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Reoperations in anorectal malformations

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Cloacal malformation

Abstract

Aim of Study: Significant advances have occurred in the management of anorectal malformations, yet many patients still have technical, frequently catastrophic, operative complications that are potentially avoidable. We chose to analyze our experience in patients who have previously undergone a repair which was unsuccessful and required a reoperation, to detect the technical problems that led to complications and to try to establish a set of recommendations to avoid them.

Methods: From a series of 1806 cases of anorectal malformations, 212 were reoperated on after a failed procedure done at another institution. The operative reports of the original procedure were analyzed, as well as our own operative findings, in an attempt to understand the causes of the complications.

Results: We found 303 indications for reoperation, with many patients reoperated on for more than 1 problem. Complications requiring reoperation included stricture or acquired atresia of the rectum (87 patients), mislocated rectum (76), recurrent, persistent, or acquired fistula from the rectum to a neighboring urogenital structure, or to the perineal skin (67), persistent urogenital sinus in cases of cloacas (23), rectal prolapse (21), stricture or acquired atresia of the vagina (16), stricture or acquired atresia of the urethra (8), and persistent cloaca (4). The analysis of the original operative report and/or our operative findings indicated that the most common causes of these complications were (*a*) insufficient rectal mobilization owing to a dissection performed in a wrong plane, or (*b*) in the presence of or inadequate colostomy located too distally, (*c*) a tense anastomosis owing to inadequate mobilization, (*d*) rectal devascularization caused by rectal wall damage, (*e*) an error in diagnosis because of lack of a distal colostogram, (*f*) incomplete separation of the rectum from the genitourinary tract, (*g*) failed attempts to repair a cloaca with a common channel longer than 3 cm, or those with a very high rectum.

Conclusions: The complications we observed usually had a clear explanation. They can be considered preventable as adherence to specific principles in technique avoids them. Key technical maneuvers are discussed to prevent these complications.

During the last 25 years, significant advances have occurred in the management of anorectal malformations. Despite this, there are still many patients who experience technical, frequently catastrophic, complications that are potentially avoidable. The purpose of this review was to analyze our experience with reoperations for patients born with anorectal malformation who were subjected to attempted but failed repairs and had complications, to analyze the technical problems that led to complications and to try to elaborate a set of recommendations to avoid them.

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Table 1 Indications for reoperations

Stricture/acquired atresia of rectum	87
Rectal mislocation	76
Fistula (recurrent, persistent, or acquired)	67
Persistent urogenital sinus (in cloacas)	23
Rectal prolapse	21
Stricture or acquired atresia of the vagina (in cloacas)	16
Stricture or acquired atresia of the urethra (in cloacas)	8
Persistent cloaca	4
Total[a]	303

[a] Some patients had more than 1 indication.

Since August 10, 1980, the authors have operated on 1806 cases of anorectal malformations. From this, 1420 were primary and 386 were secondary procedures. One hundred seventy-four were operated on secondarily to try to improve bowel control, an experience we have previously reported [1,2]. Two hundred twelve were reoperated on because they had significant complications related to the previous repair done at other institutions. This group of 212 patients represents the material analyzed for this publication.

1. Material and methods

The medical records of 212 patients reoperated on by us were retrospectively reviewed. The ages of the patients varied from 2.75 to 45 years. One hundred fifteen were males and 97 were females. This study was performed following the requirements and with the approval of our hospital's institutional review board.

Special emphasis was placed on the analysis of the operative reports of the original procedure, as well as our own operative findings in an effort to understand the main causes of the complications.

2. Results

We found 303 specific indications for reoperative surgery in these 212 cases. Many patients were reoperated on for more than 1 indication (Table 1). Eight-seven patients had stricture or acquired atresia of the rectum. Seventy-six patients had their rectum completely mislocated outside of the sphincter mechanism. Sixty-eight patients had recurrent, persistent, or acquired fistula from the rectum to a neighboring urogenital structure, or to the perineal skin. "Recurrent fistula" (32 cases) indicates that the fistula was present since birth; the primary surgeons tried to repair it, but the fistula was still present during our reoperation. "Persistent fistula" (19 cases) was one that was present when the baby was born, and when the primary surgeons operated on, they repaired the rectal component of the malformation but were unaware of the presence of the urethral fistula, which remained untouched. An "acquired fistula" (16 cases) was one that was not present at birth, but was inadvertently created during the primary operation.

