a cloaca

the possibility of a vaginal switch and eventually a vaginal replacement.

#### 16.1.4.3 Acquired Urethral Atresia or Stricture

Very occasionally, one may find a patient who comes after a failed attempted repair of cloaca; the patient developed an acquired atresia or severe stricture of the urethra. Depending on the location of this, we may follow a different strategy. Certainly, one alternative to repair this is the transpubic approach that will be described in another chapter. If the urethral stricture is located low enough, we may try to repair that posterior sagittally. First, we have to go through the mobilization of the rectum, as previously described, then the vagina, and eventually the urethra. Sometimes, the acquired atresia of the urethra is so severe and the length of separation of both healthy portions of the urethra is so great that the decision is made not to repair the urethra but actually to perform a Mitrofanoff type of procedure, creating a conduit for the patient to empty her bladder with intermittent catheterization through an orifice created in the abdomen.

#### 16.1.4.4 Sequelae from Catastrophic Complications

Some patients underwent an attempted repair and almost everything went wrong; the patients had dehiscence, infection, retraction, and fistula formation. The operative reports are frequently confusing and only illustrate the disorientation suffering and anxiety of the surgeon exploring the pelvis of a patient with an anatomy that seems to be completely unknown to him or her. As a consequence, we do not know exactly what happened, but we may see, for instance, that the

patient has a blind perineum. In other words, there is no urethra, no vagina, and no rectum. Or, they may have one or two orifices. A study with contrast material injected through the different orifices of the patient (colostomy, cystostomy, vaginostomy, vesicostomy, or rectum) helps us to make the anatomic diagnosis. More recently, at our institution, the Interventional Radiology Department has helped, performing a study called three-dimensional, rotational scan, injecting contrast material simultaneously, through the stomas of orifices of the patient's perineum. Videos 16.1, 16.2 and 16.3 illustrates the beautiful images provided by this kind of study. We often use this modality in primary complex cloaca cases as well. These help us to understand the sequelae in these patients. It is not unusual for us to find that the patient has what we call a "frozen" or "cement type of pelvis." These patients are extremely difficult to approach and based on the diagnostic tests, already described, the surgeon has to make a decision as to how to approach this kind of patient. The first possibility would be to repair these posterior sagittally. However, sometimes posterior sagittally, the operative findings are very frustrating, and it is very difficult to mobilize the structures. The next possibility is an abdominal midline incision. Yet, through the abdomen, sometimes it is also very difficult to find the plane of dissection in the pelvis. At that point, the last alternative and more aggressive approach that provides the best exposure is the transpubic route.

### 16.1.5 Transpubic Approach

The transpubic approach is part of our armamentarium to approach complex pelvic problems [120]. If the surgeon considers that the lesion, fistula, mass, or structure that he or she wants to reach is surrounded by excessive fibrosis and is located in a place impossible to reach from below or from above, then another alternative is to consider the transpubic approach. This type of approach provides excellent exposure. It is particularly indicated in a patient who has a functional rectum but a significant number of problems in the urogenital tract, in other words, urethrovaginal and bladder vaginal fistula (Fig. 16.65a–c).

A midline incision running from the umbilicus down toward the genitalia is performed, all the way down to the urethra. The clitoris is divided exactly in the midline with needle-tip cautery. The pubis is exposed and dissected, in order to identify what is bone and what is cartilage, to be sure that the cartilage is divided exactly in the midline (Fig. 16.65). The needle-tip cautery divides the pubic cartilage very easily. We must keep in mind that behind the pubis are venous plexuses that may bleed significantly, particularly in teenage or adult patients. In those cases, we should use sutures to stop the bleeding. Once we divide the pubic cartilage, the pubic bones spontaneously separate, giving us usually enough space to work. If that is not the case, we can use a retractor to separate the pubic bones more. With this approach, we achieve a great exposure to the urogenital tract (Fig. 16.65b); we can work comfortably to deal with the specific complications that the patient has. We have operated on 20 patients with this transpubic approach.

At the end of the procedure, the pubic cartilage is reapproximated using 0, 00, or 1 Vicryl sutures. We keep the patient in bed for a week before we allow her to walk making sure to avoid external rotation of the hips. The main inconvenience of the transpubic approach is the fact that it produces significant pain postoperatively. The patients usually refuse to walk for a week or 2 because of the pain. One patient suffered from an acute dehiscence of the pubic cartilage that had to be resutured on an emergency basis. Surprisingly, this occurred in a little baby and not in an ambulatory patient. Another patient suffered from high fever; a bone scan suggested that the patient was suffering from an infection in that area. The patient received antibiotics for 6 weeks to treat osteomyelitis and healed normally. Other than that, all the patients have done very well. We do not recommend the transpubic approach except under the circumstances already mentioned.

The functional results in reoperations for cloacas vary from case to case and depend very much on the type of complication and the origi-

