Recto-bladder Neck Fistula

11

11.1 Definition and Frequency

Imperforate anus with a recto-bladder neck fistula is the highest of all anorectal malformations seen in male patients. The rectum connects to the bladder neck (Fig. 11.1). It is relatively common to see that these patients have a rather narrow pelvic space. We interpret this like a manifestation of a significant degree of caudal regression (Animation 11.1). The sacrum may be normal, but frequently, it is very abnormal or even absent. The frequency of associated defects is much higher than in all the other malformations. Fortunately, in our experience, this defect only occurs in approximately 10 % of all anorectal malformation patients in males [1]. Unfortunately, this defect runs with the worst functional prognosis for bowel control and occasionally for urinary control. In our experience, this particular defect is the only one that requires a laparotomy or laparoscopy in order to be repaired. In other words, the rectum is located so high in the pelvis and connected to the bladder neck (very high in the urinary tract) that it is not possible to be reached posterior sagittally.

In fact, some of the worst unfortunate catastrophes that we have seen occurred in babies that were born with this defect and a surgeon tried to reach the rectum posterior sagittally; he obviously could not find it, but in the process, he damaged the vas deferens, seminal vesicles, and/or prostate. In some cases, the surgeons divided the entire urethra or the bladder neck and pulled down a megaureter or even the entire bladder thinking that they were dealing with the rectum. These catastrophic events occurred only in patients that were operated on without a preoperative high-pressure distal colostogram (Animation 11.2).

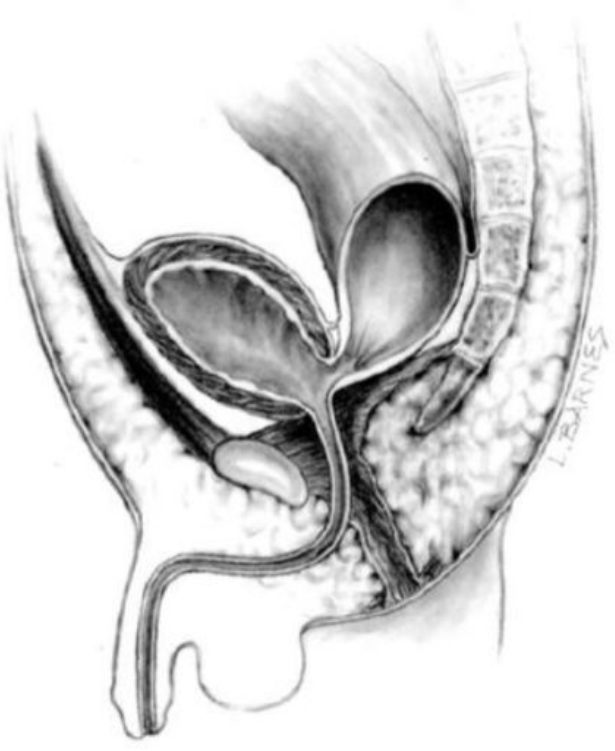

Fig. 11.1 Diagram showing a bladder neck fistula

Electronic supplementary material Supplementary material is available in the online version of this chapter at 10.1007/978-3-319-14989-9_11.

A. Peña, A. Bischoff, *Surgical Treatment of Colorectal Problems in Children*,
DOI 10.1007/978-3-319-14989-9_11, © Springer International Publishing Switzerland 2015

11.2 Associated Defects

11.2.1 Sacral Defects

The average sacral ratio in patients with recto-bladder neck fistula is AP 0.51 and lateral 0.6 (normal AP is 0.74 and lateral 0.76). It is not uncommon to find these patients to have, in addition, sacral hemivertebrae in approximately 35 % of the cases.

11.2.2 Spinal-Associated Defects

Fifteen percent of these patients have suffered from hemivertebra, which produces scoliosis of different degrees in magnitude. The most frequent location of the hemivertebra is the lumbar spine, followed in frequency by the thoracic spine.

11.2.3 Urologic-Associated Defects

As expected, the incidence of associated urologic defects is the highest in these particular types of patients. In a recent evaluation of 110 patients operated on by us with recto-bladder neck fistula, 99 (89 %) of them suffered from some sort of urologic malformation or vesicoureteral reflux. The frequency of absent kidney in these patients is 37/111 (33 %). Vesicoureteral reflux was present in 40 patients (36 %). Eighteen percent of them suffer from hypospadias. Thirty patients (26 %) suffered from an undescended testicle. Eighteen percent of them have a bifid scrotum. Two patients have penile-scrotal transposition. Thirty-three patients (29 %) were born with hydronephrosis. In addition, there is a specific group of patients born with recto-bladder neck fistula that have a single kidney with hydronephrosis, megaureter, and massive reflux. This is a very bad situation because usually they have poor sacrum and therefore poor prognosis for bowel and urinary control. In addition, the fact that the patients are born with a single kidney with hydronephrosis indicates that they already have a significant degree of kidney damage and there is a high chance that these patients will end up with a kidney transplant when they grow up.

Table 11.1 List of urologic abnormalities in patients with bladder neck fistula

Anomaly	No. (%)
VUR	36
Absent kidney	33
Hydronephrosis	29
Undescended testis	26
Bifid scrotum	18
Hypospadias	18
Intravesical verumontanum	45
Urethral stenosis	13
Neurogenic bladder (congenital)	11
Megaureter	9
Other	24

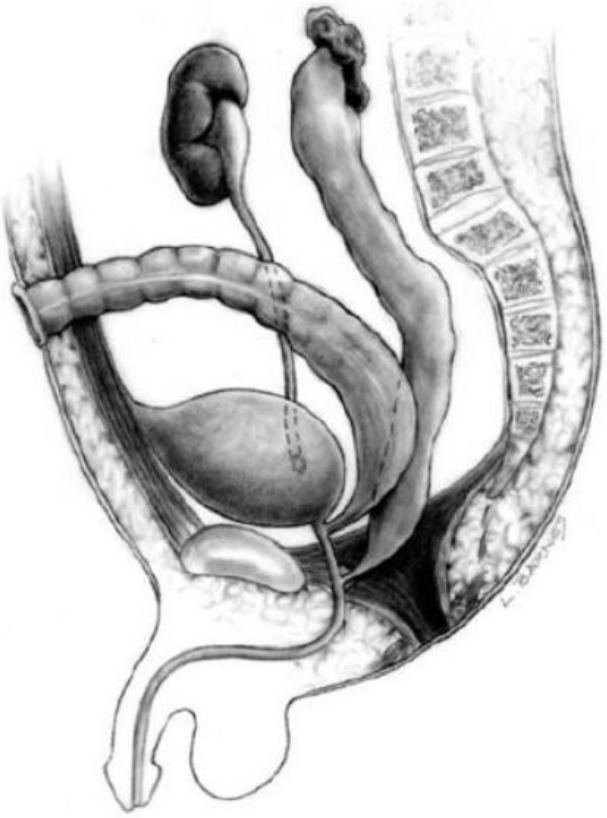

Fig. 11.2 Diagram showing a case of bladder neck fistulas with an ectopic ureter

Table 11.1 shows the list of urologic abnormalities.

Ectopic ureters were present in ten cases (9 %). The ectopia occurs most commonly toward the bladder neck and occasionally in the posterior urethra. This last group of ureters connected to the posterior urethra usually originates from completely damaged kidneys (Fig. 11.2).

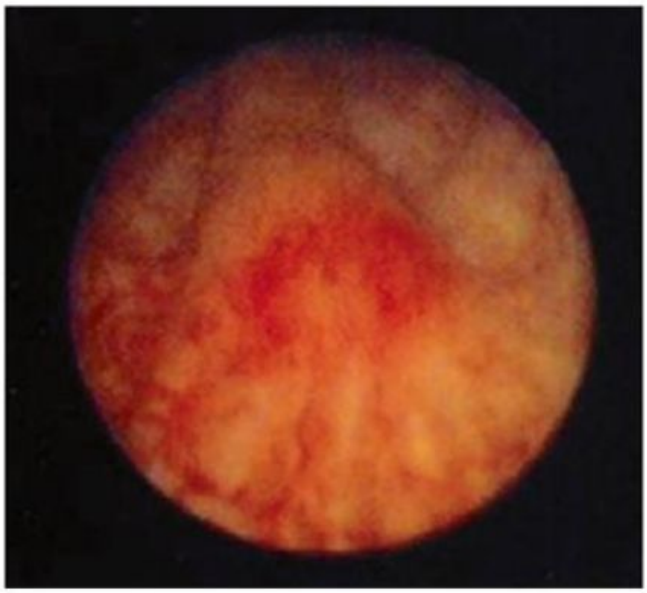

Fig. 11.3 Cystoscopic aspect of a verumontanum located at the trigone

A serious, not previously known anomaly found in 45 % (15 cases) of those patients in whom we performed a cystoscopy was an ectopic verumontanum. Six of them were located in the trigone (Fig. 11.3), five in the bladder neck, and four immediately below the bladder neck. Based on this experience, we now consider it mandatory to do a cystoscopy on these patients to avoid unpleasant future surprises for the patient and the family. When these patients reach adolescence, they have erections and orgasms but they ejaculate into the bladder. Theoretically for these patients to have children, it will require special maneuvers to retrieve the sperm from the urine followed by artificial insemination.

When the clinician makes the diagnosis of the recto-bladder neck fistula type of malformation, he/she should be aware of the fact that he/she is dealing with a patient with a potential serious urologic condition.

11.2.4 Gastrointestinal-Associated Defects

The incidence of esophageal atresia is 15 %, Meckel's diverticulum 2 %, and abdominal wall defects 4 %.

11.2.5 Neurosurgical-Associated Defects

The incidence of tethered cord in these patients is 32 %. In addition, 5 % of the patients suffer from other neurosurgical conditions, including a lipoma and blunted conus.

11.2.6 Cardiovascular-Associated Defects

Fifteen percent of these patients are born with an atrial septal defect and 10 % with a patent ductus arteriosus, 5 % with tetralogy of Fallot, 2 % with tricuspid atresia, and 2 % with pentalogy of Fallot.

11.2.7 Other Associated Defects

Five percent of these patients suffer from hand abnormalities and 5 % from lower extremity abnormalities (equinovarus).

11.3 Diagnosis

We must suspect the presence of these very complex malformations when we see a newborn baby with imperforate anus and flat bottom. The midline groove in between the buttocks that we see in normal children is not present. In addition, we frequently see in these patients a sphincter mechanism located right at the base of the scrotum (Fig. 11.4). The presence of a bifid scrotum (Fig. 11.5) also suggests that the malformation that the baby has is rather complex, most likely very high (recto-bladder neck fistula). The diagnosis is confirmed with a distal colostogram and subsequently by cystoscopy that is performed at the time of the main repair. The distal colostogram can be performed after the colostomy is opened (Fig. 11.6).

