

13.1 Introduction

The minimally invasive approach to the repair of congenital malformation is here to stay. All of us, pediatric surgeons, are concerned about inflicting pain and/or being too invasive to our patients. Every effort aimed to reduce the suffering of our patients, as well as the length of stay in the hospital and subsequently the cost of the operations, is welcomed. We embrace ideas that have, as the end result, a less traumatic operation without compromising the standard of care or provoking more sequelae in our patients. That explains why the minimally invasive approach to repair malformations had so much impact in pediatric surgery and the surgical field in general.

Since the introduction of this new technology, there are now operations in which the laparoscopic or thoracoscopic approach represents the gold standard. There are others in which these new approaches have not been applied, and there is another group in which the approach is controversial. Anorectal malformations have not been an exception, and many pediatric surgeons have used and continue trying to use more and more the laparoscopic approach to the repair of anorectal malformations, many of them with the correct indication, others in malformations that can be

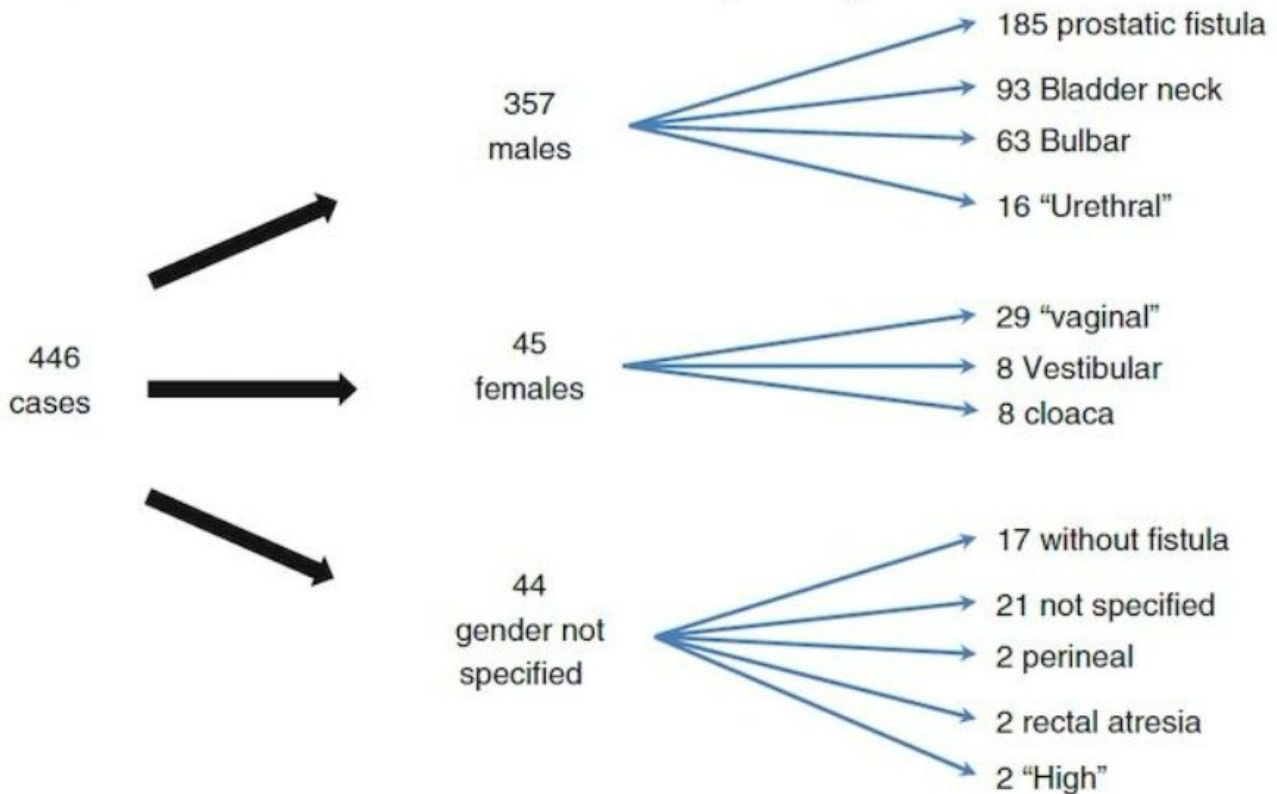
repaired either way laparoscopically or posterior sagittally, and others in cases in whom we believe that laparoscopy is formally contraindicated.