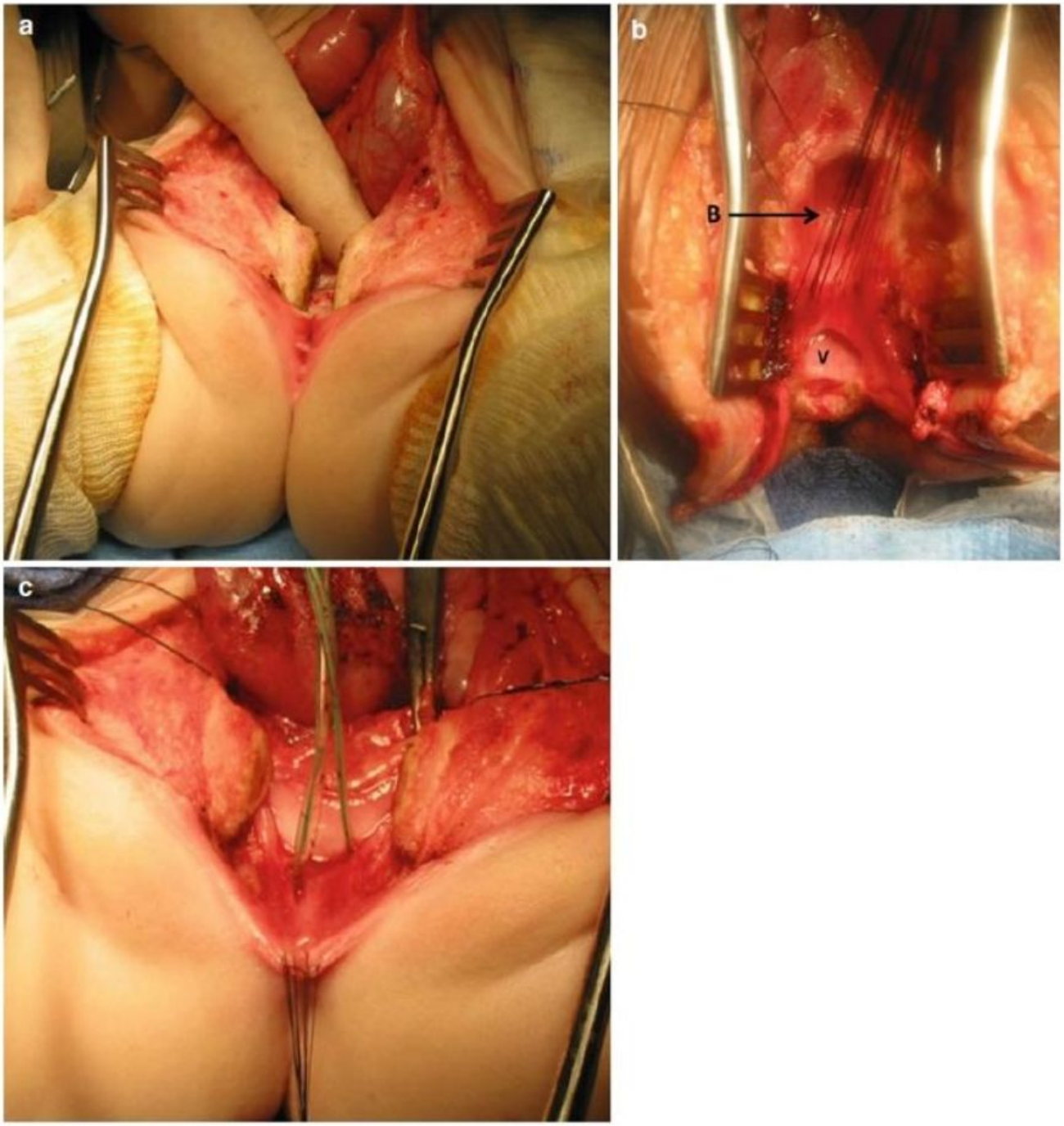


**Fig. 16.65** Transpubic approach. (**a**) Dividing the pubis. (**b**) Absent bladder neck. Wide communication between vagina and bladder. *B* bladder, *V* vagina. (**c**) Ectopic ureters opening below the bladder neck

nal complexity of the malformation. In a patient with a normal sacrum, common channel shorter than 3 cm, and an intact persistent urogenital sinus, we can expect an excellent result, including normal urinary control. Of course, when the previous surgeon was too invasive, performed a lot of dissection, damaged important nerves and structures, and produced more fibrosis, the functional prognosis cannot be predicted. This has to be informed to the parents.

As part of the long-term follow-up, we explain to the parents of our patients that they must pay special attention when their daughter shows signs of puberty, has no menstruation, and complains of abdominal pain with monthly exacerbations; when these are observed, they must contact us. In such case, we order a pelvic ultrasound and an MRI to rule out the presence of menstrual blood in the peritoneal cavity. To alleviate the symptoms, sometimes it is beneficial to administer

medication to suppress the menstruation, which gives us time to plan the surgical treatment.

The next time to be concern is when the patient is expected to have sexual intercourse. We encourage them to have an examination under anesthesia. We want to be sure that the vagina has the adequate characteristics (size and elasticity) to avoid unpleasant experiences.

Sometimes, we find a narrow external orifice with a large, elastic vagina. That problem is solved with an external vaginoplasty (see Fig. 16.38). Other times, we find a more serious problem, a patent vagina, but very inelastic due to scarring. For that problem, we designed an operation consisting in patching the posterior wall of the vagina with the most distal piece of rectum. Figure 16.66a–g shows the surgical technique that we use to patch the vagina with rectum.

This procedure is only recommended when the patient has no bowel control. We must keep in mind that the resection of the distal rectum in a patient with bowel control may provoke fecal incontinence in a patient that had bowel control. If the patient is fecally incontinent, the removal of the most distal part of the rectum has no functional repercussions. We have done this operation in 11 of our patients.

## 16.2 Posterior Cloaca and Absent Penis Spectrum

"The case report of M. Louis from Paris, has become famous. In his thesis in 1753, he described a girl with an anus cloacal who menstruated per anus. Happily married to an impetuous young man, she finally confided her secret to him. At the height of passion, the latter persuaded his wife to engage in coitus. She agreed to it and became pregnant. The birth of the infant occurred at term and caused a tear of the anal sphincter."

"The presentation of this thesis before the surgeons, led to legal prosecution of M. Louis in Paris. During the legal proceedings, it was decided that moral theologians should resolve the issue of whether the conduct of this woman and that of M. Louis was illegal. At last, the Pope himself was called upon to decide. He, however, more farsighted than the parliament of Paris and the physicians of the Sorbonne, granted M. Louis absolution, and the thesis was finally published in 1754."

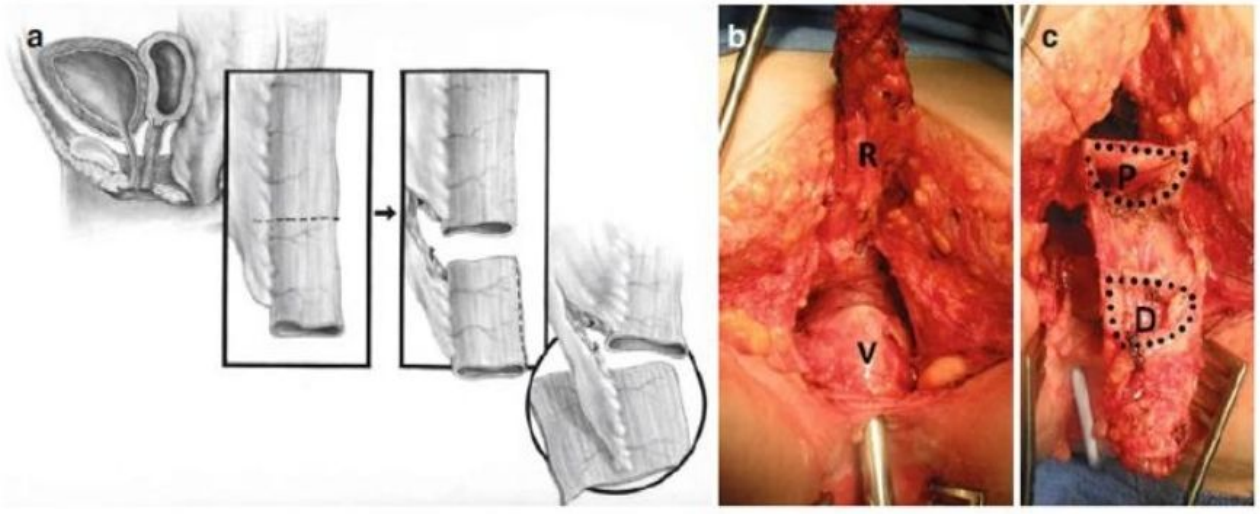


**Fig. 16.66** Surgical technique to patch a narrow vagina with rectum. (**a**) Diagram showing the creation of a rectal patch, preserving the blood supply. (**b**) Operative view. The rectum has been mobilized and is pulled up. A Hegar dilator is introduced in the vagina. *R* rectum, *V* vagina. (**c**) Operative view. The rectum has been divided, preserving the blood supply of the distal segment. *D* distal, *P* proximal. (**d**) The distal segment is open to tailor the patch. *P* patch. (**e**) The rectal patch is being sutured to the narrow vagina. The cervix is clearly seen in the vagina. *V* vagina, *P* patch. (**f**) Diagram showing the finished operation. (**g**) Diagram showing a technical modification to increase also the length of the vagina

