

Two-stage repair for delayed presentation of anorectal malformation with vestibular and perineal fistulae in females

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Background/aim

In spite of advances in knowledge and techniques in the management of anorectal malformation (ARM), delayed presentation of female ARM with vestibular and perineal fistulae is quite common especially in developing countries. In this literature, there is no standardized surgical algorithm for late presentation of these female ARM.

This study aimed to present the experience of two pediatric surgery tertiary centers regarding delayed presentation of vestibular and perineal fistulae using two-stage procedures.

Patients and methods

This prospective study was conducted on 28 female patients with late presentation of vestibular and perineal fistulae from November 2014 to April 2019. Lower contrast study was done for all patients. Laxative, frequent rectal wash enemas, and Hegar's dilators were used for 2–3 weeks preoperatively. All patients were repaired by anterior sagittal anorectoplasty (ASARP) with covering high sigmoid loop colostomy, and colostomy closure was performed after 2–3 months. Krickenbeck continence score was used for assessment of anal continence.

Results

Of 28 patients included, 25 patients completed the study. There were 14 patients with perineal and 11 with vestibular fistulae. The age of patients ranged from 8 months to 18 years. The main reasons for delay were socioeconomic, unawareness, and overlapping causes. Sphincter-saving ASARP was used in 15 patients and the classic ASARP was used in 10 patients. Tapering proctoplasty was needed in one patient. Minor wound infection developed in three patients. Constipation occurred in 11 (44%) patients. Follow-up period ranged from 6 to 50 months (median 30 months). Good continence was obtained.

Conclusion

Although delayed presentation of vestibular and perineal fistulae in female patients is accompanied by comorbidities, with good surgical management, successful results can be obtained by using two-stage procedure.

Keywords:

anal continence, anorectal malformation, anterior sagittal anorectoplasty, constipation, delayed presentation, vestibular fistula

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Introduction

Anorectal malformations (ARM) are congenital correctable anomalies with good outcome in most cases [1]. In female patients, imperforate anus with rectovestibular fistula, rectoperineal fistulae, and cloacal anomalies are the most common ARM [2,3].

Despite advances in knowledge and techniques in surgical correction of ARM, in developing countries, delayed presentation of vestibular and perineal fistulae in female patients is still quite common [4].

In this literature, there is controversy in the lines of surgical treatment of late presentation of ARM with vestibular or perineal fistula in female patients. The purpose of the present study was to present the outcome of two tertiary pediatric surgery centers in

the surgical management of delayed presentation of vestibular and perineal fistulae in female patients older than 6 months using two-stage procedure.

Patients and methods

This prospective study included female patients with late presentation of vestibular and perineal fistulae. The study was conducted in the period from November 2014 to April 2019 in two referral pediatric surgery tertiary centers in Upper Egypt.

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The study was carried out after the Local Ethical Committee reviewed and approved the study protocol in both institutions. The parents of all patients signed a detailed written informed consent form before enrollment in the study. The following data were collected: the age at presentation, cause of delay, associated anomalies, previous attempts of repair, rectal dilatation on contrast study, procedure selected for surgical correction, operative and postoperative complications, continence assessment, and follow-up period data for at least 6 months.