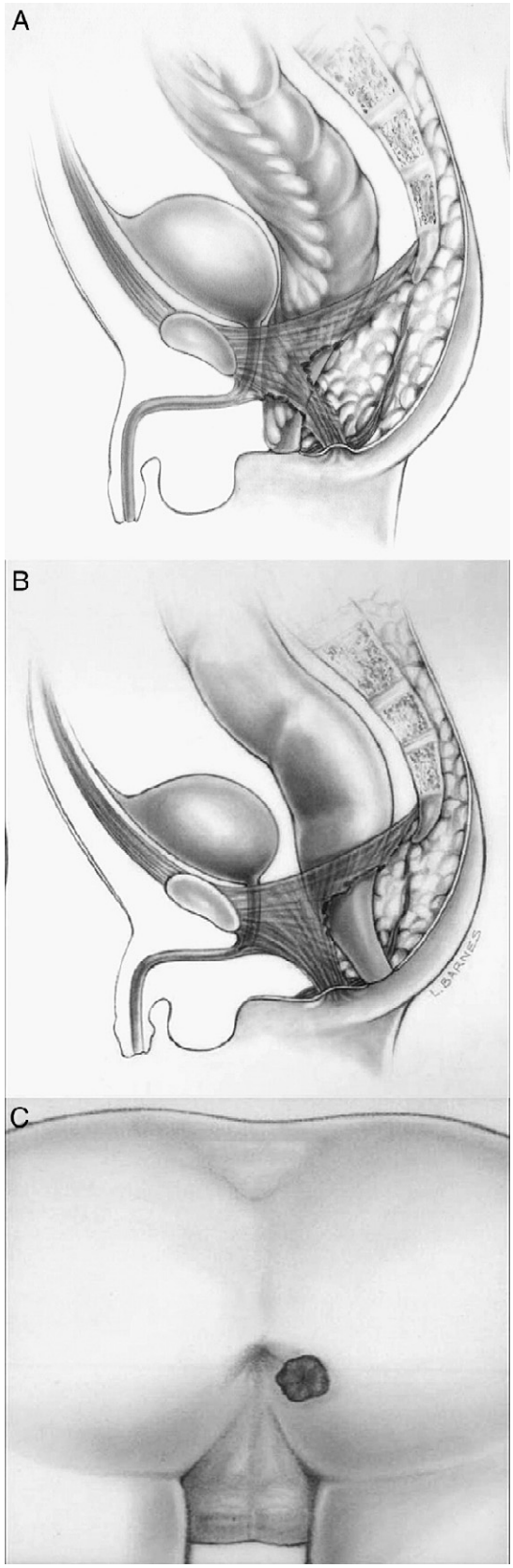

Fig. 1 Rectal mislocation: (A) anterior, (B) posterior, (C) lateral.

Twenty-three patients with cloaca were left with a persistent urogenital sinus when only the rectal component was repaired. Twenty-one patients had severe rectal prolapse larger than 2 cm. Sixteen patients with cloacas had a stricture or acquired atresia of the vagina and 8 had the same problem but of the urethra (Table 1). Four patients had a persistent cloaca.

The operative reports as well as our operative findings of the patients who had stricture and/or acquired atresia of the rectum consistently demonstrated the presence of an untouched perirectal fascia that normally surrounds the rectum of patients with anorectal malformations. Most likely the surgeons were trying to dissect and mobilize the rectum in a plane outside of that fascia; they performed a limited dissection that most likely resulted in tension at the anastomotic site.

The most common type of mislocation seen in this series was an anterior one (53 cases), followed by a posterior mislocation (16 cases). Seven patients had a lateral mislocation of the rectum (Fig. 1).