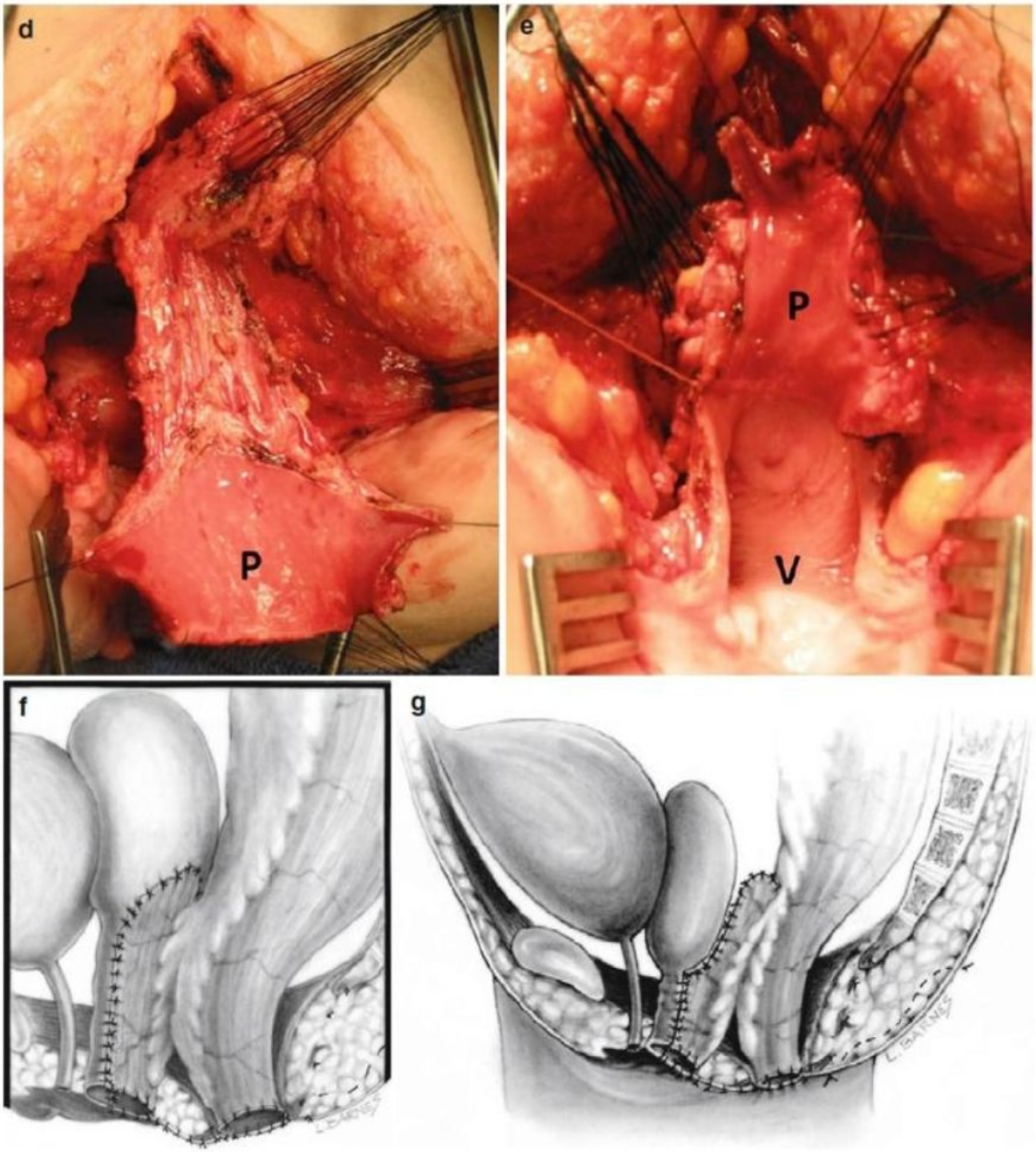


**Fig. 16.66** (continued)

These two paragraphs were taken from Scharli [121], from a publication entitled "Malformations of the Anus and Rectum and Their Treatment in Medical History," published in *Progress in Pediatric Surgery*, volume 11, 1978. Dr. Scharli took this information from a publication of M. Louis himself [122] who was also quoted by Bushe [123].

Two-hundred and thirty-two years later, in 1986, the senior author of this book was invited by Dr. Fred Leditschke to Brisbane, Australia, to operate on a little girl who was born with "an unusual anorectal and urinary malformation" [124]. That particular girl had a very unusual history. She was born and considered normal; therefore, she was discharged and went home. The mother noticed that she was passing many, very frequent, liquid bowel movements and went through a series of consultations with different

pediatric specialties who could not find the reason why the girl suffered from this "chronic diarrhea." Subsequently, the patient was referred to Dr. Fred Leditschke (pediatric surgeon) who performed a more thorough examination and also an endoscopy; by doing that, he was able to find that the patient had no vaginal opening and no urethral opening (Figs. 16.67 and 16.68). She was passing stool and urine through a single orifice located in the same location of a normal anus! In addition, the endoscopy disclosed the presence of an orifice in the anterior rectal wall, and introducing the scope through that orifice, he was able to find a urethra and vagina; in other words, it was a urogenital sinus, posteriorly deviated and connected to the anterior rectal wall (Fig. 16.69). Interestingly enough, the little girl went through 18 months of her life suffering from this "pseudodiarrhea" and nobody had seen directly the external genitalia. This is very important because