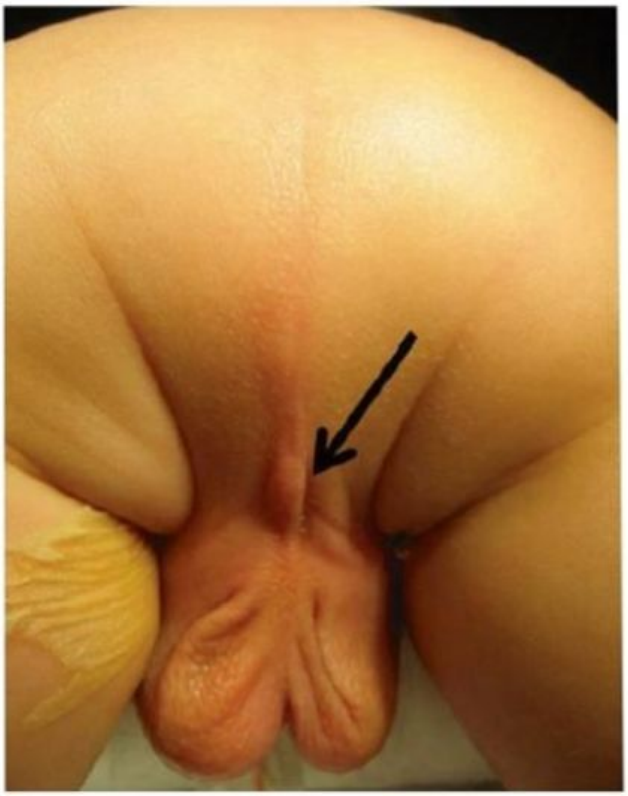

Fig. 11.4 Sphincter located next to the scrotum, frequently seen in cases of bladder neck fistula. *Arrow* showing the center of the sphincter

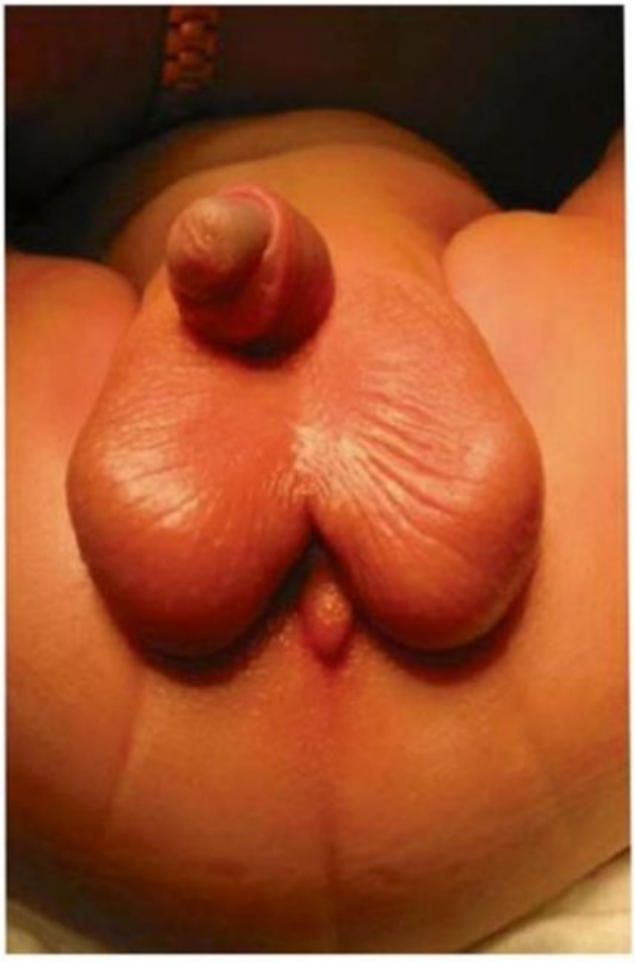

Fig. 11.5 Bifid scrotum, frequently seen in bladder neck fistula

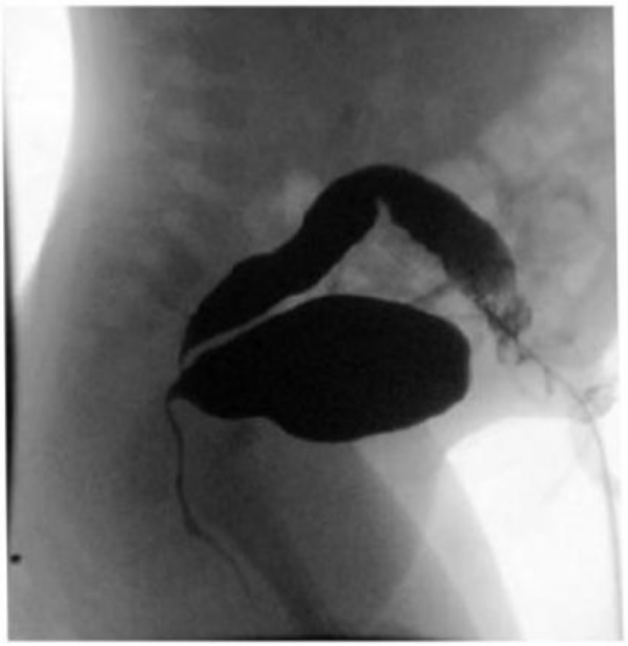

Fig. 11.6 Distal colostogram showing a bladder neck fistula

11.4 Treatment

11.4.1 Colostomy

The type of colostomy that we recommend for patients with recto-bladder neck fistula is the same one recommended for the other types of anorectal malformations. However, emphasis must be placed on being sure that enough length of distal colon is left beyond the mucous fistula (distal stoma) (Fig. 11.7), in order to have enough length of bowel for the pull-through without interference by the colostomy (Fig. 11.8). The fact that these patients have the highest of all defects means that the surgeon will need more length of bowel for the pull-through. Unfortunately, in this particular type of defect is where we have seen more often the most common type of error in making a colostomy (making the stoma too distal in the bowel), leaving a very short piece of bowel for the pull-through (Fig. 11.8).

11.4.2 Main Repair

We perform these operations as soon as we see that the baby is growing and developing normally. If the baby happened to be full term, had a good colostomy, and did not have important

associated defects that interfered with his growth and development, then the patient can be operated, at our institution, within a month after the baby is born. However, as we previously mentioned, many of these patients come to our institution when they are much older, and that is why we have experience with the main repair at different ages. These patients should never be approached surgically without a good-quality, high-pressure distal colostogram that shows how much bowel is available distal to the stoma as well as the exact location of the fistula.

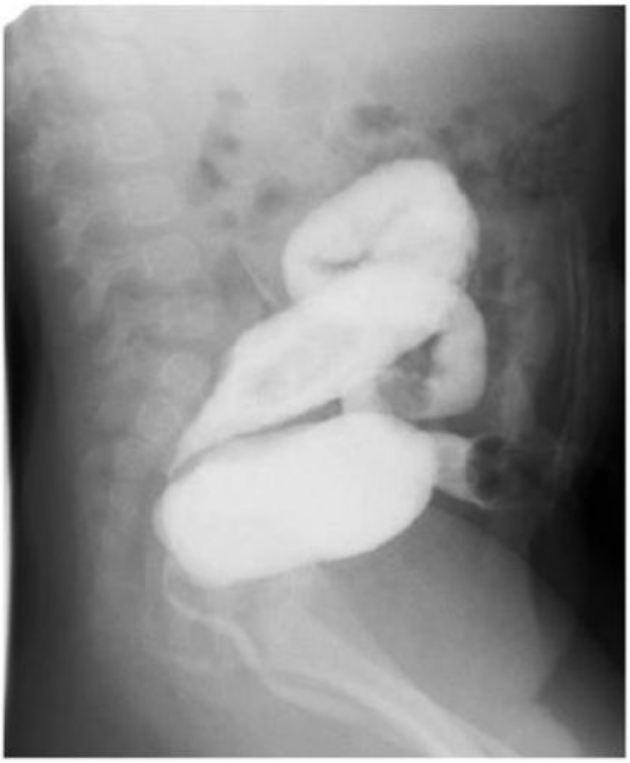

Fig. 11.7 Distal colostogram showing a good length of bowel left distal to the colostomy

If we are dealing with a patient that has a very short piece of bowel distal to the stoma, there is a reason to believe that we will not have enough bowel for the pull-through and that we may have to mobilize the proximal stoma. This is extremely important because knowing this in advance will allow us to plan an adequate procedure. More specifically, we have to prepare the entire gastrointestinal tract (administration of GoLYTELY; see Chap. 7). On the other hand, if the distal colostogram shows that we have enough distal bowel from the mucous fistula, then we are certain that we will not be disturbing the proximal stoma, and all that the patient needs is irrigation of the distal stoma in preparation for the main repair.

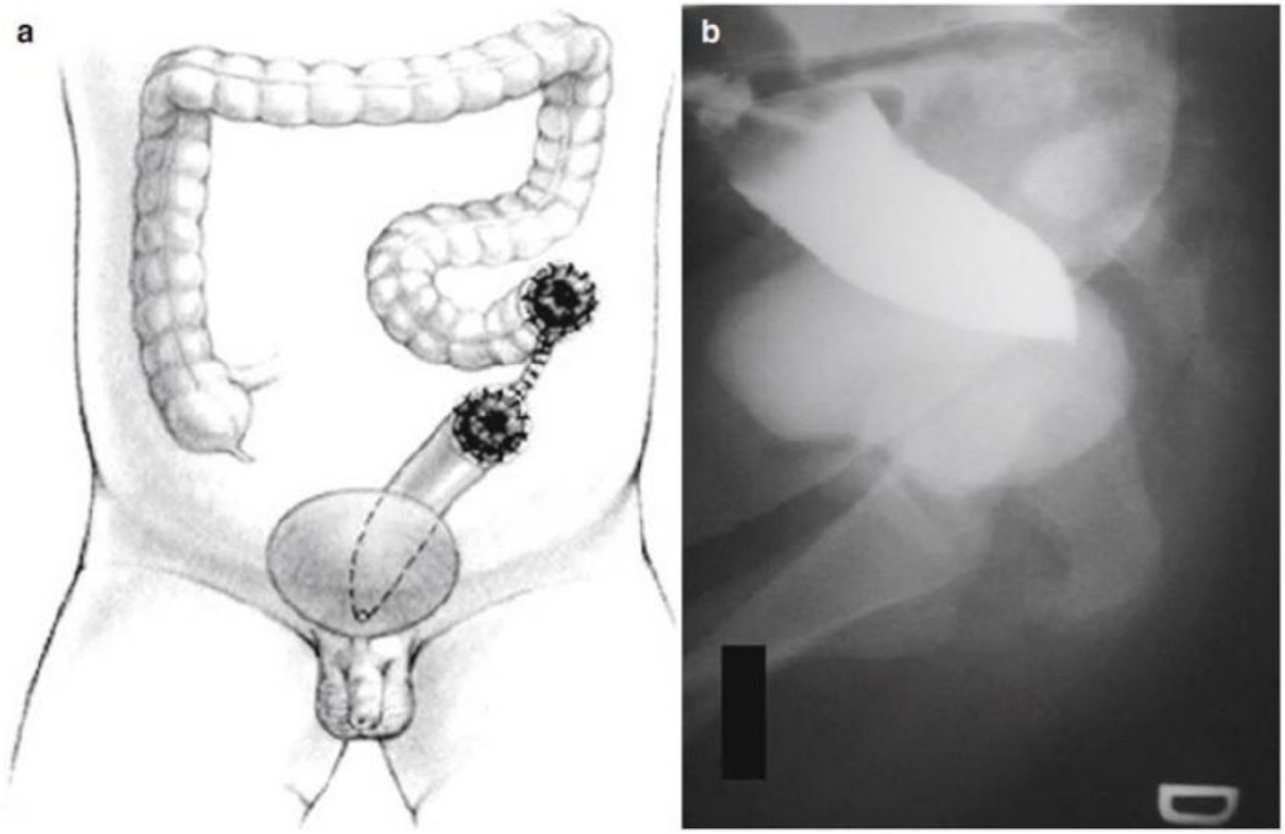

Fig. 11.8 Distal colostogram showing a very short piece of bowel distal to the colostomy (insufficient length for a pull-through). (a) Diagram. (b) Image