The most common type of fistula was the recurrent one (32 cases). It was rectourethral in 16 cases, vaginal in 12, vestibular in 3, and perineal in 1. Analysis of the operative reports of these patients, and/or our operative findings, indicated that the rectum was not fully mobilized after the fistula was closed. In other words, the surgeons separated the rectum from the urogenital tract, closed the fistula, but did not mobilize the rectum enough to leave a normal rectal wall in front of the repaired fistula's suture line. In some cases, a damaged anterior rectal wall was left in front of the suture line of the repaired urogenital structure.

Fig. 2 Inadequate pressure during a distal colostogram giving the false impression of a high rectum with no rectourethral fistula.

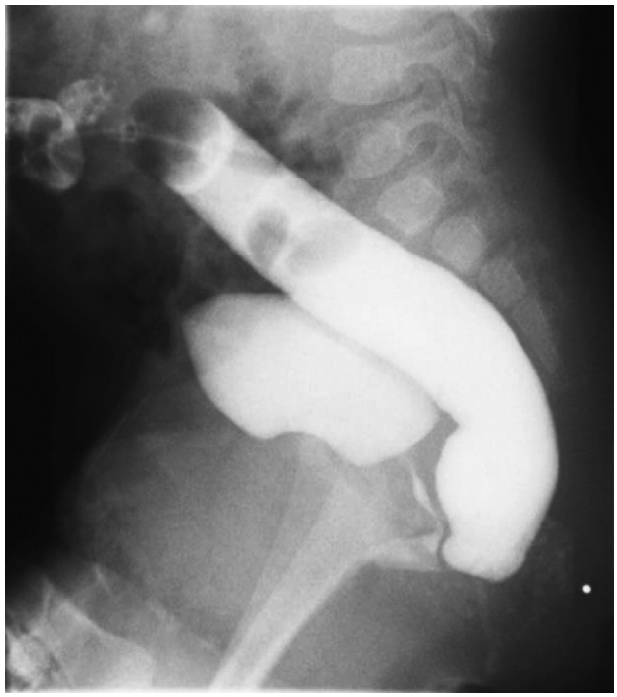

Fig. 3 Distal colostogram of the same patient, performed with enough hydrostatic pressure showing the rectourethral fistula.

The second largest group of fistulae was the persistent rectourinary fistulas in males (19 cases). In these cases, the surgeons were not aware of the presence of the fistula, which was left untouched. The patients did not have a preoperative high-pressure distal colostogram or they had one, but it was not done correctly. The surgeons incorrectly thought that the patients had no fistula (Figs. 2 and 3). During the operation, they found the rectum, pulled it down, but left the fistula intact.

Acquired fistula (16 cases) included 8 to the vagina, 3 to the perineum, and 3 to the urethra. Rectovaginal fistulae occurred in patients born with rectovestibular fistula. It was

Table 2 Reoperation in cloaca malformations

Indication	Number of cases
Persistent urogenital sinus	23
Stricture or acquired atresia of	
vagina	16
rectum	12
urethra	8
Mislocated rectum	18
Recurrent rectovaginal fistula	7
Persistent cloaca	4
Total[a]	88

[a] Some patients had more than 1 indication.