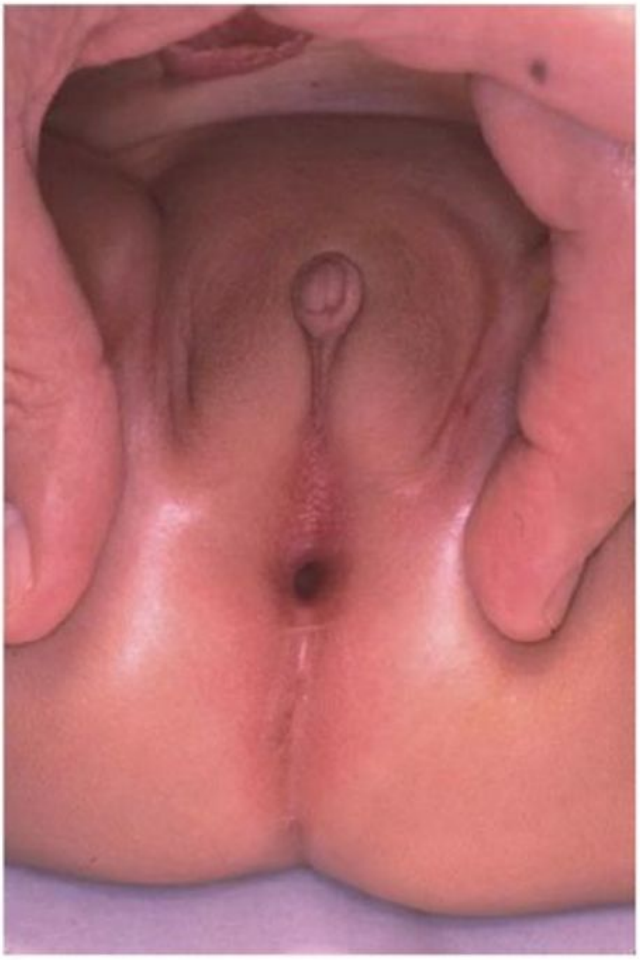

Fig. 16.68 External appearance of a case with a posterior cloaca, separating the labia majora

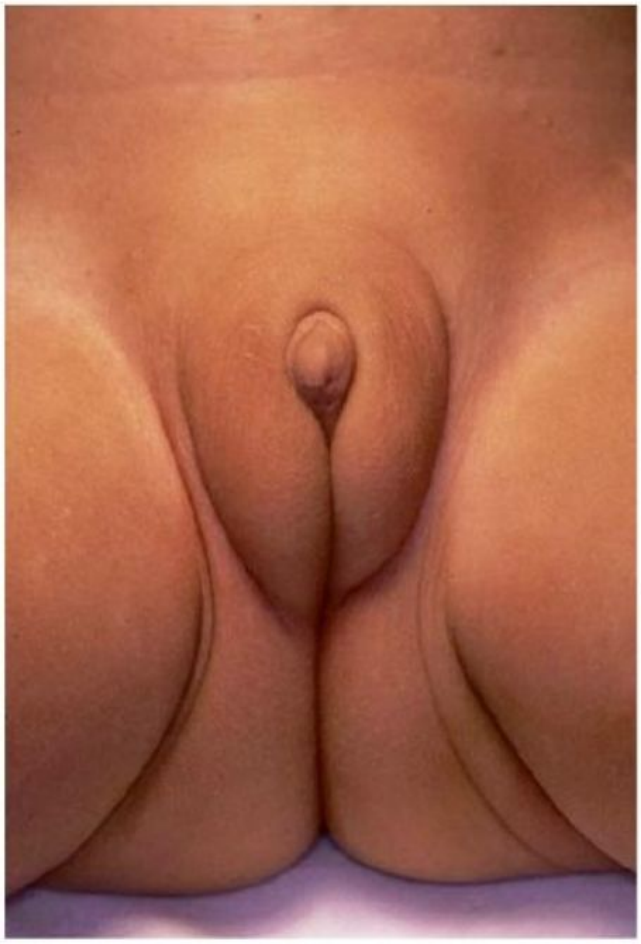

Fig. 16.67 External genitalia of a patient with a posterior cloaca

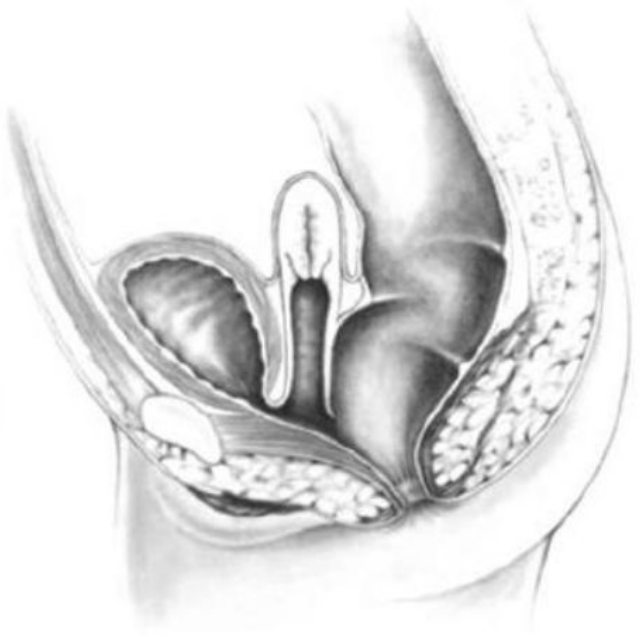

Fig. 16.69 Diagram of a posterior cloaca

many of these patients, when examined externally, they look normal; it takes a special interest to separate the labia of the genitalia in order to see the anomaly (Figs. 16.67 and 16.68).

The senior author, together with Dr. Leditschke, repaired that very unusual malformation successfully. We are happy to say that that little girl is now a beautiful young lady, happily married, and also delivered a baby by cesarean section.

Based on the external and internal findings of this particular malformation, we decided to call this "posterior cloaca." These patients have a single perineal orifice, but what makes this defect unique is the fact that the single orifice is located in the same location as a normal anus. A typical cloaca has a single orifice located in the same location of a normal urethra. As a consequence of the repair of this malformation, we suspected that perhaps there would be other similar cases; in retrospect, we found that there were more cases than what we originally thought, except that they had not been named and had been included in the category of cloaca.

As a consequence of the repair of that initial defect, two ideas came into our minds. Number one is the idea of dividing the entire rectum, what we now call "trans-anorectal approach." In other words, the fact that the patient has a normal anus with normal pectinate line and therefore normal anal canal makes that patient fecally continent by definition. The surgical technical implication of this is that we should not mobilize the anus and the rectum. We can divide the entire rectum in the midline, including both posterior and anterior walls, repair the urogenital sinus anteriorly with an excellent exposure, and reconstruct the rectum. We had evidence that this does not harm fecal control (see Chap. 26). The second idea that came to our minds as a consequence of dealing with this malformation is the "total urogenital mobilization." In order to move the urogenital sinus from its posterior mislocation, seen in posterior cloacas, it is necessary to mobilize it and switch it forward to be able to place the urethra in a normal location and the vagina behind the urethra. That maneuver has demonstrated to be extremely useful in the repair of cloacas with a common channel shorter than 3 cm and also in urogenital sinus cases with normal rectum (see Sect. 16.1 and Chap. 26).