The fact that the patient has a recto-bladder neck fistula means that we have to go into the abdomen either by laparotomy, laparoscopy, or both, in addition to the posterior sagittal approach. We start the operation by putting the patient in the lithotomy position and performing a cystoscopy. The cystoscopy is extremely valuable. We have been learning a great deal about the anatomy of the male urethra, bladder neck, and trigone in these patients. As previously mentioned, it is not uncommon to find that these patients have no verumontanum located in the posterior urethra. Rather than that, we find the verumontanum located in the trigone. In retrospect, now we have an explanation for the adult patients that were born with these kinds of defects and have no ejaculation. Further studies demonstrate that they actually ejaculate in the bladder. This must be differentiated from the concept of retrograde ejaculation. We are referring to a patient that has the verumontanum located in the trigone and ejaculates directly into the bladder. This is demonstrated later in life, by finding sperm in the urine after an ejaculation, as well as the cystoscopy (Fig. 11.3). It is not unusual in this type of patients to find, also, ectopic ureters. We have learned that the higher the malformation, the more chances of the patient to have ectopic ureters. The ectopia in this type of defect usually means that the ureters are located closer to the bladder neck or even below the bladder neck into the posterior urethra. Sometimes, the ureters open in the bladder neck and provoke either vesicoureteral reflux, ureterovesical obstruction, or urinary incontinence. When the ureter is ectopic and located into the posterior urethra, usually it is associated with a severe stricture, megaureter, and severe renal damage (Fig. 11.2). It is common for these patients to end up with a nephrectomy. The location of the fistula is frequently visualized at the bladder neck with the cystoscope. A Foley catheter is placed in the bladder.

11.4.3 Laparotomy

A total body preparation is performed on these patients (Fig. 11.9). This means to wash, prep, and drape both lower extremities, the perineum, buttocks, perianal area, lower abdomen, and lumbar portion; in other words, the entire lower body is included in the sterile field. The cautery plate is placed in the back of the patient and is protected with a plastic drape. The arms of the patient are placed in the upward position because they belong to the nonsterile part of the field. Both legs of the patient are covered with stockinettes or with an elastic bandage to avoid loss of temperature. The proximal stoma of the colostomy is packed with packing gauze impregnated with an antiseptic solution to avoid contamination. The skin of the abdominal wall is covered with a plastic drape.

We can start the operation either from below or through the abdomen. We more often now

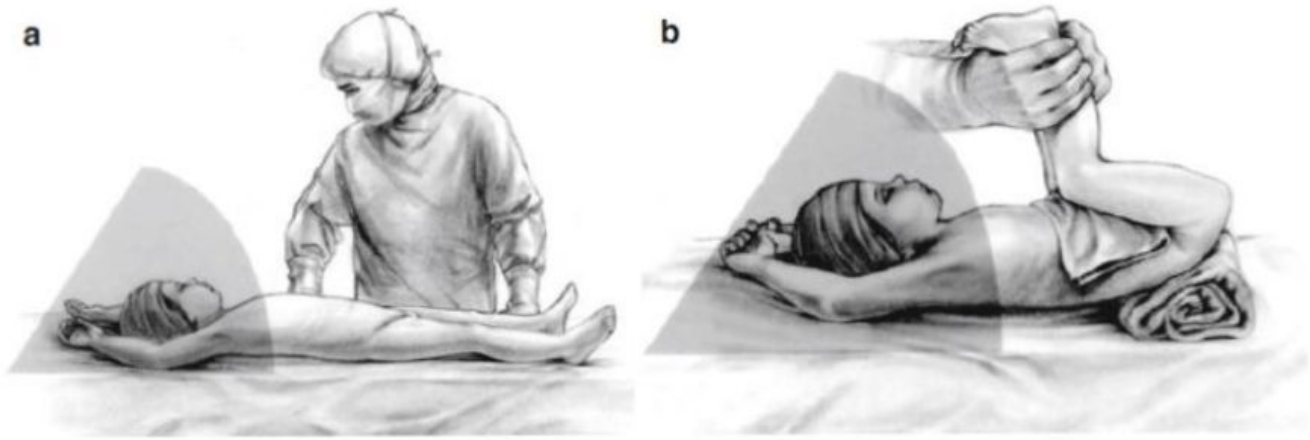

Fig. 11.9 Total body preparation – diagram. (a) Supine. (b) Legs up. (c) Sequence of photographs of bowel preparation. (*a*) Holding legs up. (*b*) Cautery plate up in the back. (*c*) Packing gauze in proximal stoma. (*d*) Wash and prep the entire body below the chest. (*e*) Wash and prep the back. (*f–h*) Sterile sheets on table. (*i, j*) Covering stomas (*k*) Foley catheter inserted

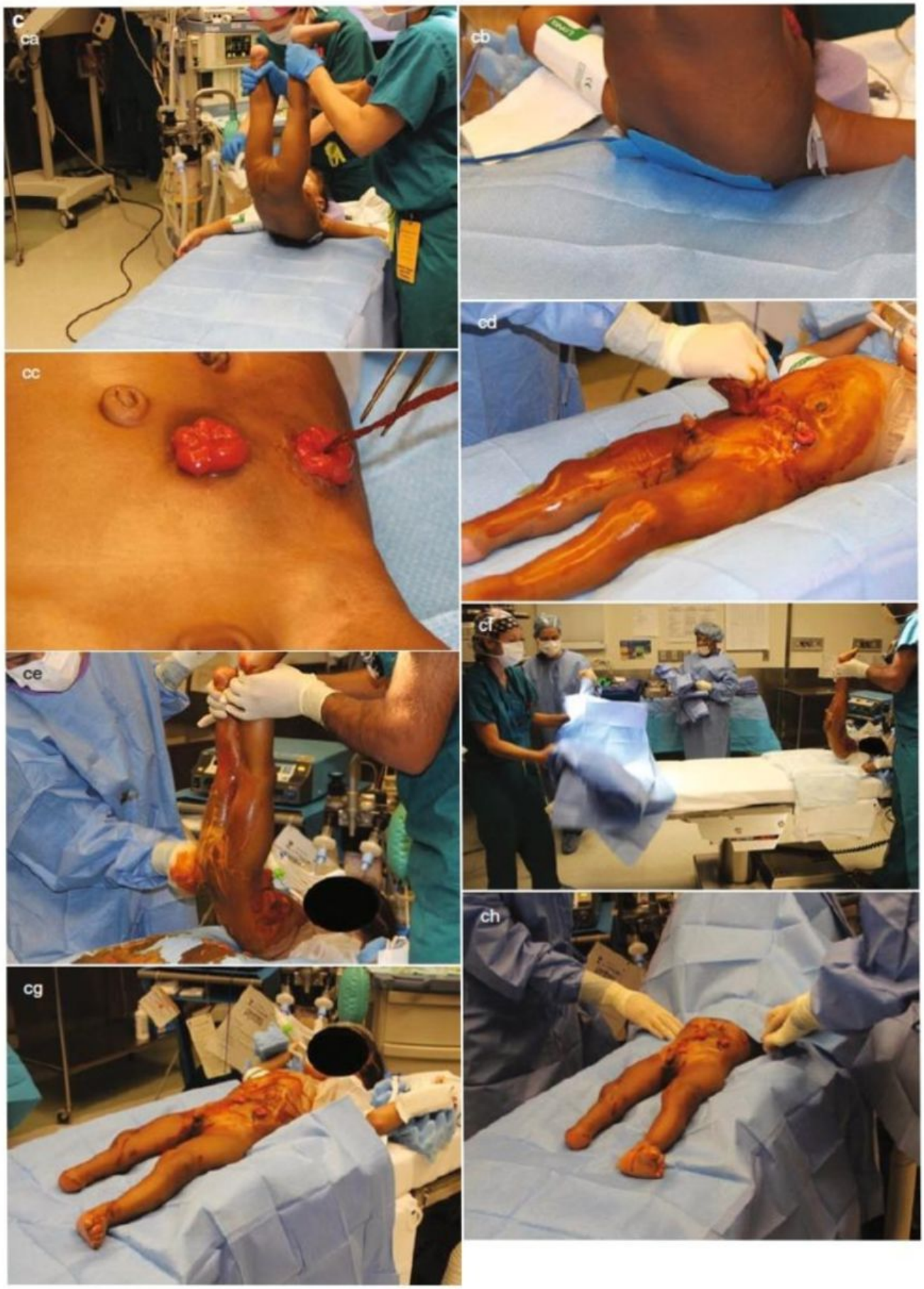

Fig. 11.9 (continued)