Our basic contention and belief is that laparoscopy is primarily indicated to replace a laparotomy. In other words, a classic indication is a procedure that is usually done opening the abdomen and now can be done equally well, through the small orifices of the ports necessary for minimally invasive operation. In male patients with anorectal malformations, we have to open the abdomen 10 % of the times, specifically in those cases that have a recto-bladder neck fistula. We consider that group a good potential indication for laparoscopy. We use the word “potential” because even in those particular types of defects, laparoscopy has certain limitations, as will be shown later.

Approximately 30 % of patients with cloacas require a laparotomy. Yet, all of them require a posterior approach in order to repair the urogenital component of the malformation. Conceivably, a laparoscopic approach could be useful to separate the rectum from the bladder neck in those unusual cases that have the rectum connected to the bladder neck. Yet, those patients require rather sophisticated and technically demanding maneuvers to repair the urogenital component of the malformation (see Chap. 16).

We decided to review the literature related with the laparoscopic approach of anorectal malformations. We found 52 papers published between 1998 and 2014. Forty-one of them [1–41] presented

Electronic supplementary material Supplementary material is available in the online version of this chapter at [10.1007/978-3-319-14989-9_13](https://doi.org/10.1007/978-3-319-14989-9_13).

Table 13.2 Summarized results of a literature review on minimally invasive approach to anorectal malformation

series of cases (Table 13.1). Ten papers [42–52] were rather informative, written by experts but without discussing specific cases.

The total number of cases reported in the literature according to our review is 446. These included 357 males, 45 females, and 44 in whom the gender was not specified (Table 13.2).

The group of male cases included 185 recto-prostatic fistulas, 93 bladder neck, 63 bulbar, and 16 labeled as “urethral” without specifying the location of the fistulas.

The female group included 29 “vaginal fistula,” 8 vestibular, and 8 cloacas. The group of patients without gender specification included 21 cases, 17 without a fistula, 2 recto-perineal, 2 “rectal atresia,” and 2 cases labeled simply as “high” (Table 13.2).

13.2 Males

Anorectal malformations in males are divided into the following categories:

- Recto-bladder neck fistula
- Rectourethral prostatic fistula

- Rectourethral bulbar fistula
- Anorectal malformation with no fistula
- Perineal fistula

We believe that mainly those patients with recto-bladder neck fistulas may benefit from the laparoscopic approach. In our own experience, the laparoscopic approach provides an excellent exposure of the peritoneal floor and the distal part of the bowel as it approached the bladder neck (Animation 13.1). The dissection of the rectum, as well as the ligation of the fistula, is an easy maneuver. The main reason for this is that in anorectal malformations, the higher the malformation, the shorter the common wall existing between the rectum and the urinary tract, and therefore the rectum ends in the bladder neck in a T fashion, which makes the ligation of the fistula an easy task, without any risk of damaging neighboring structures. To repair this malformation, the next step after the ligation of the fistula is the mobilization of the rectum. We found that sometimes this is feasible, and the entire procedure can be completed after making a small incision in the perineum. Yet, sometimes the mobilization of the rectum (as explained in Chap. 11) requires a

delicate and selective ligation of mesenteric vessels, preserving the necessary arcades in order to preserve the blood supply of the distal rectum. Laparoscopically, this maneuver is done using cautery, which is a less than optimal way to divide vessels, because the extension of the burn is not controlled accurately and may damage collateral vessels necessary to preserve the blood supply of the distal rectum. In addition, some of these patients have a very dilated rectum that requires tapering. We found difficult to perform the tapering of the bowel laparoscopically, and therefore we have to open the abdomen. It is important to remember that unfortunately, these groups of patients (recto-bladder neck fistula) have a very poor functional prognosis. Only 20–25 % of our patients in this category have voluntary bowel movements, and about half of those suffer from soiling. These patients are born with serious congenital deficiencies in their sphincteric mechanism and frequently are associated to very poor sacra.

Ninety-three cases of our literature review belong to this category of recto-bladder neck fistula. We believe that all pediatric surgeons agree that this malformation represents an excellent indication for a laparoscopic approach.

Perineal fistulas are malformations that can be repaired with an operation that takes less than 45 min with 100 % chance of bowel control. Therefore, we do not see the reason to attempt a less invasive procedure than the one that we do at present time. Yet, we found two publications that included perineal fistula repaired laparoscopically [2, 12].