Very soon, in the management of these cases, we realized that there was another very conspicuous anomaly present in these cases, and that was the fact that the pubis is extremely thick. In other words, the urogenital sinus is posteriorly deviated, and we do not know if that is because of the presence of an extremely prominent pubis or is just coincidental. The fact is that it is necessary to carve the pubis, removing part of it, to be able to create the necessary space to move the urogenital sinus forward and create urethral and vaginal orifices in a normal location.

As we gained experience in the management in these cases, we found another variant of these malformations. We were able to see girls that are born with the anus normally located or slightly anteriorly mislocated and a second orifice (urogenital sinus) located immediately anterior to the anus (Fig. 16.70). The fact that they have two perineal orifices would prevent us to call that a "cloaca"; yet, the urogenital sinus is posteriorly deviated and mislocated; therefore, we believe that we should consider that malformation as a part of the spectrum of posterior cloaca [125]. In addition, we found cases that had a posterior cloaca, but in addition, they have an accessory micro-urethra that runs from the bladder toward the tip of a pseudophallus or clitoris (Fig. 16.71). We also include that in the spectrum of posterior cloaca because of the common denominator that is a normally located anus or slightly anteriorly mislocated with a posterior location of the urogenital sinus.

We could not find in the literature a report using the term posterior cloaca. However, looking at the very few publications on the treatment of cloaca, prior to 1982, we found that sometimes the authors of those papers referred to cloaca patients using the term "urogenital sinus and imperforate anus." Looking at the specific pictures that the authors presented in the publications, we were able to see that some of those cases were actually posterior cloacas, but that term was not mentioned and the specific posterior deviation of the urogenital sinus was not described. We found one specific paper in 1981

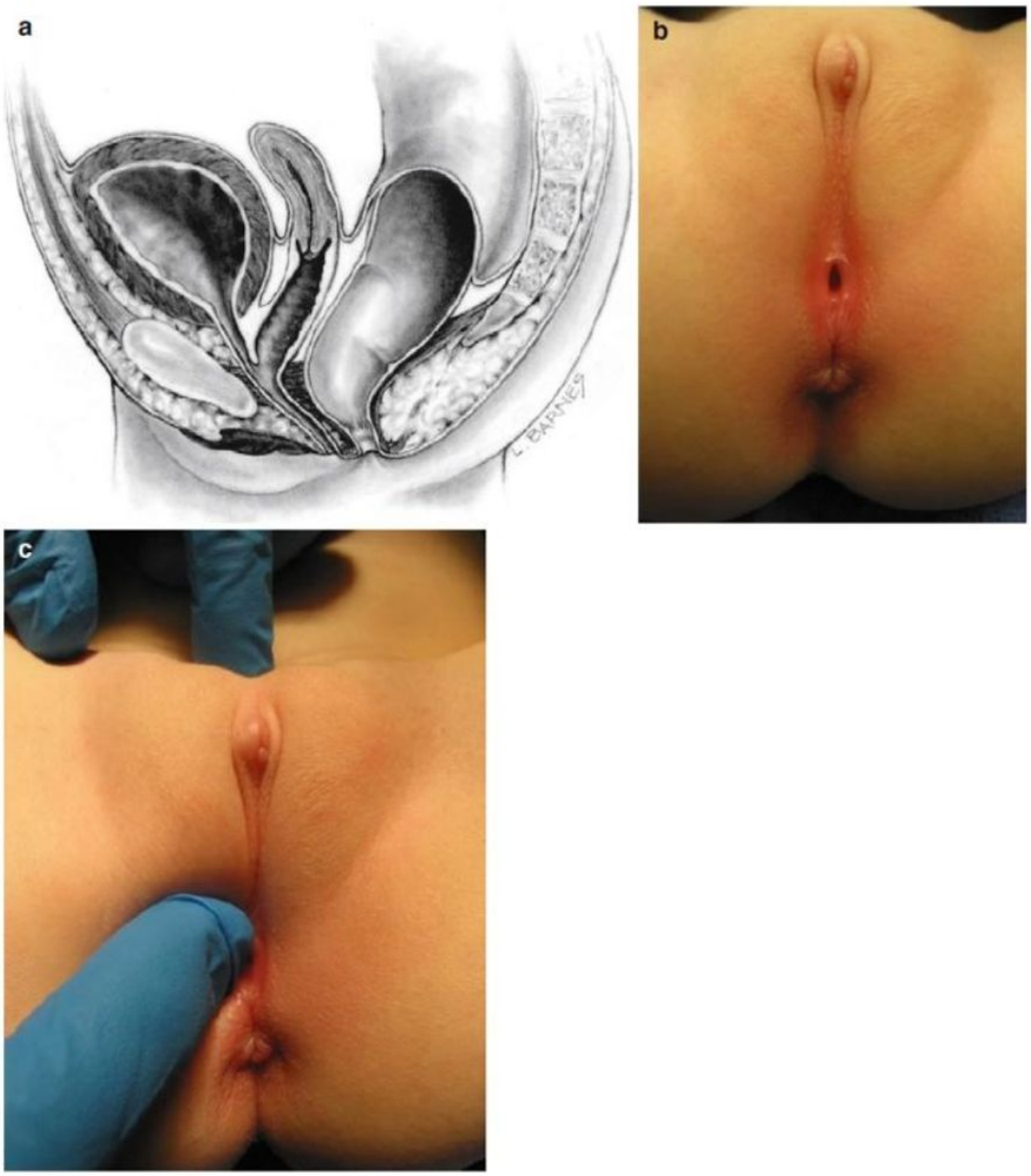


**Fig. 16.70** Posterior cloaca variant. The urogenital sinus is located immediately anterior to the anus, but far away from the clitoris. (a) Diagram of sagittal aspect, (b) external appearance of the perineum, (c) demonstration of large pubis

by John Duckett and Ballinger. They entitled their paper "Accessory Phallic Urethra in the Female Patient," and actually they described three cases that are posterior cloacas according to the pictures that we saw in the paper [126]. In 1994, Chatterjee published a case described as clitoromegaly, duplex urethra, and dysplastic vagina, but the photograph of the patient shows the 2-orifice variant of the posterior cloaca spectrum [127].