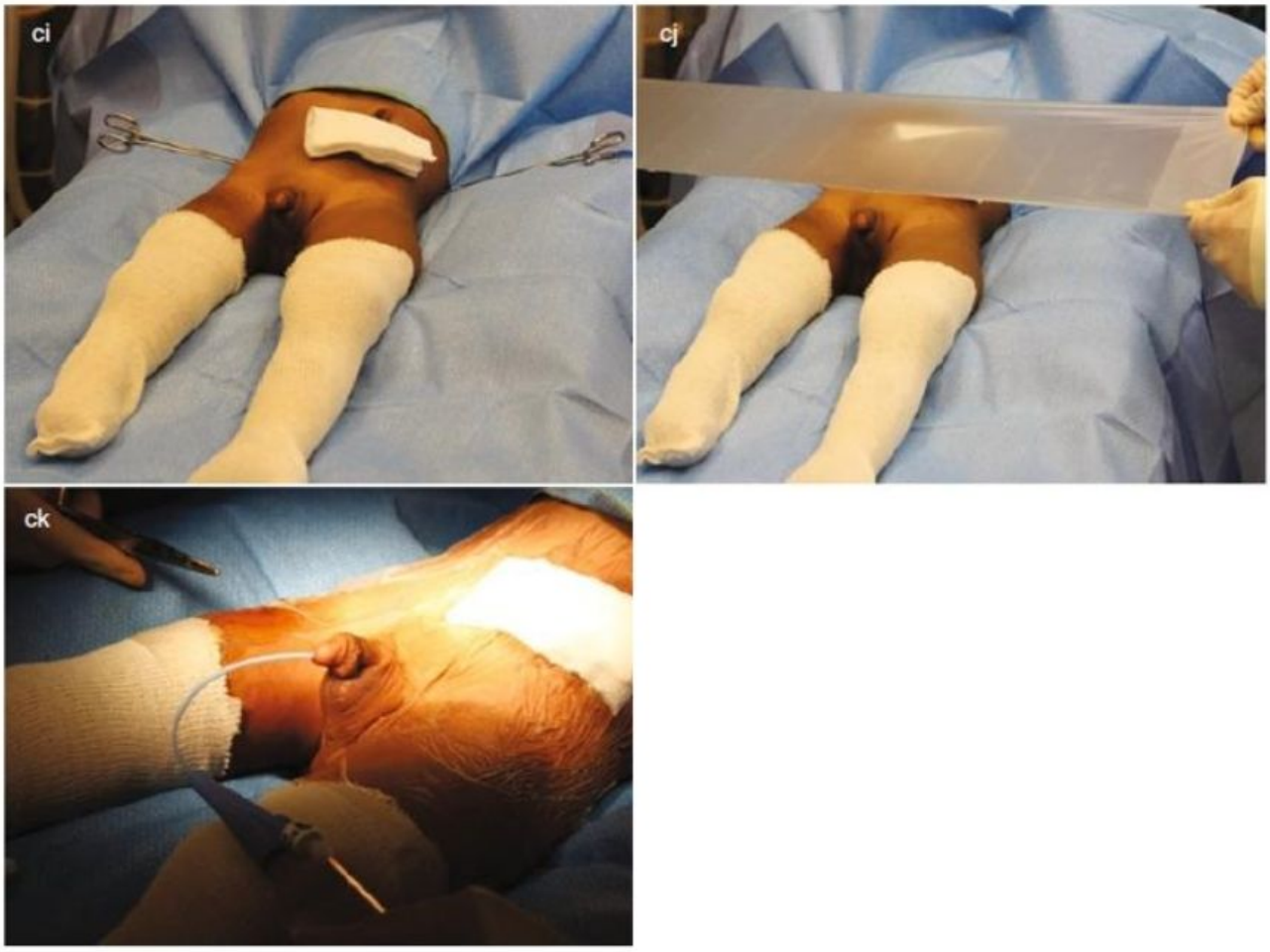

Fig. 11.9 (continued)

approach the abdomen first which can be done by laparotomy or laparoscopy. We consider this particular defect to be a good indication for a laparoscopic approach. More information related with the indications for laparoscopy in anorectal malformations can be found in Chap. 13. The abdomen is entered through a midline incision running from the umbilicus down to the pubis. A needle-tip cautery is used changing from cutting to coagulation to provide meticulous hemostasis. The peritoneal cavity is entered. The urachal remnant and obliterated umbilical arteries are identified and divided. A clamp is placed on the urachal remnant of the bladder to apply caudal traction. The lateral avascular attachments of the bladder to the abdominal wall are divided with cautery to have easy access to the lower pelvis. By pulling on the bladder out of the abdomen and toward the pubis, caudally, we can see the posterior wall of the bladder as well as the peritoneal floor, sigmoid, both vas deferens, and ureters (Fig. 11.10). Both vas deferens seen behind the bladder run distally toward the bladder neck; the ureters are seen retroperitoneally, and they also seem to be running toward the bladder neck. Our specific recommendation is to place a 4-0 silk stitch on the anterior wall of the sigmoid to apply traction. About 1 or 2 cm from the peritoneal floor, the serosa of the anterior wall of the sigmoid is divided in order to create a plane of dissection as close as possible to the bowel wall, but without damaging it. This plane of dissection is followed all around the bowel, separating the mesenteric fat from the sigmoid. Once we have created a plane all around the bowel, a Silastic vessel loop is passed around the rectum in order to have a more effective handle for traction (Fig. 11.11a). Applying traction on the vessel loop, it is very easy to continue a circumferential dissection of the bowel distally. Very soon, within a centimeter or two from our initial dissection, one can appreciate that the bowel decreases in size and becomes

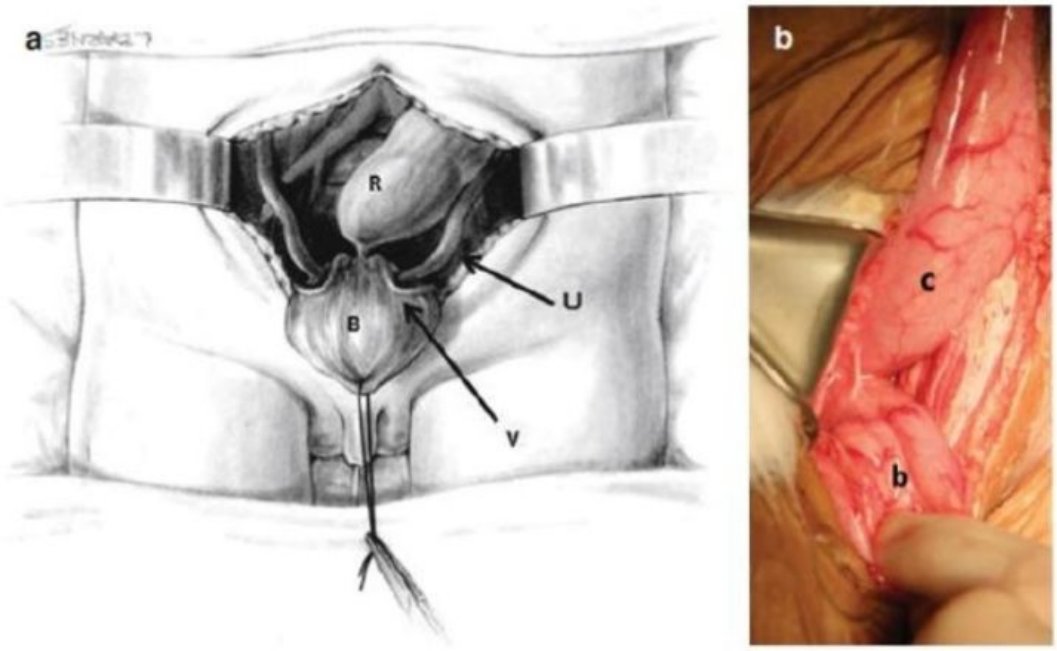

Fig. 11.10 View of the peritoneal floor in a bladder neck fistula. (a) Diagram – *b* bladder, *r* rectum, *u* ureter, *v* vas deferens. (b) Photograph – *c* colon, *b* bladder

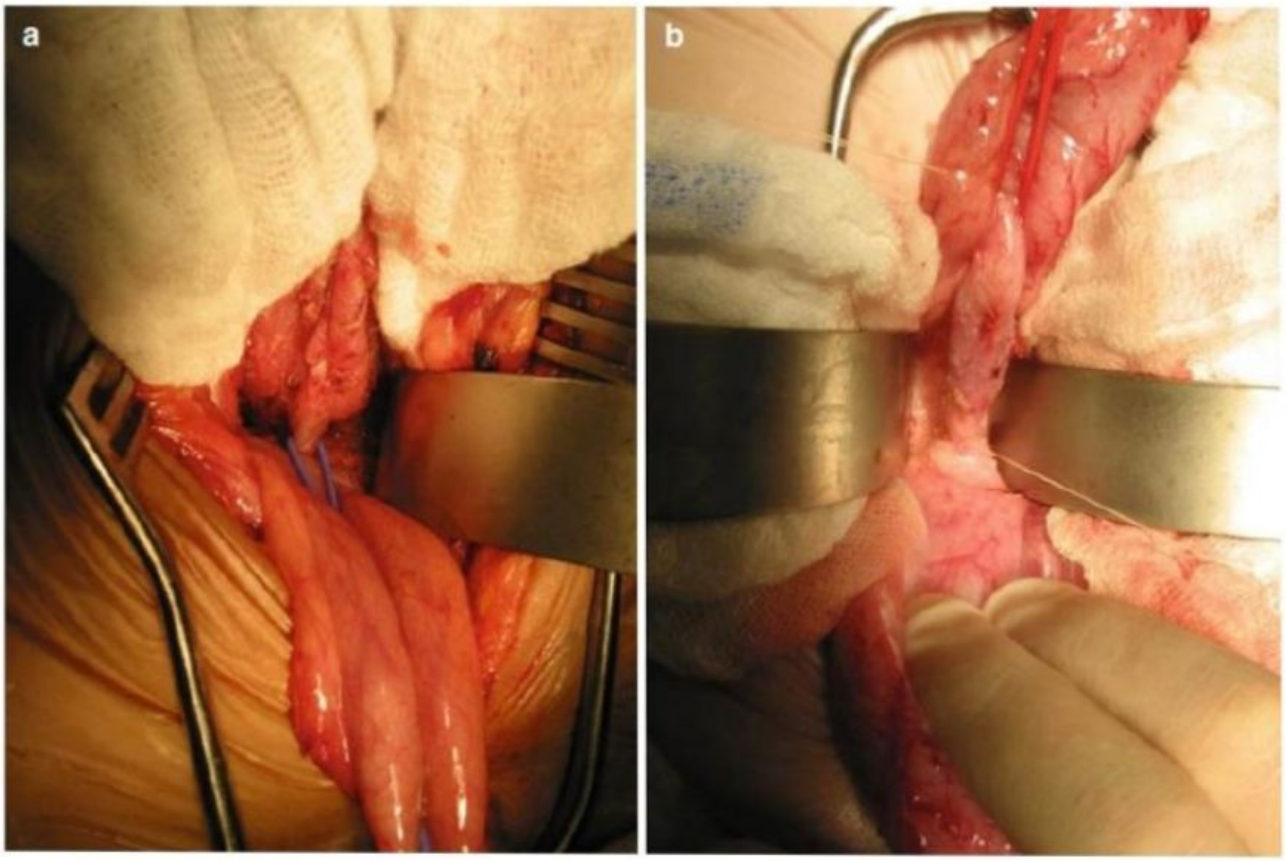

Fig. 11.11 Distal rectum dissected down to the fistula. (a) Vessel loop surrounding the fistula. (b) Sutures to close to the fistula

very narrow, indicating that it is reaching the bladder neck. One does not have to be very precise in trying to determine exactly the location of the end of the fistula and the beginning of the urinary tract. Once the rectum starts being narrow, it reaches a point where the diameter is about

3–4 mm, obviously not useful for the reconstruction of an anus, and, therefore, that means that we can divide the rectum right there. We must keep in mind that because we are applying traction on the bowel, there is a possibility that we are kinking the bladder neck or the upper posterior urethra, and therefore, when we divide the fistula, actually, we will be dividing the bladder neck or the posterior urethra. Therefore, the traction must be gentle. Two 5-0 Vicryl stitches are placed in both sides of the fistula site in order to avoid retraction, and the fistula is divided. The distal end of the rectum is also sutured with a running 5-0 Vicryl to avoid contamination from mucus or meconium previously left in the bowel. The fistula is closed with three to five 5-0 Vicryl sutures (Animation 11.3) (Fig. 11.11b).