Imperforate anus with no fistula is a malformation in which the rectum is blind; it has no communication with the urinary tract. Half of these patients have Down syndrome. Our experience shows that this is a benign condition if we consider the good functional prognosis. Ninety percent of our patients without Down syndrome have bowel control, and 70 % of our Down syndrome patients also have voluntary bowel movements.

We found that 17 patients (from our literature review) with this defect underwent a laparoscopic repair [2, 7, 10, 17, 22, 23, 25, 34, 37]. We believe

that patients with anorectal malformation without fistula should not be approached laparoscopically. The rectum is located very close to the perineum, and therefore it requires a minimal dissection to be mobilized. A laparoscopic approach implies in these cases a full dissection of the rectum, which is actually unnecessary. In our series, the operation takes 2 h, the patients have minimal pain, the blood loss is negligible, and the patient can be discharged 24 or 48 h after the operation.

Rectourethral bulbar fistula is fortunately the most common defect that we see in male patients. The rectum is located approximately 2 cm above the skin and connects to the bulbar urethra at the same level of those cases of imperforate anus with no fistula. Eighty-five percent of our patients with normal sacrum have bowel control, and 94 % of them have urinary control. The operation takes approximately 2½ h, and the patients eat the same day, have minimal pain, and can be discharged the next day. We find it very difficult to have a less invasive procedure than the one that we perform. Again, trying to repair these malformations laparoscopically represents a maximally invasive approach since the entire rectum has to be dissected from above. When these malformations are repaired posterior sagittally, we only dissect the distal part of the rectum as much as necessary to bring the rectum down; we do not have to go all the way to the peritoneum. As a consequence of attempting the repair of this malformation laparoscopically, a significant number of patients suffered from recurrent rectourethral fistula or a large persistent posterior urethral diverticulum which represents a piece of rectum left attached to the urethra [53, 54] (Animation 13.2) (Fig. 13.1). We believe that laparoscopically the surgeons found it impossible to reach the bulbar fistula site and amputated the rectum leaving a piece of rectum that later gave significant symptoms. We believe that the laparoscopic approach of this malformation is contraindicated. Unfortunately, many surgeons keep trying [5, 6, 11–13, 17, 24–26, 30, 34, 35, 37–39].

Rectourethral prostatic fistula is the second most common malformation that we have seen in male patients. For us, this malformation

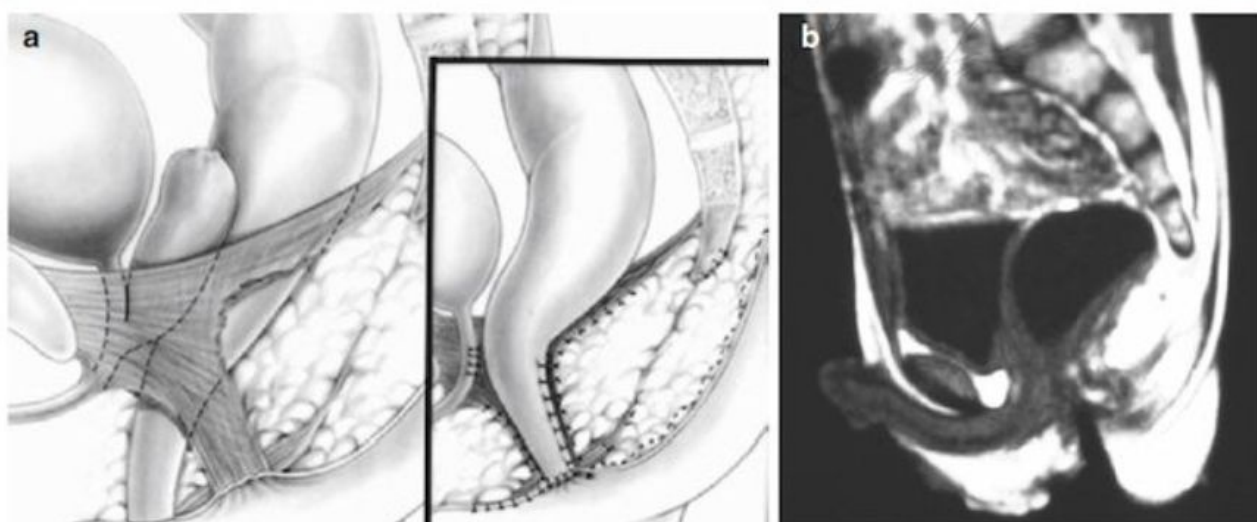


Fig. 13.1 Posterior urethral diverticulum. (a) Diagram. (b) MRI study showing a giant posterior urethral diverticulum