More recently, the term posterior cloaca seemed to be becoming more popular and we were able to

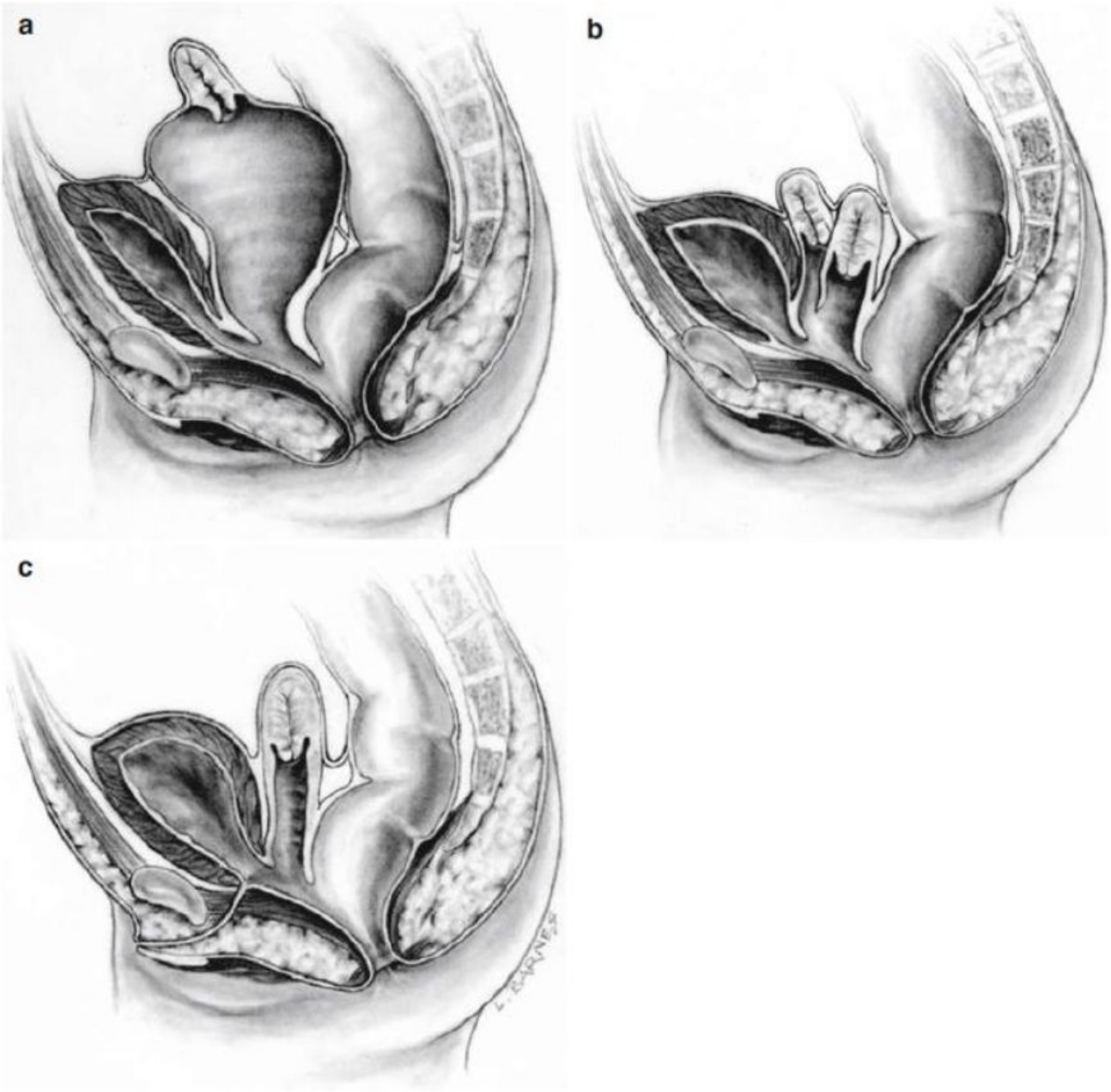


**Fig. 16.71** Diagrams showing a sagittal view of three different variants of posterior cloaca. (a) With hydrocolpos, (b) with double vagina and (c) with an accessory quasi-atretic urethra opening at the tip of a pseudophallus (clitoris)

find publications referring to this specific defect using the term posterior cloaca [128–131].

## 16.2.1 Surgical Repair

Most of these patients came to us with a colostomy performed at another institution during the newborn period. Patients born with a posterior cloaca also may have hydrocolpos with all of the negative consequences related to this defect such as megaureters and hydronephrosis. That should be treated at birth.

### 16.2.1.1 Typical Cloaca with a Single Perineal Orifice in the Anal Location

The patient is placed in the prone position with the pelvis elevated. A posterior sagittal transanorectal incision is performed. As described in Chap. 26, the incision includes the skin, subcutaneous tissue, parasagittal fibers, muscle com-

plex, posterior rectal wall, anterior rectal wall, and anterior sphincter mechanism. With the use of self-retractors, we have beautiful exposure (Fig. 16.72). The urogenital sinus that is connected to the anterior rectal wall, usually about 1 or 2 cm from the anal verge, can be seen as soon as we open the posterior rectal wall. Multiple 5-0 silk sutures are placed in the circumference of the single orifice of the urogenital sinus, in order to be able to apply uniform traction to facilitate its dissection. The urogenital sinus is separated from the anterior rectal wall. The perineum anterior to the anus is also divided in the midline all the way up to the clitoris. At this point, we can evaluate and feel if the pubis is interfering and does not allow us to find space to place the urethra immediately behind the clitoris and the vagina behind. If that is the case, in babies, we can resect more than 50 % of that very heavy thick cartilage of pubis with needle-tip cautery, with minimal bleeding. By doing that, we create enough space to switch forward and mobilize the urethra and vagina together (total urogenital mobilization). As previously described in the Sect. 16.1, if the common channel is shorter than 3 cm, the overwhelming majority of cases can be repaired without opening the abdomen, placing the urethra immediately behind the clitoris, with the vagina behind, without separating the vagina from the urethra. Figure 16.72 shows pictures and diagrams of this procedure.

The urethral opening is sutured with 6-0 long-term absorbable sutures to the tissue immediately behind the clitoris as described in the repair of cloacas. The vaginal walls are sutured with interrupted 5-0 long-term absorbable sutures, to the skin of what is going to be the neolabia. The divided rectum is already perfectly visible (Fig. 16.72). A meticulous reconstruction is performed, bringing together corresponding portions of the sphincter mechanism anterior to the anus. The anterior rectal wall and the anal wall are sutured with two layers of interrupted long-term absorbable sutures. The posterior rectal wall is reconstructed in the same manner followed by a meticulous reconstruction of a normal sphincter mechanism located behind the rectum and anus.