Design of the study

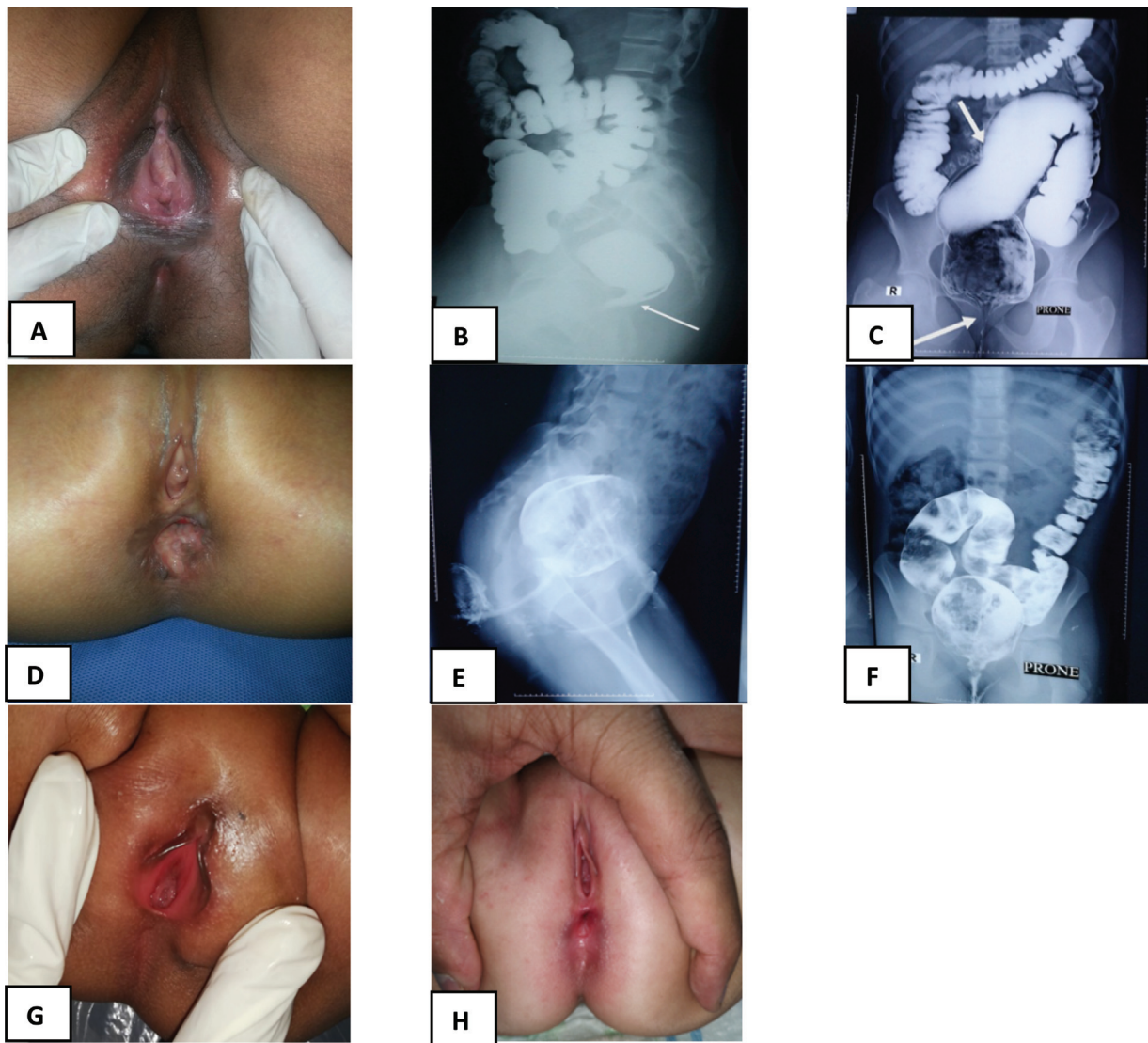
Inclusion criteria were female patients with ARM with vestibular and perineal fistulae who first presented after the age of 6 months (Fig. 1).

Exclusion criteria were patients who presented with colostomy.

The included patients were subjected to the following

- (1) Thorough history taking and examination to detect the cause of delay and associated anomalies.
- (2) Contrast enema to measure fistula length and the degree of rectal dilatation (Fig. 1).
- (3) Preoperative preparation with frequent rectal wash enemas and Hegar dilatation for all patients 2–3 weeks before surgery to decrease rectal dilatation.
- (4) All patients were repaired by anterior sagittal anorectoplasty (ASARP) with covering high sigmoid loop colostomy, and colostomy closure was done after 2–3 months later.

Figure 1



Different patients with delayed presentation (a) an 18-year-old patient with vestibular fistula; (b, c) contrast enema show the fistula and rectal dilatation; (d) perineal fistula in a 8-year-old patient with history of previous unsuccessful surgery; (e, f) contrast enema of the same patient showing rectal dilatation; (g) a 15-month-old patient with vestibular fistula; (h) perineal fistula in a 12-month-old patient.