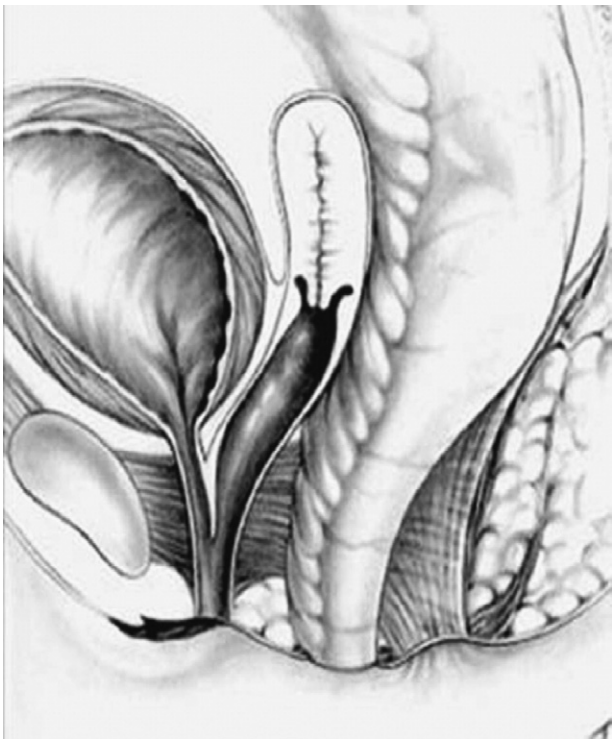

Fig. 4 Persistent urogenital sinus in a patient with a cloacal malformation in whom only the rectal component was repaired.

clear that the surgeons failed to complete the separation of the rectum from the vagina. In addition, they frequently had problems in that separation, making holes in the posterior vaginal and anterior rectal walls. As a consequence, a damaged rectal wall was left in contact with a damaged vaginal wall, which led to an acquired rectovaginal fistula.

The 3 acquired urethral fistula occurred in cases born with a rectoperineal fistula (the most benign of all anorectal defects); these patients had an attempted repair, and the surgeon damaged the posterior urethra creating the fistula.

Twenty-one patients had rectal prolapse larger than 2 cm. When analyzed by the type of the original malformation, it became clear that this problem occurred more often with higher malformations.

Fifty-four of the 212 patients reoperated on by us were born with a cloaca (Table 2). The most common indication for reoperation was a persistent urogenital sinus (23 cases) (Fig. 4). A stricture or acquired atresia of the vagina occurred in 16 patients, the rectum in 12 patients, and urethra in 8. Eighteen patients with cloaca had a rectal mislocation. Seven patients had a recurrent rectovaginal fistula and 4 patients came to us with a persistent cloaca; meaning that, in spite of an attempted repair, the patients were left, with the rectum, vagina, and urethra still confluent. Analysis of the operative reports showed that these patients had a common channel longer than 3 cm; the surgeons unsuccessfully tried to repair the malformation posterior sagittally. The operative reports reflected many difficulties during the mobilization and separation of these structures, which explain the complications seen.

3. Discussion

Our series may not be representative of the frequency with which complications occur in the majority of institutions in the world, as ours is a referral center for the management of these defects, which may explain the high incidence of complicated cases.

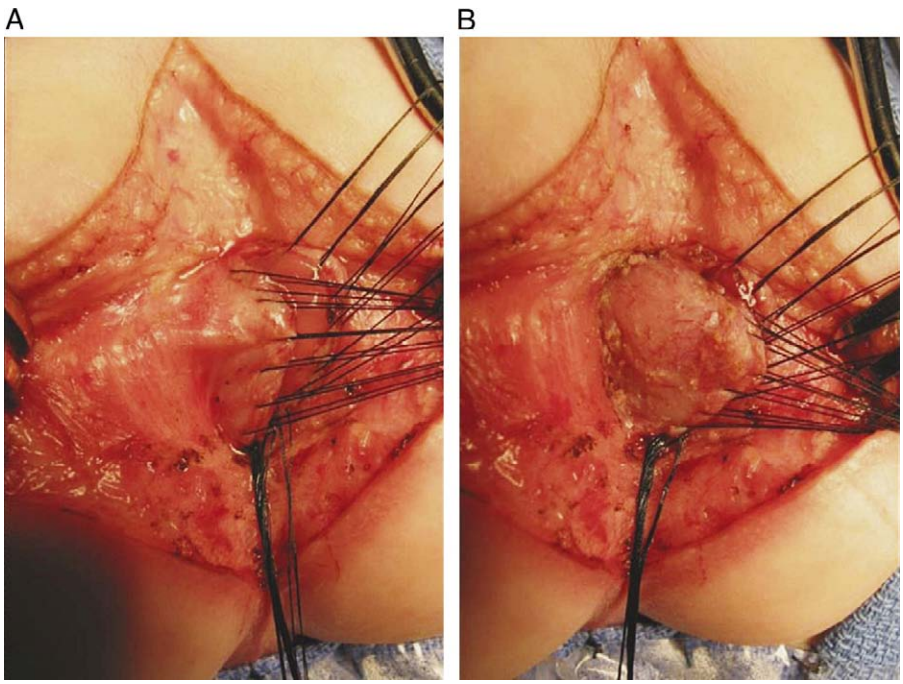

Fig. 5 Rectal wall (A) with fascia; (B) with fascia dissected off.