Once the rectum has been separated, the next step is to divide the avascular attachments of the distal rectum to evaluate and determine the location of the mesenteric vessels (Fig. 11.12). At this stage, it is very easy to appreciate that the main limitation for the pull-through of the rectum is its blood supply provided by the branches of the inferior mesenteric vessels.

Traditionally, we surgeons learn that we can mobilize different parts of the colon (up to the neck or down to the perineum) provided we are familiar with the blood supply of the colon in normal individuals, which is represented by three main sources: (1) the ileocecal vessels, (2) the mesocolic vessels, and (3) the left colic vessels (Fig. 11.13). Once the rectum goes below the peritoneal reflection, its blood supply is provided by the hemorrhoidal vessels, which are branches of the internal iliacs. We also know that the three main vessels that irrigate the colon are intercommunicated by a vascular arcade. As a consequence, we can easily divide, let's say, the middle colic vessels without interrupting the blood supply of the rest of the colon, provided we preserve intact the other two sources of blood supply (ileocolic and left colic) and the vascular arcade that intercommunicates the three systems. We can equally divide the left colonic and inferior mesenteric vessels, preserving the arcade; the most distal part of the colon will survive receiving blood from the middle colic vessels. We can do the same with the ileocolic on the right side. This is a general notion. However, we must warn surgeons about the limitations that this concept has in patients with anorectal malformations. We must keep in mind that we are dealing with patients whom already had a colostomy. Most of the time, the opening of a descending or sigmoid colostomy included the ligation of the colonic vascular arcade. This means that the most distal portion of the rectosigmoid receives all of its blood supply from the inferior mesenteric vessels. The obvious recommendation is do not ligate the inferior mesenteric vessels and to bring the rectosigmoid down because doing that may represent the loss of that bowel (Fig. 11.14). We have learned that fortunately, the rectum has an excellent intramural blood

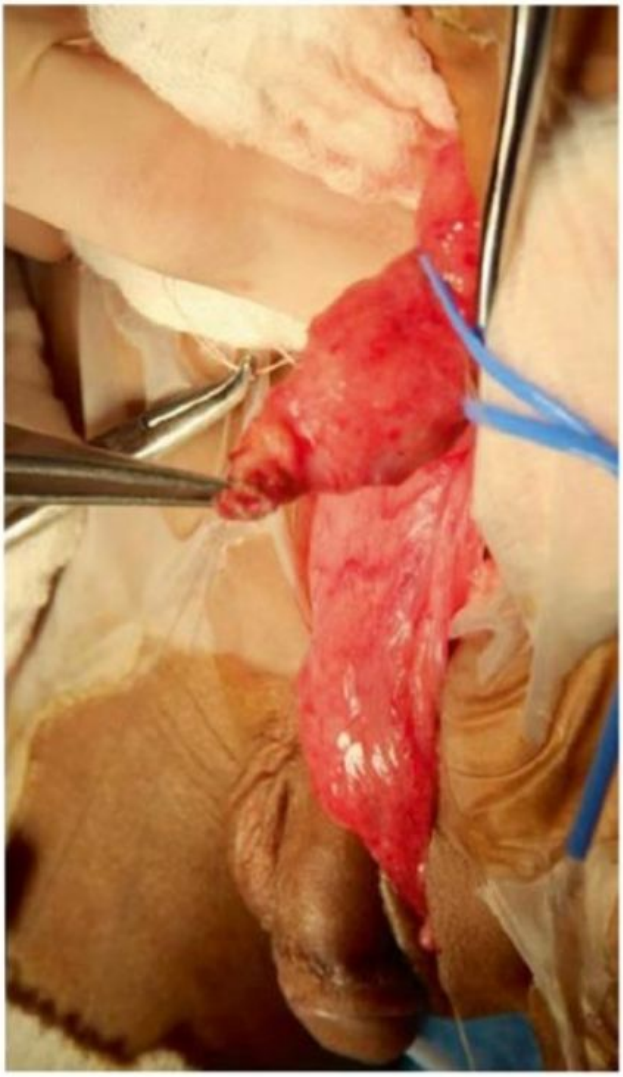

Fig. 11.12 Photograph of rectum separated from the urinary tract, very high, does not reach the perineum

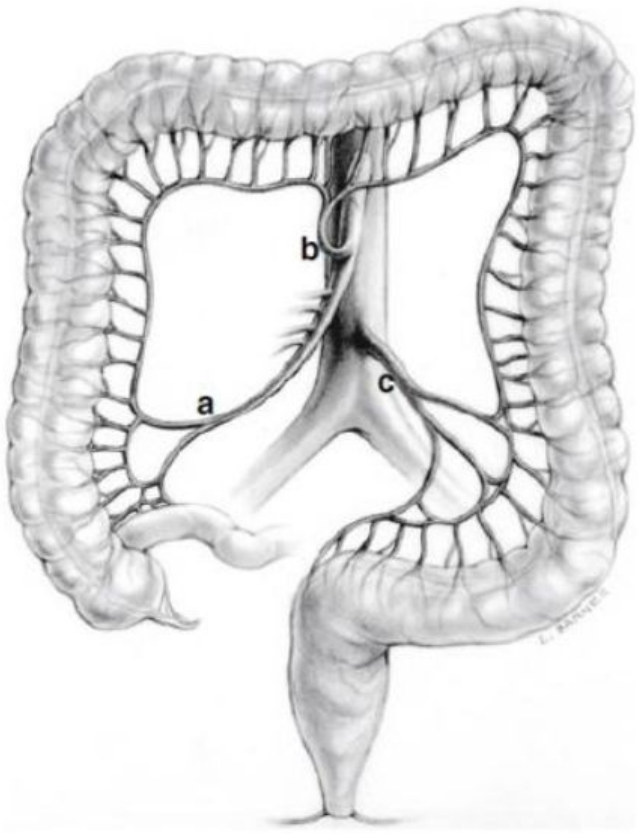

Fig. 11.13 Diagram showing the normal blood supply of the colon. (a) ileocecal vessels, (b) Middle colic vessels, (c) Left colic vessels. A vascular arcade, joins the three systems

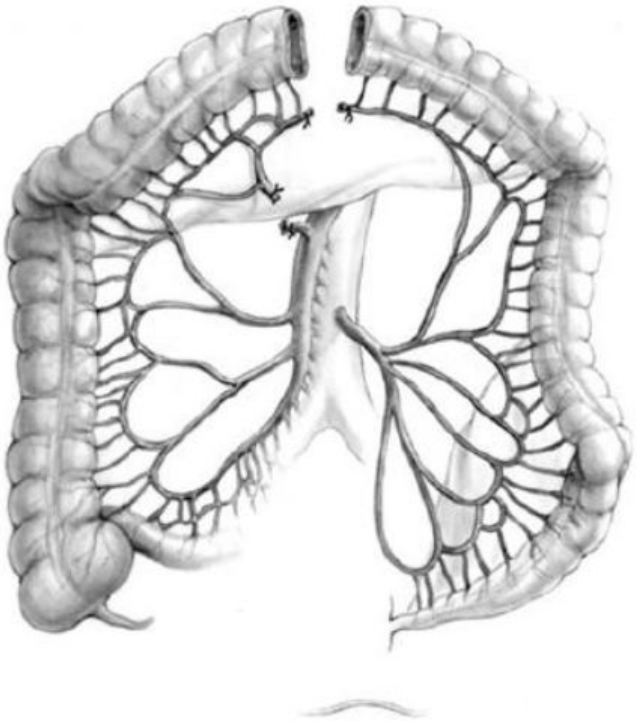

Fig. 11.14 Diagram showing the blood supply of a rectosigmoid in a case with a colostomy and ligated vascular colonic arcade

supply, which allows sacrificing all of its extrinsic vessels without compromising its vascularity provided the bowel wall is maintained intact and the inferior mesenteric vessels are not ligated. In other words, we can ligate several peripheral branches of the inferior mesenteric vessels, being sure to preserve at least one or two proximal branches (Fig. 11.15 and Animation 11.3). All this, provided we maintain intact the integrity of the rectal wall. Damaging the rectal wall interferes with the intramural blood supply, and the distal blood supply suffers. Every time we ligate one of the peripheral branches of the inferior mesenteric vessels, we do it very close to the rectal wall, and that allows us to gain length. We must be sure to visualize that at least one good branch from the inferior mesenteric vessels remains intact, reaching the bowel wall and that the bowel wall remains intact; by doing that, the bowel blood supply is going to be good. Following those recommendations, we have been able to pull down all of these rectums, even when they were located very high (Figs. 11.15 and 11.16).

When we separate the bowel from the bladder neck, as previously mentioned, we divide the avascular attachments of the bowel in order to identify the mesenteric vessels. It is very easy to pull on the bowel and identify exactly what is limiting the pull-through. At the beginning, one can see that what is limiting us are the vessels, and we can selectively ligate the peripheral branches of the mesenteric vessels as close as possible to the bowel and see how we gain more and more length until we have the necessary length for the pull-through. However, occasionally in the process, we find that we are no longer limited by the vessels but rather limited by the colostomy itself. Under those circumstances, we must take the colostomy down. To do that, multiple 5-0 silk stitches are placed at the mucocutaneous junction of the mucous fistula. If the patient has a loop colostomy (which we consider contraindicated in anorectal malformations), then, unfortunately, we had to take down the entire colostomy, which makes the procedure more complex. If, on the other hand, the patient