- (5) During the follow-up period, cooperative children were advised to perform pelvic floor exercise.
- (6) Anal continence assessment was done using Krickenbeck continence score [5] 1 year after surgery and/or patient's age older than 3 years (Table 1).

Operative technique

All patients were repaired either by classic ASARP as described by Okada *et al.* [6] in patients with rectal dilatation or sphincter-saving ASARP in patients without rectal dilatation (in which the mobilized rectum pass through the center of intact sphincter complex).

Operative steps

All patients were operated under general anesthesia with caudal analgesia.

Regarding position of the patients, lithotomy position (or supine position with hanging and supporting the legs) was used, with insertion of urethral catheter.

Using muscle stimulator (Peña stimulator) or low-current diathermy (needle diathermy cutting mode), the position of the neoanus was marked at the center of the sphincter muscle complex, traction sutures were applied around the fistulous opening with circumferential incision, and strict midline perineal incision was done for careful dissection of perineal muscle. Meticulous dissection was needed for separation of the rectum from posterior vaginal wall, especially in these late cases owing to presence of fibrosis and adhesions.

Then mobilization of the rectum to reach the perineum without tension was done. This was followed by passing the rectum in the center of muscle complex and finally

perineal body reconstruction and anoplasty. In vestibular fistula, more meticulous dissection was needed for separation of the rectum from posterior vaginal wall.

In sphincter-saving technique, the center of the intact sphincter complex was identified using Peña stimulator and then with gradual dilatation by Hegar's dilator without its incision or laceration. The mobilized rectum then was passed through the center of sphincter complex, and finally, anoplasty and perineal body reconstruction was done [7,8] (Fig. 2).

Loop colostomy at the junction between the descending and sigmoid colon was done at the end of surgery.

Oral feeding was started on the second postoperative day, and urethral catheter was removed after 5 days. Discharge from the hospital was done on the sixth day after training the parent on how to deal with both stoma and perineal wound.

Anal dilatation was started after complete wound healing. Colostomy closure was done after 6–8 weeks after complete healing of the perineal wound.

Results

Of 28 patients included in the study, one patient (aged 20 years with vestibular fistula) refused surgery and two girls with vestibular fistula presented with colostomy (done elsewhere), so they were excluded from this study. A total of 25 patients fulfilled the inclusion criteria. There were 14 patients with perineal and 11 patients with vestibular fistulae.