Stricture and/or acquired atresia of the rectum is, we believe, a preventable complication. In all cases of anorectal malformations operated on primarily by us, we have documented the presence of a white fascia covering the posterior and lateral walls of the rectum (Fig. 5). It is our impression that it is necessary to remove this fascia to carry out the dissection of the rectum, in intimate contact with the rectal wall itself. Even when this maneuver sacrifices much of the extrinsic blood supply of the rectum, the intramural rectal blood supply provides enough perfusion to the most distal end of the rectum, provided the rectal wall itself is not damaged. Trying to mobilize the rectum without removing this fascia (peripheral to the fascia) frequently ends in an unsuccessful mobilization, which results in a tense anastomosis between the rectum and the perineum. In addition, it may be the cause of postoperative neurogenic bladder [3].

Anterior rectal mislocation (Fig. 1) occurred mostly in older patients. Before the popularization of the posterior sagittal approach in 1982 [4,5], the specific recommendation was to pull the rectum as close as possible to the genitourinary structures to preserve the so-called puborectalis sling [6-9]. This recommendation was based on the idea that this muscle would provide bowel control, and it was thought to be the only sphincteric structure available in the so-called "high" anorectal malformations. Below this structure, it was felt that these patients did not have any significant sphincteric mechanism. As a consequence, surgeons tried to create a tunnel as close as possible to the genitourinary structures and did not care much about the final location of the rectum in the perineum. Rectal mislocations are, in our experience, rarely seen today. We believe this is because surgeons are more meticulous in trying to put the rectum in the center of the sphincteric mechanism.

A completely mislocated rectum in a patient with an intact and good sphincter mechanism as well as a normal sacrum represents an indication for reoperation in patients with fecal incontinence. These patients benefit from the relocation of the rectum and may experience improved bowel control [1,2].

Urologic injuries are common [3]. Recurrent fistulas occurred as a consequence of lack of rectal mobilization, the rectum retracted and reconnected to the vagina or urethra. The failure to mobilize the rectum was again a consequence of trying to dissect it outside the fascia, already described. Other times, it was impossible to mobilize the rectum because the patient had a colostomy located too distally in the rectosigmoid tethering the colon to the anterior abdominal wall and interfering with the pull-through (Fig. 6). This is the most common error seen in sigmoid colostomies [10]. The surgeon was not aware of this fact, because he or she was operating on the patient without a high-pressure distal colostogram, or with a technically deficient one [11].

Patients with persistent fistula (untouched) were originally evaluated when they were newborns, using the traditional invertogram. The surgeons found that the image of rectal gas was located very close to the perineal skin; they did not know that these patients had a fistula because a high-pressure distal colostogram was not done. Based on old diagnostic criteria [12], the patients were then operated on through the perineum; the distal rectum was found and pulled down leaving a persistent fistula untouched (Fig. 7). The term *high pressure* was intentionally created to emphasize the need to generate enough hydrostatic pressure during the injection of contrast material to demonstrate a fistula, which is usually tiny. That explains perhaps the high incidence of imperforate anus with "no fistula" shown in the early literature [13]. In our series, imperforate anus with no fistula represents only 5% of our cases [14]. A colostogram done with lack of hydrostatic pressure understandably fails to demonstrate a fistula, and then the surgeons do not look for it.

Acquired rectourethral fistula occurred in male babies born with rectoperineal fistulas and subjected to an anoplasty during which the urethra was injured. These patients did not have an intraoperative Foley catheter. To repair this defect, most surgeons, including us, place multiple stitches in the fistula site to exert traction to perform a circumferential dissection of the fistula and to comfortably relocate the anal opening within the limits of the sphincter. This traction on the rectum of a baby with no Foley catheter conceivably may pull on the urethra that may look like a simple thickening of the anterior rectal wall. Unless the surgeon suspects the presence of the urethra, this can be inadvertently injured or totally divided (Fig. 8) [3].