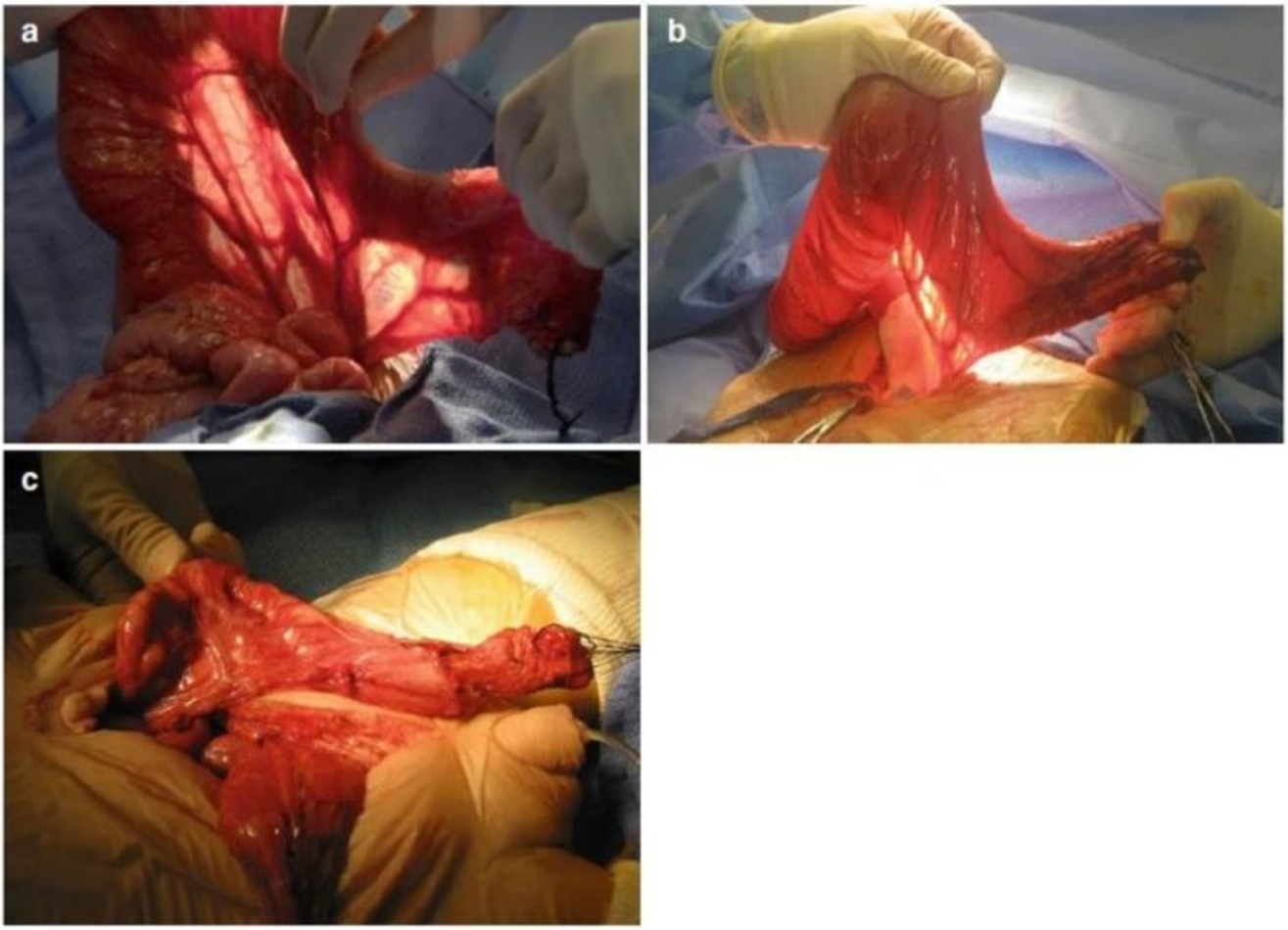

Fig. 11.15 Intraoperative photographs showing how to evaluate the blood supply of a very high rectum. (a) Before dividing vessel. (b) Dividing vessels, preserving the arcade. (c) Gained length

has separated stomas, we only have to take down the mucous fistula. By doing that, we may have enough length of bowel to reach the perineum. We can go ahead with the pull-through and decide whether to leave the upper part of the pulled-through bowel closed, as what is called "Hartmann pouch," or to close the colostomy and do the pull-through, leaving the patient without a protective colostomy and a colonic anastomosis in the pelvis (Fig. 11.19). If one decides to leave it as a "Hartmann pouch," we want to be sure that the length of the distal bowel is enough for the blind upper end of the bowel to be found above the peritoneal reflection at the time of the colostomy closure; otherwise, it may become a technically demanding type of procedure.

In order for us to learn whether or not there is enough length of colon for the pull-through, we can open from below and see exactly if we have enough length passing the rectum behind the posterior urethra. If the perineum has not been opened yet, we can guess whether or not there is enough length by pulling the bowel outside of the abdomen caudally toward the genitalia. We have learned that we have enough distal bowel to reach the perineum if the distal end of the bowel reaches about 4 cm below the lower edge of the pubic bone (Fig. 11.16). If we do not have this kind of length, that means that we have to work more on the blood supply or to take down the colostomy in order for the bowel to reach.

The perineal approach can be done in two ways. One is simply lifting the legs up, putting a bulky roll below the pelvis of the patient. By doing that, the perineum of the patient is well exposed, horizontally, and we can work comfortably (Fig. 11.17c). The incision that we make in these patients does not have to be a full-length posterior sagittal one. An incision that runs from the base of the scrotum and about 5 or 6 cm posteriorly usually provides plenty of exposure to create a safe abdominal perineal path. Our

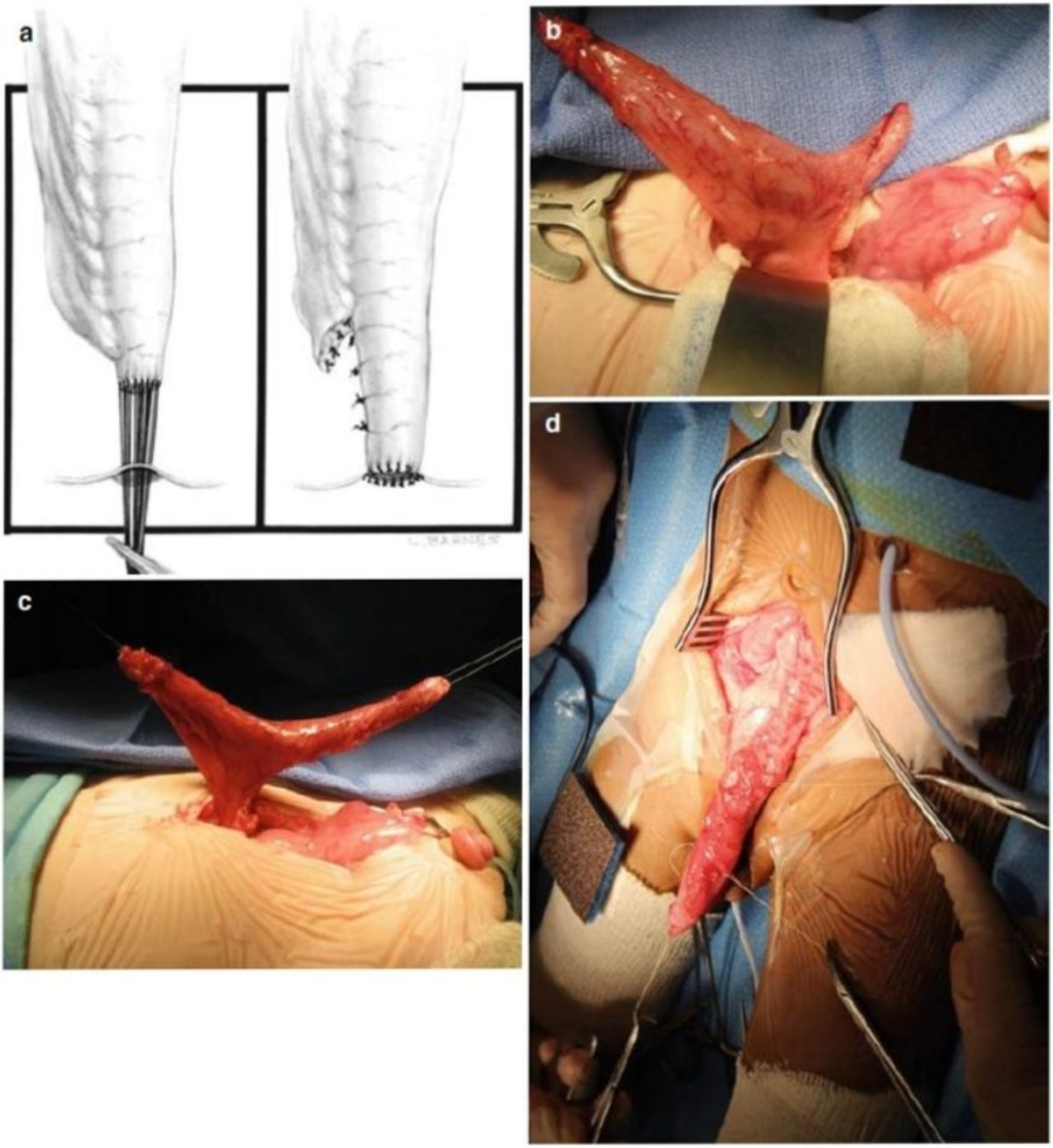

Fig. 11.16 How to gain length in a case of a very high rectum. (**a**) Diagram – divide peripheral branches of the inferior mesenteric vessels. Maintain intact rectal wall. Blood supply of the rectum is provided by intramural vessels. (**b**, **c**) Intraoperative picture of the same maneuver. (**d**) Bowel reaches the perineum

incision goes through the skin, parasagittal fibers, muscle complex, and levator mechanism. However, these patients often have very poor sphincter mechanism, and sometimes it is very difficult to identify each one of the components of the sphincter mechanism. In addition, we find different degrees of "caudal regression." This means that the pelvis may be extremely narrow, making a very difficult task to accommodate a rectum through it. The entire procedure is done with a Foley catheter in place. In the process of opening the perineum, we frequently stop to palpate the catheter in the urethra located in the deepest portion of the "V" formed by the pubic bones. As we progress deeper through the posterior sagittal incision, after we have divided the