### 16.2.2 Surgical Repair of the 2-Perineal-Orifice Variant of the Posterior Cloacal Spectrum

When patients are born with two orifices, one is the anal opening located in the normal location or slightly anteriorly and the second one located anterior to the anus, but far away from the clitoris, it represents the urogenital sinus (Fig. 16.70a, b). In that type of malformation, provided the common channel is not too long, it is conceivable, as many authors like to say, that we could repair the urogenital sinus without necessarily dividing the anorectum. Of course, we agree that if that is feasible, it should be done that way, but also we believe that we should not compromise the quality of the repair trying to avoid the opening of the healthy rectal walls (See Chap. 26, Fig. 26.13a, b). The urogenital sinus is mobilized as described in the case of cloacas. When the pubic bone and cartilage represent an obstacle, it should be resected, part of it, to create the necessary space, and the reconstruction is done following the same steps already described.

Our experience with the repair of posterior cloaca and its variants includes 30 patients [132]. The results in terms of urinary function, bowel control, and sexual concerns are very similar to those found in the repair of cloacas and depend, like in cloacas, on the length of the common channel that influences mainly the urinary function and the presence, absence, or abnormalities of the sacrum.

### 16.2.3 Posterior Cloaca and Absent Penis

The reader must be surprised that we are discussing here a malformation called absent penis in a chapter dedicated to female patients with posterior cloacal malformations. The reason for this is that we found interesting striking similarities between both malformations: posterior cloaca in females and absent penis in males.

Absent penis through history is a very well-known malformation. We believe that the defect

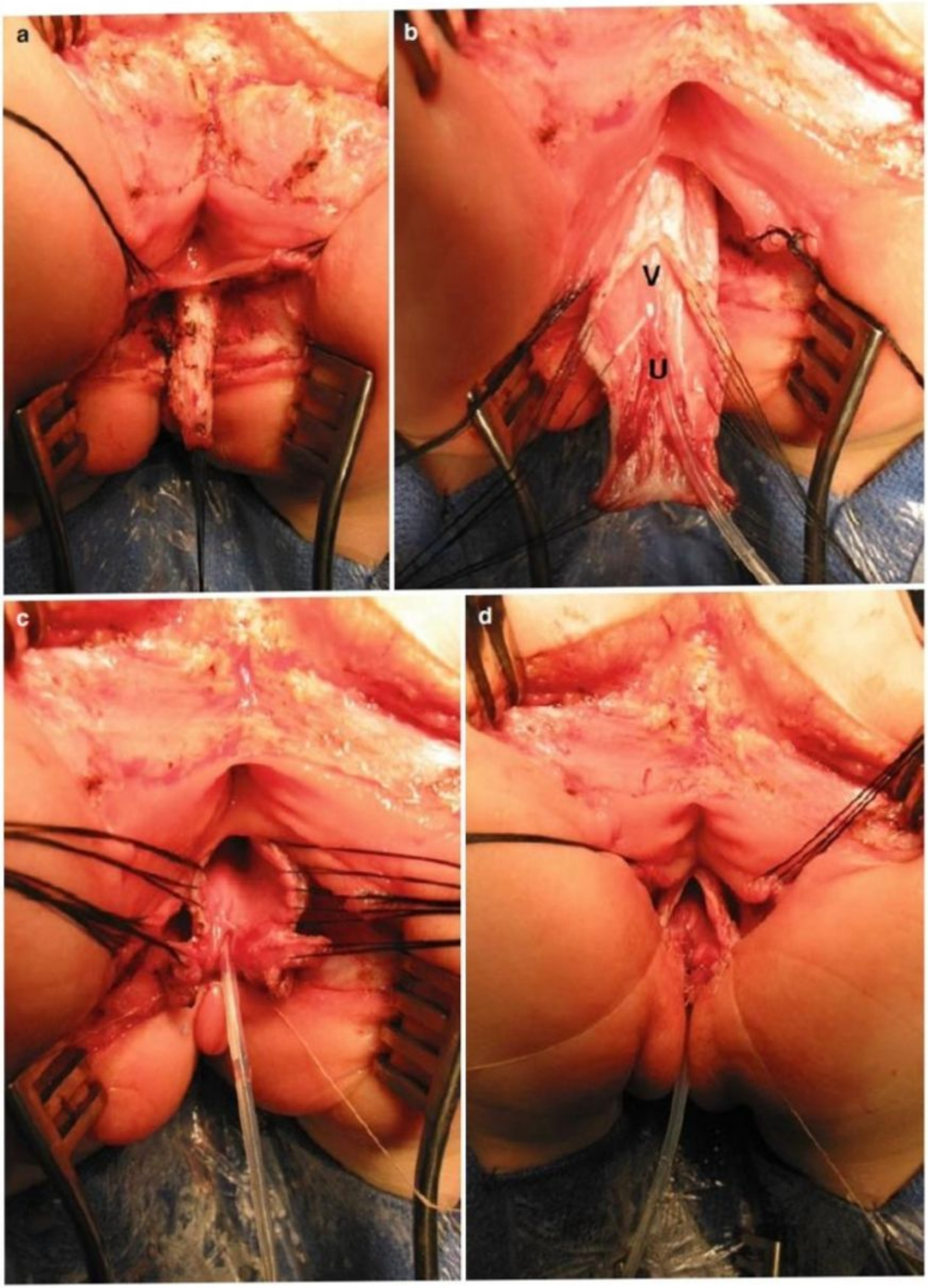


**Fig. 16.72** Operative pictures of the posterior approach of a posterior cloaca. (**a**) The rectum has been divided in the midline, and the urogenital sinus is already mobilized. (**b**) The posterior wall of the urogenital sinus has been opened. "*V*" is vagina and "*U*" is urethra. (**c**) The urethral opening is sutured a few millimeters behind the clitoris. (**d**) The lateral walls of the vagina are sutured to the neolabia

has been recognized for many years, due to the fact that it is a very obvious, conspicuous defect. It is not the purpose of this presentation to review the historical information related with this defect; we found references even from the last two centuries.

We included in this review a few references that we consider important related with this defect [133–138]. These babies are chromosomally males, have testicles, and are born without a phallus. The posterior urethra is posteriorly deviated and opens into the anterior rectal wall or immediately anterior to the anus (Fig. 16.73). Looking carefully to this diagram, one can see the remarkable similarity with the anatomy of a posterior cloaca in female patients.

Historically, and occasionally even now, these babies are raised as females. In other words, they are subjected to bilateral orchiectomies, the urethra is mobilized to be placed similar to a female patient, and a neovagina is created [139–150].

There is another very interesting malformation, seen in male patients, that is known with different names such as "urethral atresia with urethrorectal communication," "duplication of the urethra," "dystopic urethral orifice into the anus," and "Y-duplication of the male urethra." All these names refer to a malformation very similar to the absent penis, except that the penis is present. In other words, the posterior urethra is posteriorly deviated and opens into the anterior rectal wall or immediately anterior to it; the patient has a very hypotrophic, usually useless, tiny penile urethra that opens in the tip of the penis, and that is the reason why a lot of people call this "double urethra" or "Y urethra" (Fig. 16.74). Again, one can see the striking similarity between this defect and absent penis and also with the posterior cloaca in females [151–154].