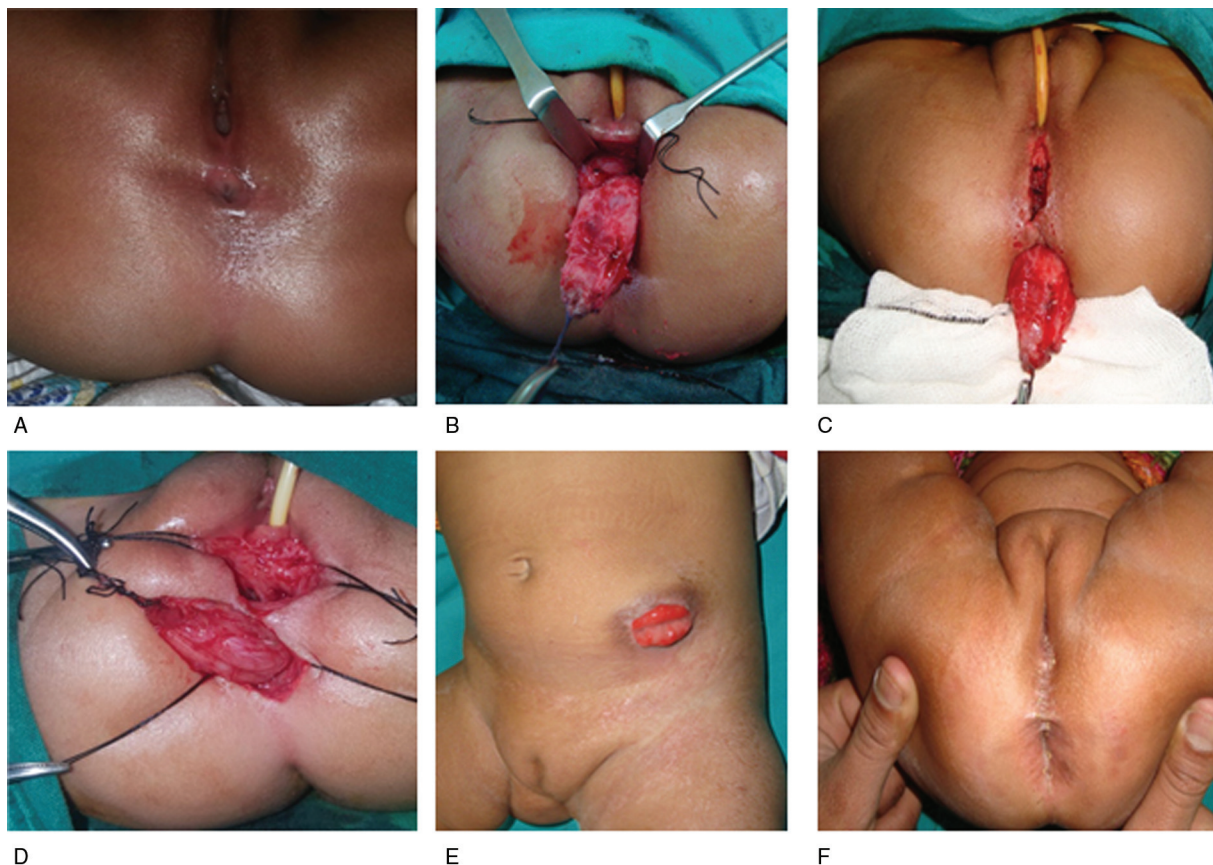
By reviewing the medical record in both centers, it was found that 28 patients included in this study presented

Table 1 Krickenbeck continence score [5] of postoperative results^a

Items	Score ^a	Number of patients (n=18)
1. Voluntary bowel movements feeling of urge, capacity to verbalize, hold the bowel movement (yes/no)	Yes 1 No 0	18 0
2. Soiling (yes/no)	No 3 Yes	8 10
Grade 1: occasionally (once or twice per week)	2	6
Grade 2: every day, no social problem	1	3
Grade 3: constant, social problem	0	1
3. Constipation (yes/no)	No 3 Yes	7 11
Grade 1: manageable by changes in diet	2	8
Grade 2: require laxative	1	3
Grade 3: resistant to laxatives and diet	0	0

^aTotal score: seven points.

Figure 2



Classic and sphincter-saving ASARP technique. (a) Patient aged 11 months with perineal fistula; (b) rectal mobilization and separation from posterior vaginal wall; (c) ASARP before anoplasty; (d) sphincter-saving ASARP in another patient with the rectum passed through the intact sphincter; (e) the colostomy before closure with no related colostomy complications except for mild pigmentation; (f) the perineum after complete healing. ASARP, anterior sagittal anorectoplasty.

with late presentation among 200 female patients with ARM who presented to both centers during the study period. In the first year of this study, of 44 cases of female ARM, 11 (25%) cases of them were late presentations. The presentation of these late cases gradually decreased and now has become one to two cases per year.

As shown in Table 2, the age of studied patients ranged from 8 months to 18 years (median 3 years) at presentation. Five patients presented initially by fecal impaction with accidental discovery of perineal fistula. Causes of delay are summarized in Table 2, which shows the associated anomalies and causes of delayed presentations. Laxatives, rectal wash enemas, and Hegar’s dilators were done 2–3 weeks before surgery, which was successful for improvement of rectal dilatation in all except one patient. Sphincter-saving ASARP was used in 15 patients without rectal dilatation, and the classic ASARP was used in 10 patients with rectal dilatation, as shown in Fig. 3. Tapering proctoplasty was needed in one patient owing to dilated rectum, and this patient developed