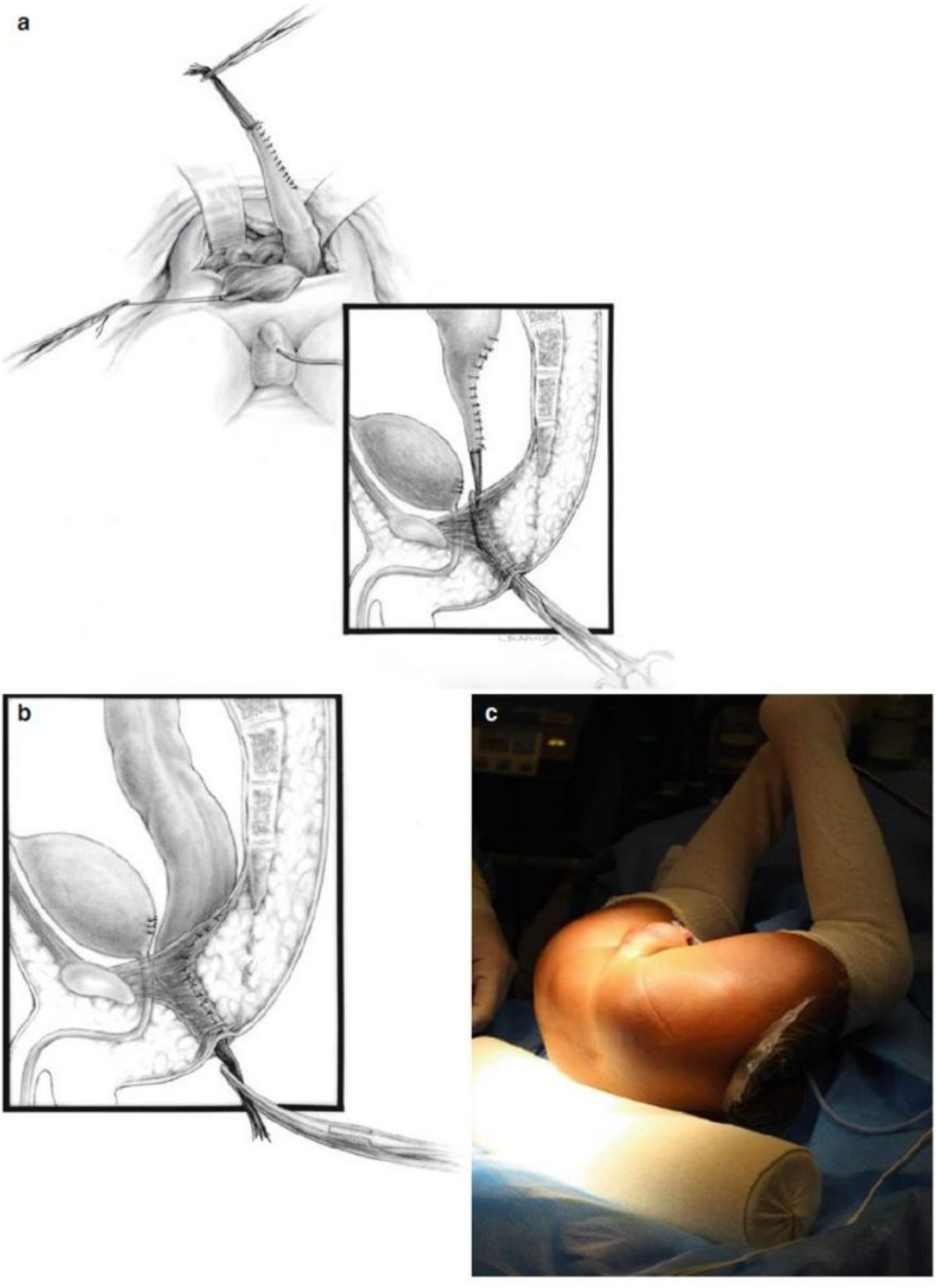

Fig. 11.17 Diagram showing the pull-through. (**a**) Pulling the rectum. (**b**) Rectum pulled down. (**c**) Photograph showing the approach to the perineum. Legs up. (**d**) Posterior sagittal incision. (**e**) Rectum pulled down. (**f**) Anoplasty

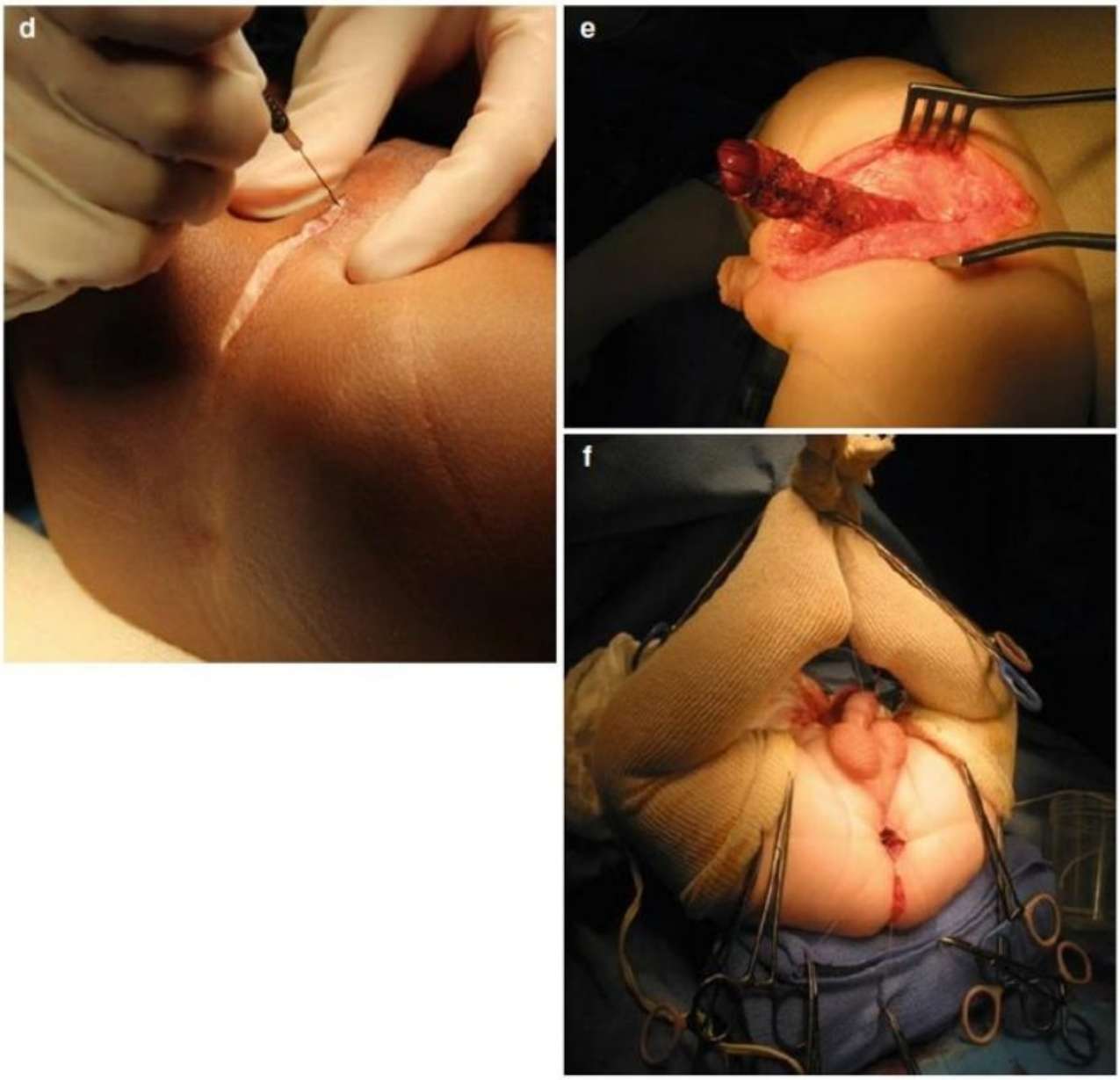

Fig. 11.17 (continued)

entire striated sphincter mechanism, we find a whitish fascia which represents the entrance to the abdominal cavity. A safety path is obtained remaining as much as possible in the midline. Once we enter into the abdomen, we pass a clamp to grasp the distal end of the bowel when dealing with a laparoscopic approach or to grasp the sutures holding the distal rectum when the abdomen was opened (Fig. 11.17). Looking from the abdominal side, we must remain away from the ureters and the vas deferens. This space has to be wide enough to avoid compression of the rectum. The rectum then is pulled under direct vision, and the anoplasty is performed within the limits of the sphincter as previously demonstrated in the other chapters. Sometimes, in these operations, we find a minimal amount of a sphincter mechanism, and therefore, the location of the anus is determined in a rather arbitrary way. Many of these patients will have a poor prognosis anyway, due to the lack of a sphincter, poor sacrum, tethered cord, and other spinal abnormalities.

Prior to the abdominal closure, we must close the defect created between the mesentery of the pulled bowel and the posterior abdominal wall (Fig. 11.18). We had experience with two cases in whom that space was left open and the patients

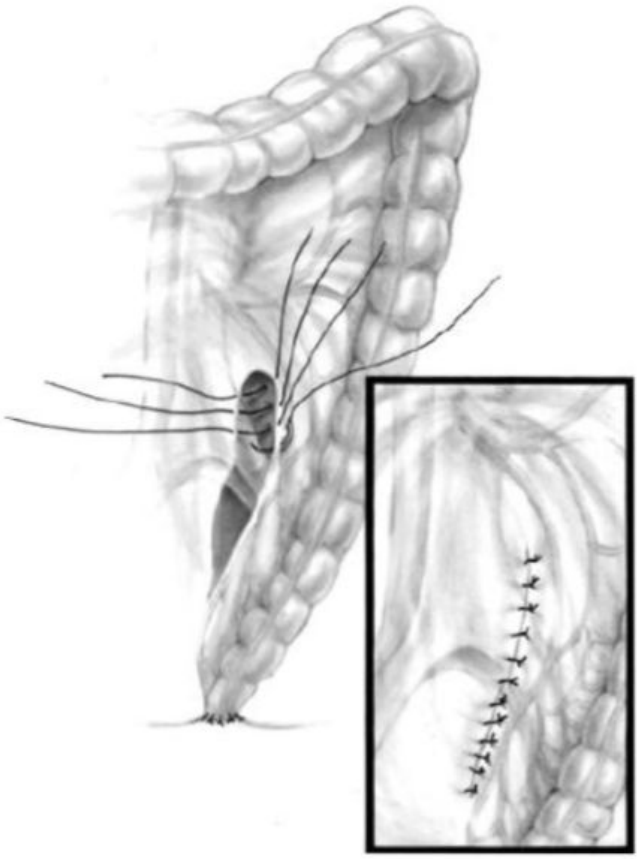

Fig. 11.18 Closing the mesenteric defect

suffered from intestinal obstruction within the first 5 days postoperatively. They required an emergency laparotomy to reduce multiple loops of small bowel which were trapped in that defect.

Also, before we pull the colon through, we evaluate the degree of dilatation of the rectum; if it is considered to be too bulky, it must be tapered resecting a portion of the posterior wall of the rectum and closing with two layers of interrupted sutures.

The posterior wall of the rectum must be anchored to the neighbor tissue with 5-0 long-term absorbable sutures. If the patient gets a good muscle complex, the rectum is anchored to the muscle complex as demonstrated in other malformations. The anoplasty is performed as previously described for other malformations.

Another way to do the posterior sagittal portion of the operation consists in packing the abdominal wound, covering it with a plastic drape, turning the patient into prone position, and opening posterior sagittally as previously described. However, more and more, we do not have to do this, but rather to lift the legs up in the way we have already described.

The abdominal wall is closed, and the patient usually starts eating as soon as the colostomy is working. The patient stays in the hospital 2 or 3 days. If, on the other hand, the patient was left with no colostomy, he will remain 7–10 days with nothing by mouth receiving parenteral nutrition.

11.4.4 Laparoscopy

The laparoscopic approach of anorectal malformation was first proposed by Willital [2]. Then he was followed and popularized by Georgeson et al. [3] and many other surgeons who are performing the laparoscopy approach for the treatment of anorectal malformations [4–31]. The classic and indisputable indication of a laparoscopy is an operation that requires an abdominal approach. In other words, the laparoscopy serves the purpose of minimizing the trauma and the pain produced by the incision in the abdomen. Because of this, the laparoscopic approach is indicated to treat this particular malformation. The exposure and view of the peritoneal floor obtained laparoscopically is excellent (see Chap. 13). The dissection of the distal rectum is easily done, as previously described, until the rectum becomes narrow. At that point, unfortunately, the division of the fistula cannot be done as accurately as when it is done with a laparotomy. Yet, we have not seen complications from the laparoscopic ligation of the fistula. It is important to keep in mind that in this particular type of anorectal malformation (recto-bladder neck fistula), the rectum reaches the bladder neck in a "T" fashion. In other words, there is no common wall between the rectum and the urinary tract located above the location of the fistula, like it happens in the cases of prostatic fistula and even more in cases of rectal urethrobulbar fistula. Our observations in 1,113 surgical repairs of anorectal malformations in male patients allowed us to learn that the lower the malformation, the longer the common wall between the rectum and the urinary tract. Therefore, in the highest of all defects (recto-bladder neck fistula), the dissection of the

rectum is easier, and it does not include the risk of injuring the urinary tract. In lower malformations such as rectoprostatic and particularly in bulbar fistula, the common wall between the rectum and urinary tract is much longer, and therefore, it is not that easy simply to ligate the fistula like in the laparoscopic approach in cases of recto-bladder neck fistula. That is one of the reasons why we consider the laparoscopic approach formally contraindicated in patients with recto-urethral bulbar fistulas.