represents a source of controversy concerning the best way to approach it. The posterior sagittal approach in these malformations renders a 66 % chance of bowel control by the age of three. Ninety-two percent of the patients have urinary control. In most of these patients, the posterior sagittal dissection of the rectum requires to open the peritoneum. In other words, the dissection is a complete one, due to the fact that the rectum is located high in the patient's pelvis. If a surgeon is particularly well trained laparoscopically and prefer to approach this type of malformation laparoscopically, rather than posterior sagittally, we do not see a real difference between going one way or the other. In addition, some of the rectoprostatic fistulas are located a little higher than others closer to the bladder neck; in those cases, we have seen complications when inexperienced surgeons try to repair this malformation via posterior sagittal. Perhaps, if they have good training in laparoscopic techniques, it will be better for them to approach those malformations that way. We were happy to find in our literature review that 185 out of 357 male cases operated laparoscopically had this kind of defect.

Unfortunately, we found in our literature review that many surgeons still use the old terminology that we consider inadequate and misleading. They still refer to "high, intermediate, and low" malformations. Sixteen of the published cases received the nonspecific diagnosis of

"rectourethral fistula" [7, 10, 13, 22]. Operating a male baby with an anorectal malformation without knowing the detailed anatomy represents a potential misadventure with serious consequences for the patient.

13.3 Females

Anorectal malformations in females are divided into the following categories:

- Perineal fistula
- Vestibular fistula
- Cloacas
- Complex malformations

Perineal fistulas in females are repaired with a minimal posterior sagittal operation that takes less than an hour; the patients have minimal pain, 100 % of them have bowel control, and therefore we do not see a justification to do a maximally invasive operation dissecting the entire rectum through the abdomen to repair these malformations. Yet, as previously mentioned, some cases are occasionally done laparoscopically.

Vestibular fistulas are by far the most common defects in female patients with anorectal malformations. In our series, 95 % of these patients with a normal sacrum have bowel control. The operation to repair this defect takes us approximately 2½h. The patients can eat the same day when operated with a colostomy and can be discharged

the next day after surgery. The most important anatomic feature in these patients is the common wall existing between the rectum and the vagina (see Chap. 15). The real challenge in the repair of these malformations is to make two walls out of one, preserving intact the posterior vaginal wall and the anterior rectal wall. It is difficult for us to imagine why somebody wants to dissect the entire rectum through the abdomen, in order to repair these malformations and still claim that it is less invasive [9, 17, 36, 37]. In some of those patients in whom the surgeons attempted a laparoscopic repair, actually, they left a piece of rectum attached to the vestibule and mobilized more colon from above, which is against the basic principles of the repair of anorectal malformations.

Cloacal malformations are the second most common anorectal malformations seen by us in females. The repair of a cloaca requires separation of the rectum from the urinary tract followed by the mobilization and repair of the urethra and vagina. For that, most of the time we use a maneuver called total urogenital mobilization. There are reports of laparoscopic attempted repairs of cloacas [7, 9, 10, 17, 23, 29]. The authors actually only repaired the rectal component of the malformation, and then they performed a posterior sagittal approach from below in order to repair the urogenital component of the malformation. In other cases the surgeons repaired the rectal component and left the urogenital tract intact! In other words, we cannot say that the laparoscopic approach has been used to repair cloacas, but, rather, we should say that it has been used to repair the rectal component of cloacal malformations. Yet, in our experience, in over 500 cloacas, the rectum can be reached posterior sagittally in the majority of cases; therefore, we do not see a reason to try to go through the peritoneal cavity to repair a malformation that can be repaired from below. Again, in theory, laparoscopy could be helpful to mobilize a rectum that can only be mobilized through the abdomen. Yet, the patient still will require a posterior sagittal approach anyway to repair the urogenital sinus.

Laparoscopy can also be useful instead of a laparotomy for the examination of the intra-abdominal anatomy in complex anorectal malformations.

We are obviously biased against the laparoscopic approach of cases with rectourethral bulbar fistula because of the cases that we see at our center, suffering from a complication consecutive to the laparoscopic approach, including urethral injuries and posterior urethral diverticula [53, 54].