Table 2 Clinical data of patients

Clinical data	N (%)
Age at presentation	
8 months to 3 years	14 (56)
3–10 years	7 (28)
10–18 years	4 (16)
(Median 3 years)	
Presentation	
Perineal fistula	14 (56)
Vestibular fistula	11 (44)
Associated anomalies	
Congenital heart disease	3 (12)
Tethered cord	1 (4)
Causes of delayed presentation	
Social and economic	5 (20)
Previous bad experience	3 (12)*
Overlapping causes	9 (36)
Congenital heart disease	3 (12)
Unawareness	5 (20)#

*Two of them were sisters. #All of them was perineal fistulae.

superficial wound infection. There was no reported rectal injury, and vaginal injury occurred in two patients with immediate intraoperative repair. Operative time

Figure 3



Preoperative, early and long-term follow-up in patients aged 6 years. (a) Perineal fistula with cicatrization due recurrent inflammation; (b,c) contrast study through the fistula to assess rectal dilatation; and (d) 3 months after anterior sagittal anorectoplasty with mucosal prolapse on right side; (e) the same patient before colostomy closure with skin excoriation around the stoma; (f,g) two-year follow-up of the same patient after closure of colostomy.

ranged from 2.5 to 4 hr, with a median of 3.25 h. Hospital stay ranged from 4 to 6 days (median 5 days). Superficial wound infection occurred in three patients with vestibular fistula (one of them tapering proctoplasty was done), with improvement with local wound care. Anal stenosis developed in three patients (two vestibular and one perineal): two patients improved with regular anal dilatation, and only one patient (with perineal fistula) needed anoplasty. One

patient needed mucosal excision before colostomy closure. Colostomy closure was done 2–3 months after the first stage. The only complication related to the colostomy in this study was skin excoriation especially in older patients (Fig. 3). Only one patient needed blood transfusion in whom tapering proctoplasty was needed. There was no mortality reported in this study. Continence was assessed by Krickenbeck continence score [5] done in 18 (72%)

patients. The result of postoperative continence is shown in Table 1. Soiling occurred in 10 (55.5%); fortunately, those patients showed improvement with time during the follow-up period. Constipation occurred in 11 (44%) patients and was controlled by diet, but three patients needed laxatives. All patients maintained regular follow-up by outpatient visits or phone calling. The follow-up ranged from 6 to 50 months (median 36 months). The overall outcomes of the studied patients were satisfactory.

Discussion

In female patients, imperforate anus with rectovestibular fistula, rectoperineal fistula, and cloacal anomalies are the most common ARM [2,3]. Although ARM can be easily detected in the neonatal period by proper clinical examination after resuscitation, there is still delayed diagnosis. This not only occurred in low-resource countries but also in developed one but with low percentage [9,10]. This delay is usually associated with more morbidity than early presentation [11,12].

In this literature, delayed presentation is defined as 'patient who presented after 7 days of birth, except for a female patient, with low-type ARM, where presentation beyond 6 months of age was considered as delayed' [1]. However, other authors considered presentation more than 48 h after birth as delayed diagnosis [12]. The higher incidence of delayed presentation of ARM in female than males may be owing to colostomy, which was done as an emergency procedure in neonatal period for males with high and intermediate ARM, with the need for definitive treatment thereafter. Poverty, inadequate knowledge or experience about female ARM, and lack of advanced health care are other causes [13].

In this study, the incidence of delayed presentation was 25% in the start of this study, but now, it has decreased owing to increased awareness in the community. According the result of this study, the reasons of late presentation were unawareness and accidental discovery of abnormal anal opening. This is the main cause of delayed presentation in perineal fistula. The main concern of parents came for consultation was the abnormal anal opening and its effect on marriage, especially those presented at adolescence age. The main problem of interest was the affection of abnormal anal opening on the future marriage. Unfortunately, one patient refused surgery assuming that she is continent and surgical correction may cause problems with her current marriage, as she was already engaged.