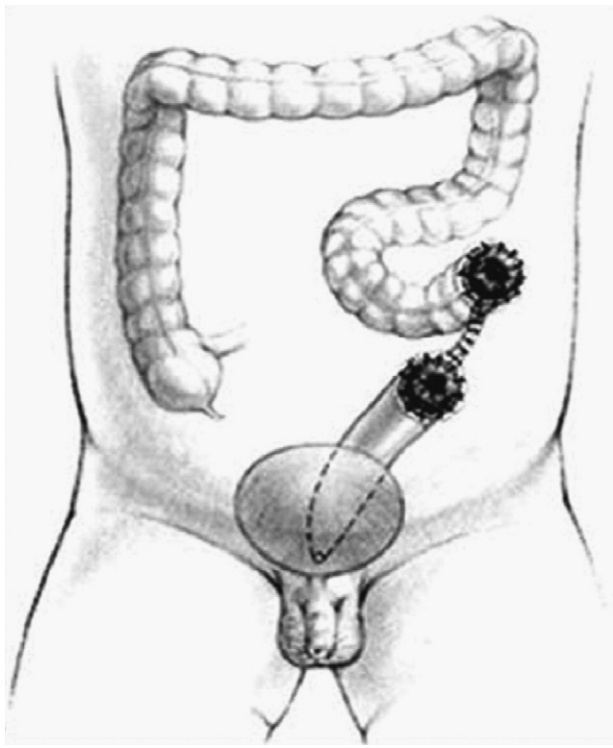

Fig. 6 Colostomy placed too distal in the sigmoid which can interfere with the pull-through.

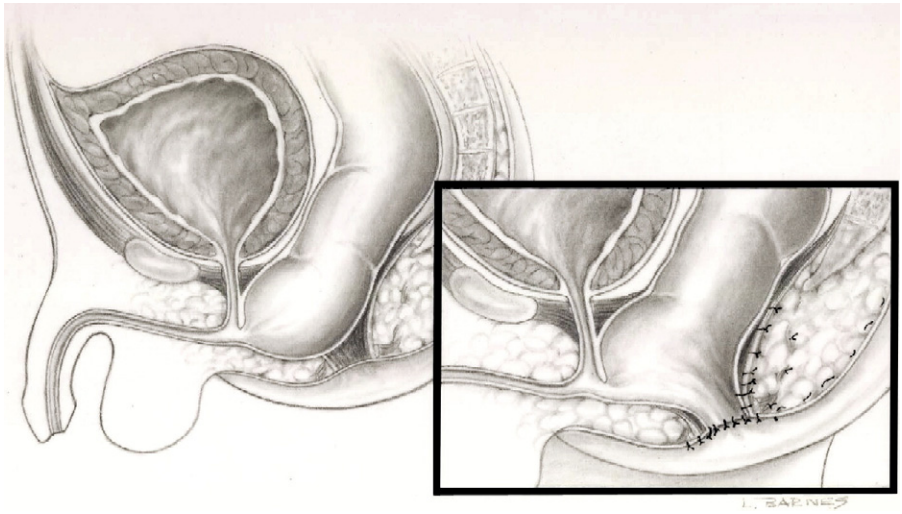

Fig. 7 Persistent rectourethral fistula.

Our recommendation is always to place a Foley catheter. Active bleeding from a sponge-like tissue (spongiosum surrounding the urethra) means that the dissection is getting dangerously close to the urethra.

The problem of a posterior urethral diverticulum was described by us previously [3]. We are concerned that this may become common again with the advent of the laparoscopic approach. There is potential for leaving behind the distal portion of the bowel attached to the urethra, leading to a posterior urethral diverticulum, particularly for malformations, such as a rectobulbar fistula [3].

Rectal prolapse occurs mostly in cases with poor sphincters. To avoid this complication, it is important to tack the posterior rectal wall to the posterior edge of the muscle complex as well as to perform the anoplasty under slight tension [15].