We are aware of the fact that these complications are not necessarily a consequence of the laparoscopy, since we have seen similar complications occurring during the repair of anorectal malformations with other techniques. However, these complications occurred mainly in patients suffering from rectourethral bulbar fistulas.

In our literature review, we found that 29 patients were operated laparoscopically with the diagnosis of rectovaginal fistula [2, 3, 6–10, 17, 23, 28, 29, 34–36].

We consider that real vaginal fistula is an almost nonexistent malformation. In our series of over 1,000 female patients, we have seen only seven real rectovaginal fistula cases (see Chaps. 15 and 16). Most of the cases that came to us with the diagnosis of “rectovaginal fistula,” in retrospect, were misdiagnosed cloacas or rectovestibular fistula. If these authors found 29 vaginal fistulas out of 45 female cases, we have reason to suspect that they were not really vaginal fistulas.

Some authors [10, 17, 35] try to evaluate the clinical results using an old score that includes “number of bowel movements” as indicative of results; “many bowel movements” were considered as bad and “few bowel movements” were considered “good”!! The number of bowel movements relates to many factors (diet, length of the colon, inflammatory factors, degree of dilatation of the rectosigmoid); none of them related to the type of repair (laparoscopic or posterior sagittal).

We were negatively impressed by the fact that only eight of the papers that we reviewed mentioned the characteristics of the sacrum of the patients [5, 13, 14, 17, 21, 28, 30]. We think that it is impossible to discuss functional results in the treatment of anorectal malformations without specifying the characteristics of the sacrum of the patients. The presence or absence of tethered

cords was only mentioned by five authors [2, 4, 17, 28, 40]. The presence of this anomaly is recognized as a negative prognostic factor, mainly for the urinary function.

One of the rationales to justify the laparoscopic approach in patients with anorectal malformations is the short length of stay. We were very surprised to find that the length of stay was only mentioned in three papers [22, 35, 41], and the authors reported 5–11 days' range of length of stay. In our series, 90 % of our male cases did not require a laparotomy, and they remained in the hospital 48 h postoperatively. In females, with the exception of complex cloacas that required a laparotomy, the length of stay was similar to the males.

Ten publications [6, 10, 17, 22, 32–35, 37, 41] compared the functional results obtained with laparoscopy and the traditional PSARP (posterior sagittal anorectoplasty). Most of the authors concluded that the differences found were not significant. Others suggested that the results obtained with the use of laparoscopy are better [17, 41].

Four of the papers [6, 10, 22, 35] evaluated their results using rectal manometry, which we consider a non-reliable method (see Chap. 25).

Some authors mentioned the "potential negative effect of the scarring produced by the posterior sagittal approach." That is a reasonable theoretical concern; however, we strongly suggest for those authors and the readers to read our publication [55] describing our experimental experience (in dogs), comparing different types of rectal operations and their effect on bowel control. The posterior sagittal incision did not affect bowel control. In addition, from the clinical point of view [56], we have operated multiple cases with normal rectum that required a complete division of the rectum and anus, including the posterior and anterior sphincter mechanism (what we call trans-anorectal approach), and bowel control was not affected. All patients have normal bowel control.

We have not found conclusive evidence that the laparoscopy is less invasive than the posterior sagittal approach for the repair of anorectal malformations in patients that do not require a

laparotomy. There is no current evidence of better clinical results. There are a significant number of cases in which laparoscopy has been used with no real justification.

We are happy to see that many authors recognize the risk of urethral damage and leaving a posterior urethral diverticulum and are showing a high degree of creativity, using sophisticated technology to avoid these complications [12, 24, 30]. Others are already limiting the use of laparoscopy to cases with bladder neck or prostatic fistula [37].

We strongly suggest to all surgeons interested in performing laparoscopic repair of anorectal malformations to follow the following guidelines:

- Use an adequate terminology; try to be accurate in the type of malformation treated.
- Refer to the characteristics of the sacrum as well as the presence or absence of tethered cord, in order to discuss clinical results.
- Avoid confusing terms such as "high," "intermediate," and "low."
- When discussing cloacal approach laparoscopically, be specific as to "repair of a cloaca" or repair of the "rectal component of the cloaca." Be specific in saying exactly what was done with the urogenital component, which represents the real technical challenge.

We firmly believe that minimally invasive techniques will play an increasing positive role in the management of many congenital anomalies including of course anorectal malformations. However, we must be cautious to avoid negative results as a consequence of excessive enthusiasm or the desire of notoriety.

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