We have no experience with the treatment of absent penis. However, we have experience in the treatment of the variation that we just described, namely, the posterior urethra deviated to the anterior rectal wall and a quasi-atretic penile urethra (Fig. 16.74). This malformation is repaired by stages. The first procedure is done with the patient in prone position, mobilizing the urethra from the anterior rectal wall or from the very anterior perineum, forward, to put it in a more convenient perineal location away from the anus (Fig. 16.75a). This operation can be done dividing the anterior rectal wall and the posterior rectal wall or in more simple cases without touching the rectum. After the operation, basically, the

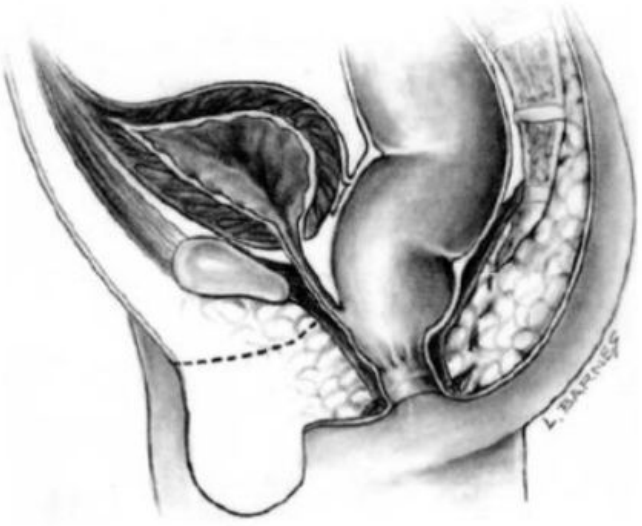

**Fig. 16.73** Diagram showing a sagittal view of an absent penis. The posterior urethra is connected to the anterior rectal wall

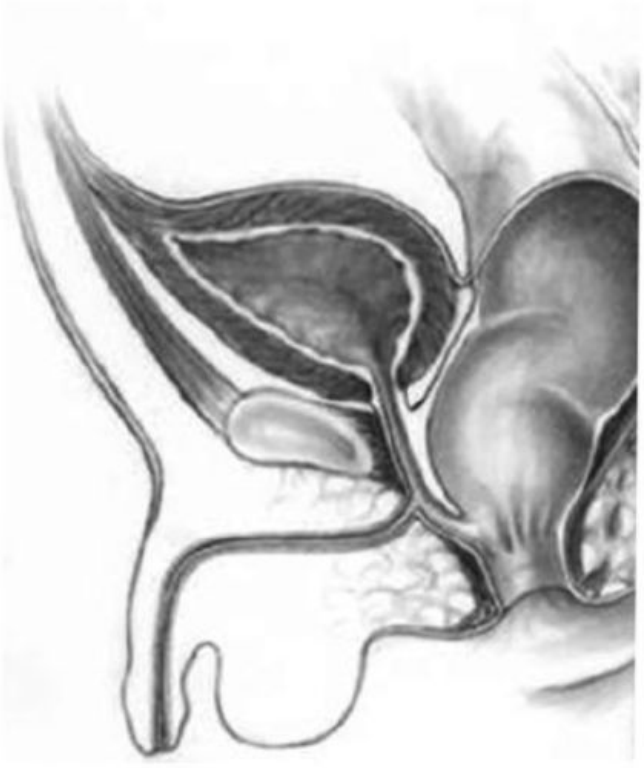

**Fig. 16.74** Diagram showing a sagittal view of a malformation considered a variant of the absent penis. The posterior urethra is connected to the anterior rectal wall and the penile urethra is very structured

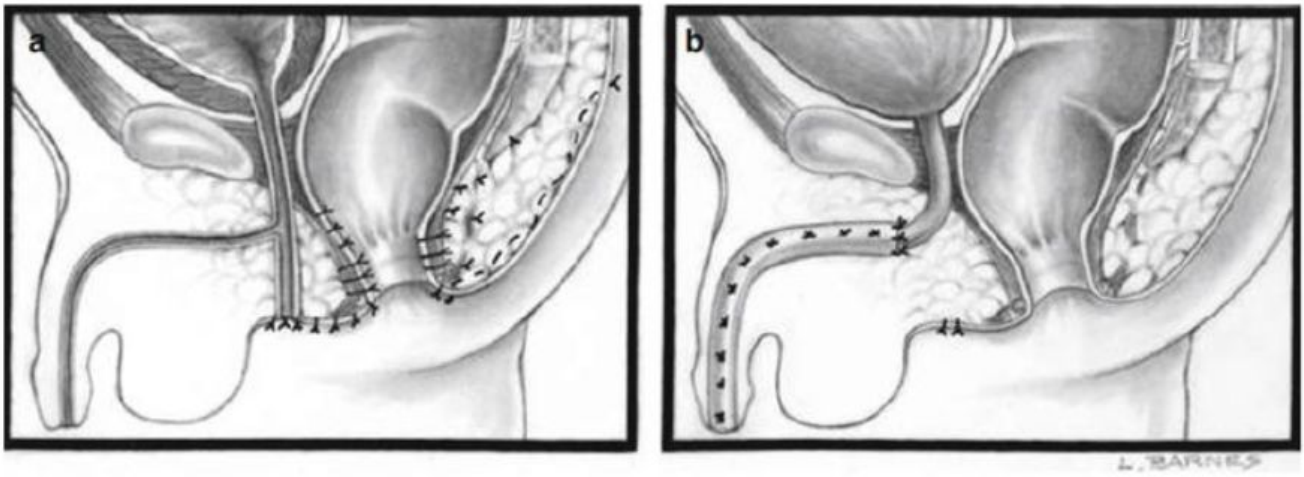


**Fig. 16.75** Diagrams showing the repair of the "absent penis variant." (**a**) The posterior urethra has been disconnected from the anterior rectal wall and switched forward like a perineal urethrostomy, (**b**) another stage of the repair, the penile urethra has been reconstructed with a patch (neighbor tissues or buccal mucosa)

patient would have the equivalent to a severe perineal hypospadias. The penile urethra is usually very narrow. Sometimes we see urine coming out through the tip of the penis, but certainly most of the urine comes from a wide posterior urethra originally connected to the anterior rectal wall or to the perineum. In the subsequent stages, the penile urethra is reconstructed; the posterior urethra connected to the perineum is switched forward and is connected to the reconstructed penile urethra (Fig. 16.75b). This part of the procedure (urethral reconstruction) must be done by skilled, dedicated pediatric urologists.

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