In a study by Rawat *et al.* [14], illiteracy, wrong advice, and poverty were the main causes of late presentation. They added that their patients came from villages with low social and economic standard. In another study, Sinha *et al.* [1] stated that 'the reasons for a delayed presentation were either delayed awareness on the part of parents or poor access to the appropriate facilities.'

Major congenital anomalies sometimes may be the cause of delay in the current study. A total of three (12%) patients had congenital heart disease that needed correction before definitive surgery for low ARM. Mittal *et al.* [15] reported that associated congenital anomalies were found only in one patient (10%).

In this literature, female ARM with vestibular and perineal fistula according to Krickbeck classification are corrected either by one or three stages [5]. Many authors prefer correction of early presentation of vestibular and perineal fistula in the neonatal period or beyond the neonatal period [16,17]. Peña [18] proposed surgical correction at 2–3 months. However, there is no universal standard of surgical management of late presentation of perineal and vestibular fistulae.

Most of the reported studies used either one or three stages for surgical correction. According to this study and the previous experience of our centers, two-stage repair is more convenient for these patients with late presentation. Colostomy first performed in the three-stage repair, can be replaced by preoperative good rectal wash to decrease rectal dilatation. One-stage repair carries the increased risk of perineal wound complications with decreasing the chance of continence especially in late presenter. Moreover, repair with covering colostomy offers a safe option for decreasing perineal wound complication with good perineal and sphincter muscle function thereafter. This agreed with Khalifa *et al.* [19] in their study comparing one-stage vs two-stage procedure for rectovestibular fistula in that two-stage procedure was safer than one stage, although the age of their patients ranged from 3 months to 2 years.

Gupta *et al.* [20] compared the outcome of one-stage vs conventional three stages on female patients with perineal and vestibular fistulae, with variable ages up to 14 years. They concluded that primary one-stage definitive surgery had significant higher complications.

Although colostomy has many complications [21], according to this study, a covering colostomy is a safer way to avoid complications of perineal wound

after correction of ARM particularly in those presented late for definitive surgery. In a study by Sham *et al.* [13], they did correction of 13 female patients and their age ranged from 13 to 32 years; primary ASARP repair was done in only three cases and staged repair was done in other patients.

In this study, tapering proctoplasty was done only in one patient. Postoperatively, this patient had mild constipation and soiling. The constipation was controlled by diet modification, whereas the soiling was improved after one year and became apparent only during the attacks of diarrhea. In a study by Rawat *et al.* [14], tapering proctoplasty was needed in three cases using PASARP owing to severe rectal dilatation. Soiling and constipation occurred in these cases. The authors provided explanation that this may be due to the development of dilatation and redundancy in the rectosigmoid region with time.

In this study, three patients developed superficial wound infection that improved by local wound care; two patients were adolescents with vestibular fistula with history of previous surgery, and the third patient underwent tapered proctoplasty. In a study by Sharma and Gupta [4], 33 female patients with age ranged from 5 months to 14 years underwent one-stage PASARP. Superficial wound infection developed in two patients and reoperation was not required. In the present study, constipation was the main problem among the studied patients, which occurred in 11 (44%) patients; however, it was controlled with diet modification in eight (32%) patients and only three (12%) patients needed laxative.

Continence was assessed by different scores used to assess the surgical results after correction of ARM [22]. In this study, patients were assessed by Krickbeck continence score [5]. During follow-up period, children and adolescent patients continued wearing diapers for at least 5 months with gradual improvement. With follow-up, good results were obtained in most of patients.

Limitation of this study

This study considered short-term and intermediate-term outcomes; however, long-term follow-up of these cases is needed, and this will be done in future study. This study included vestibular with perineal fistulae as one group of patients, although they have different pathology and presentation; however, the plan of surgical correction is nearly the same to some extent. Moreover, this study targeted to discuss causes of late presentation and surgical outcome after two-stage repair.

Conclusion

Although delayed presentation of ARM with vestibular and perineal fistulae in female patients is accompanied by comorbidities, with good surgical management, successful results can be obtained. Two-stage repair offers a safe option for surgical correction, has good continence outcomes, and decreases the morbidity of multistage procedure.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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