The separation of the rectum from the bladder neck and the ligation of the fistula are easy maneuvers. The mobilization of the rectum and the cauterizing of the vessels to allow the rectum to reach the perineum without undue tension, on the other hand, may not be so easy. The burning of selected mesenteric vessels may not be as accurate as when it is done with an open abdomen; accidental burning of important neighbor vessels may occur. In addition, it is not uncommon to find that the rectum is too bulky to be placed within the limits of the sphincter. A tapering of the rectum is required in such cases. It is at this point when sometimes we have decided not to continue the laparoscopic approach and go into to a formal laparotomy. In some cases, we have concluded the entire procedure laparoscopically successfully.

11.5 Special Problems

11.5.1 Dealing with Inadequate Colostomies (Too Distal)

When the colostomy is located too distal in the colon, it is technically demanding to bring the distal rectum down to the perineum preserving its blood supply. We have learned how to do it, but sometimes the upper part of the rectum has to be detached from the abdominal wall in order to be pulled down. Once the pull-through is completed, it may occur that the upper end of the rectum ends up being located in the area of the posterior urethra. If we leave it there, it would become an impossible task to close the colostomy (Fig. 11.19). At that point, we have to make a decision. One possibility would be to resect the distal piece of bowel and take the proximal stoma down as a pull-through. In general, we do not like to do this because that means the patient will lose its natural bowel reservoir which will give him a tendency to have diarrhea, making the bowel management to keep him artificially clean more difficult. Another possibility would be to take the proximal stoma, separate it from the abdominal wall, close the colostomy, and pull together down to the perineum, the distal bowel with what used to be the proximal stoma attached and anastomosed. If we do something like that, we have to make a decision to (a) keep the patient postoperatively without a colostomy, with parenteral nutrition, and nothing by mouth for 10 days or (b) open a more proximal colostomy (Fig. 11.20). The decision is a clinical one and will depend on how secure the surgeon feels about the blood supply of the distal rectum and the surgical technique observed in general. We have only removed one rectum in cases like this, because the patient had only a 4 cm portion of bowel, and if we anastomosed it to the proximal stoma, the anastomosis would be located too low. We felt that it was an unnecessary risk to do that and preferred to pull down the colostomy itself. When we did that, we had to keep the patient 10 days with nothing by mouth or to open a more proximal colostomy. We almost never open a proximal colostomy. We have, rather, kept the patient with nothing by mouth for 10 days.

Another reason to open the abdomen even in cases of prostatic or bulbar fistulas is when the colostomy interferes with the pull-through of the rectum because this has been created too distally. We have seen this happening very often.

11.6 Functional Results

Our experience includes 110 patients. The functional evaluation is only done in patients older than 3 years of age and that have been in touch with us.

11.6.1 Fecal Control

Forty-seven patients were evaluated after the age of 3, and we found that 12 of them (25 %) had

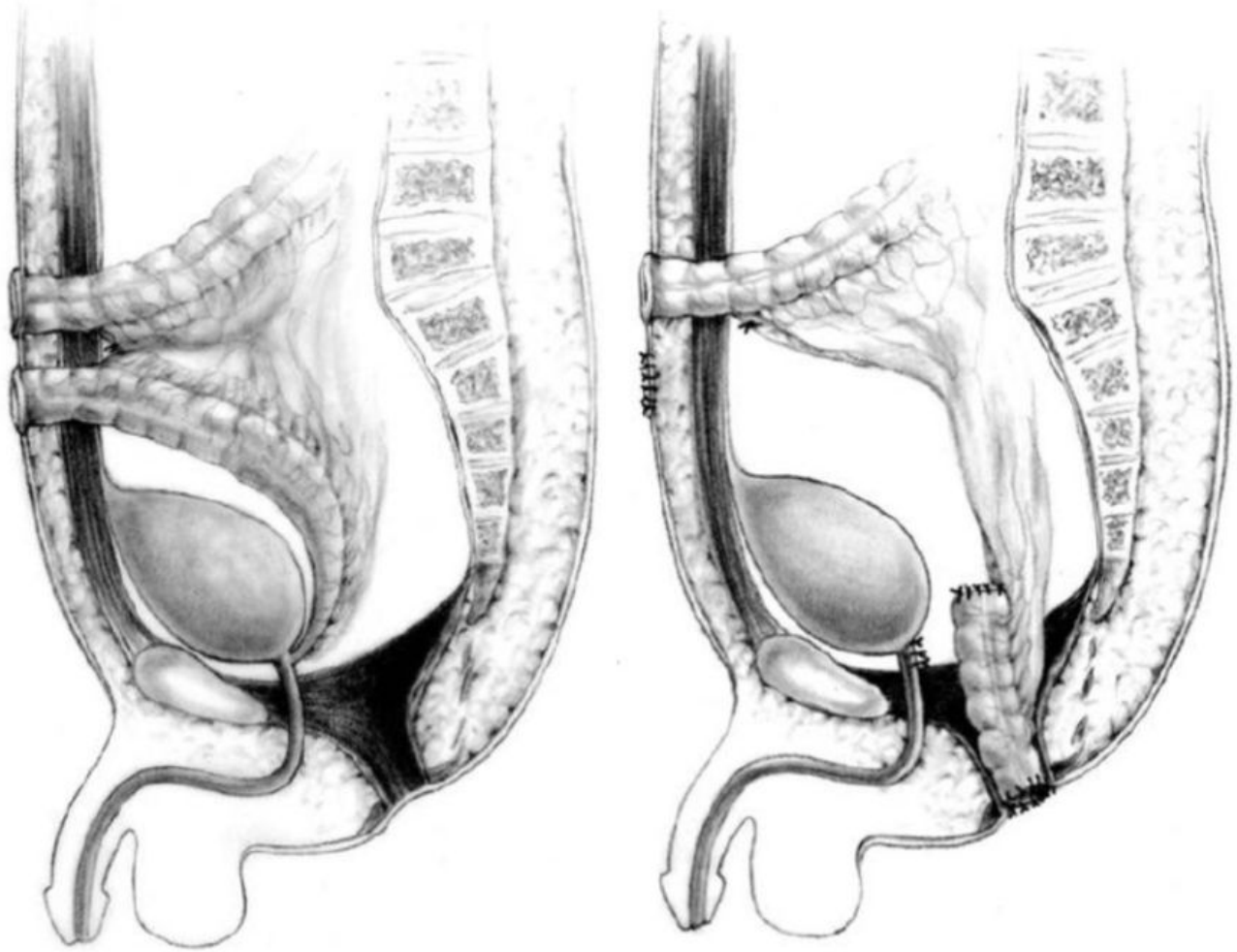

Fig. 11.19 Diagram showing the pull-through of a very short piece of rectosigmoid, which will make the colostomy closure a very difficult operation

voluntary bowel movements. Ninety percent of these patients soiled occasionally in the underwear. Only 10 % were totally continent. All patients that received bowel management were kept totally clean in the underwear. Among 21 patients with sacral ratio of 0.7 and up, 7 (33.3 %) had voluntary bowel movements. Patients with sacral ratio of 0.41–0.69, (20 %) had voluntary bowel movements. None of the patients with a sacral ratio lower than 0.4 had voluntary bowel movements.

11.6.2 Urinary Control

Forty-nine patients were available to evaluate urinary control. Thirty-nine of them (78 %) had urinary control. Among 19 patients with sacral ratio higher than 0.7, 13 (68.4 %) had urinary control. Twenty-two patients had a sacral ratio of 0.4–0.69, and 18 of them (81.8 %) had urinary control. When the sacral ratio was less than 0.4, three out of 13 patients (23 %) had urinary control. The fact that a significant number of patients have urinary control does not mean that the urinary tract is working properly. Some patients have urinary control but cannot empty the bladder well. In addition, a significant number of cases with this malformation suffer from vesicoureteral reflux. That explains why a significant number of patients 17/54 (31 %) are treated with clean intermittent catheterization.

We consider it extremely important to alert and to warn the parents about the future of these babies as soon as we make the diagnosis of recto-bladder neck fistula. This is extremely important in order to adjust the expectations of the parents concerning the future of the baby and to avoid further frustration.

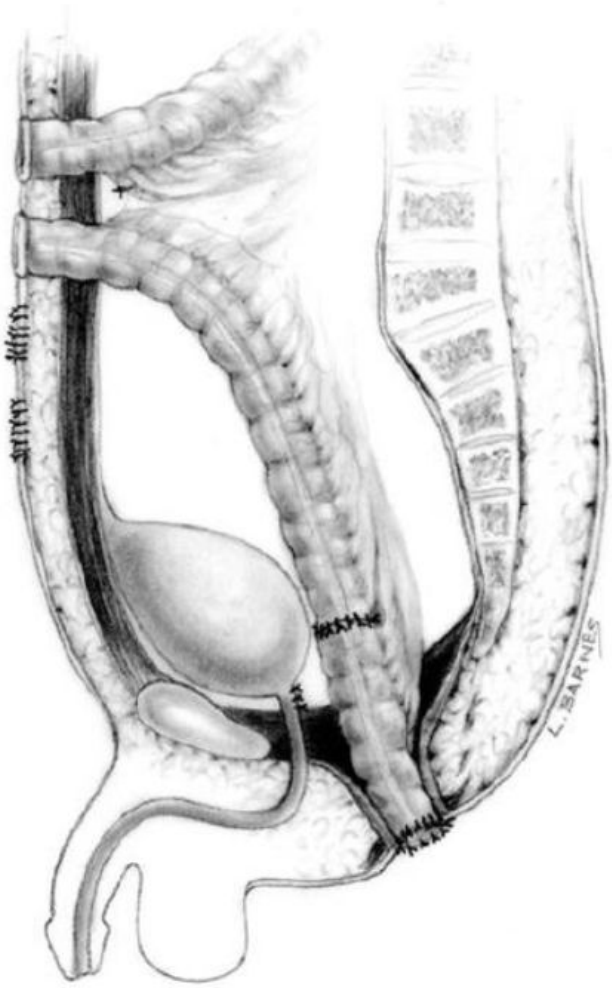

Fig. 11.20 Diagram showing a pulled-through short piece of rectum with a colostomy closure and opening of a more proximal colostomy

Once we make the diagnosis of this malformation, we tell the parents what we know about the future bowel function of the baby, and at the same time, we tell them that we will always be there to help them. We offer them our bowel movement program to be started when the patient is 3 years old in order for the patient to go to school like a normal child with normal underwear and to be adapted and accepted into the society.

When the patient has vesicoureteral reflux, we leave a suprapubic cystostomy tube at the time of the main repair. Prior to the colostomy closure, we perform a suprapubic cystogram to determine the presence and magnitude of the reflux. Also, a urodynamic evaluation will help to determine the best urologic future management.

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