In cases of cloacas with persistent urogenital sinus [16], the surgeons did not know that the patients had cloaca because they referred to a "rectovaginal fistula" and never mentioned the word cloaca. The real incidence of a rectovaginal fistula is extremely low [16]. Many of the "rectovaginal fistulas" reported in the literature in the early years [13], we believe, were actually cloacas that were not correctly diagnosed. These unfortunate patients missed the ideal opportunity to have a total repair of their cloaca and required a complete secondary procedure, which is technically more demanding.

Stricture, or acquired atresia, of the vagina, rectum, or urethra that occurred in patients with cloaca happened in those with a common channel longer than 3 cm. We believe that patients with a short common channel (<3 cm) can be repaired by most pediatric surgeons, provided they are adequately trained to perform a maneuver called total urogenital mobilization [17], which facilitates the mobilization of the urethra and vagina together, preserving an excellent blood supply. Cloacas with a common channel longer than 3 cm represent a serious technical challenge, and we believe that those patients should be referred to centers where there are pediatric surgeons and/or pediatric urologists dedicated to the repair of these complex malformations.

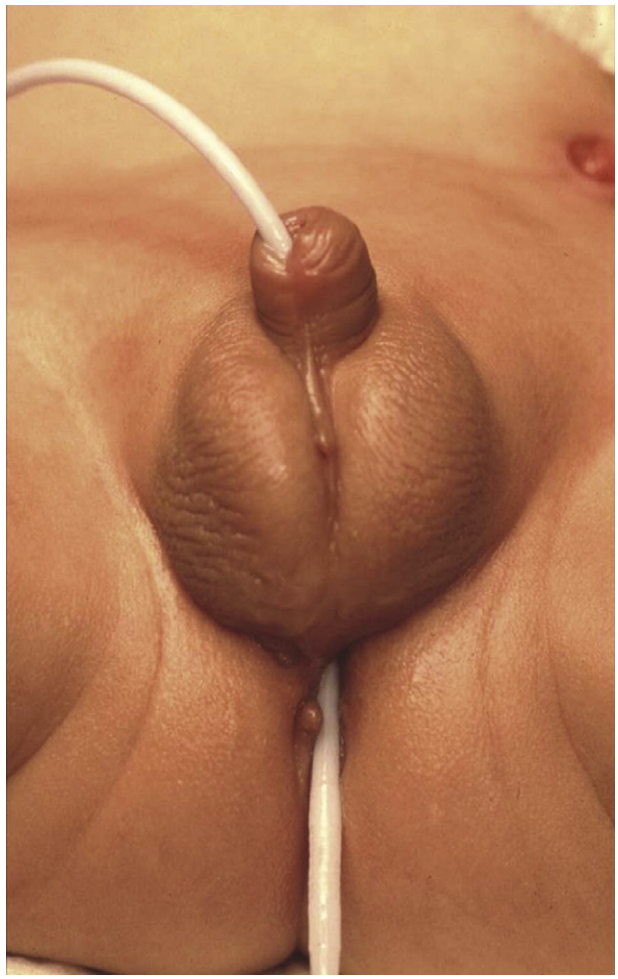

Fig. 8 Acquired rectourethral fistula in a patient with a rectoperineal fistula.

In conclusion, we believe that most complications observed by us are preventable. Rectal dissection must be performed as close as possible to the rectal wall. The rectum must be mobilized enough to achieve an anastomosis without tension and to leave a normal rectal wall in front of the fistula suture. High-pressure colostogram is the most valuable preoperative study. Perineal fistula in males must be repaired with a Foley catheter in place. Cloacas are more common than previously thought, whereas rectovaginal fistulas are extremely rare.

4. Q&A

Grosfeld, Indiannapolis, USA

Alberto that is a marvelous series and certainly gives us a lot of information regarding how to prevent these from happening in the future. What you did not mention in your study and I would like to ask you whether some of these complications occurred in the presence of vesicle fistulas where the surgeon initially pursued the operation from below and did not go into the abdomen. Did he disturb things injure things by a procedural attempt from a place where he could not really reach the problem?

A.

Many surgeons try to repair an anorectal malformation from a posterior sagittal approach (from below as you mentioned) without the right anatomic information, without a good distal colostogram. They do not know that the patient has a very high malformation and yet, they insist in looking for the rectum but instead they find the urethra, seminal vesicles, vas deferens, prostate and cause a lot of damage. When we looked into preoperative notes we found that the patient did not have a preoperative distal colostogram or, had a technically deficient one. That is why we put so much emphasis on the importance of this radiologic study.

Cohen, Sydney, Australia

I would be interested in your opinion as to whether you feel that minimally invasive surgery applied to this operation will reduce the number of complications you have outlined or whether this will just present us with a new range of complications by applying a different technology to your operation.

A.

The laparoscopic approach is indicated to replace a laparotomy. In our experience, only 10% of these patients need a laparotomy. In that 10% group we believe that laparoscopy is indicated. We do not believe that laparoscopy is necessary, justified, or beneficial in cases of rectourethral bulbar fistulae. There is controversy about the use of laparoscopy in rectoprostatic fistulae. If a young surgeon is well trained in laparoscopic procedures and does not have much experience in repairing high prostatic fistula posterior sagittaly perhaps it is not a bad idea for him to try to repair that type of defect laparoscopically. I am not sure about that. I do not think that the complications presented here are going to be necessarily worse by using laparoscopic techniques. We have seen a couple of patients operated laparoscopically at other institutions, who came to us to repair a recurrent rectourethral fistula. We can not blame the laparoscopy for those complications. Laparoscopy or no laparoscopy, the surgeon must know the anatomy of the defect and the patient must always have a technically correct, high pressure distal colostogram.

Sakalkale, Hamilton, New Zealand

Would you consider magnetic resonance emerging as the primary method of investigation for localisation of the fistula and anatomy?

A.

We do not use MRI scans in babies. I do not think that MRIs are good enough to determine the precise location of the fistula. We use MRI in older the patients for other determinations, such as location of the rectum and in cases of posterior urethral diverticulum, also to determine a pre-sacral mass, it is very good. We do not think that to determine the precise location of a fistula that MRI is the right method. In addition, to do a MRI scan in a newborn baby it is a little complicated perhaps in your institution is different. If you can do the MRI and different. If you can do the MRI and you work together with a reliable radiologist, perhaps you can use that study; but I suggest confirming the location of the fistula cystoscopically at the time of the repair.

Kim, Toronto, Canada

Following up on Dr Grosfeld's point. Over 200 out of 1800 cases were re-dos, if you will. Do you have any more information about the surgeon side rather than patient-side in view of the things we have heard about inflammatory bowel disease and you alluded to the fact that things are a little more complex than realised at the original centre. Should some of these cases then be stratified according to the surgeon's experience, perhaps it is a little controversial but in view of your vast experience I would like to ask this question.

A.

We don't discuss surgeons, because if you discuss surgeons you may get into big trouble. It is not an easy issue and it is not our role. For issue and it is not our role. For obvious reasons we get a lot of complicated cases and subsequently we are invited to participate in medico-legal cases. We refuse to participate because we do not want to be seen like a threat to other surgeons but rather as colleagues willing to help. I believe that all of us, senior surgeons, have a serious responsibility to train young surgeons well, to avoid this kind of complication, that we believe are preventable. Of course, we have personal impressions about some surgeons mainly because we receive repeated complicated cases operated by them. We do not know what to do about that, other than waiting for a new generation of better trained surgeons.

Bagolan, Rome, Italy

In female newborn patients with an anorectal anomaly and cloaca. Do you advise a primary vaginal reconstruction or a delayed one?

A.

We replace the vagina when indicated at the same time that we repair the cloaca, since we are there, at the right location. To replace the vagina later in life, will represent the need of a reoperation in a second area. We are not sure yet, about the functional results (sexual activity) of our patients. We are looking forward to find out when our patients reach the age of sexual activity.

Acknowledgments

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