



Evaluation of the functional outcome of the neonatal one stage posterior sagittal anorectoplasty (PSARP) as a procedure to treat cases of high anorectal malformation in male neonates

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ABSTRACT

Background: Male anorectal malformations have many varieties, from simple types that can be easily repaired in one stage to more complex types that need more sophisticated repair in a staged approach. The staged approach has higher morbidity than the one-stage approach and includes colostomy complications, risk of repeated anesthesia and surgery, high cost and psychological burden on parents. Based on these disadvantages of the staged posterior sagittal anorectoplasty (PSARP) for high anorectal malformations, one-stage neonatal PSARP has been developed. The aim of our work was to evaluate the functional and clinical outcome of the one-stage approach in the treatment of selected cases of male neonates with high anorectal malformations

Material and methods: Our retrospective study which included male patients who had neonatal one-stage PSARP for their high anorectal anomalies between 2006 and 2013. Inclusion criteria included patients with flat or mildly distended abdomen; those in whom the cross-table film of the abdomen showed the rectal pouch reaching down to the fifth sacral segment and patients passing meconium from the urethra. However, cases with complete sacral agenesis, major cord anomalies, and a markedly distended abdomen—possibly due to a megarectum—were excluded from the selection. Patients were evaluated for their functional and clinical outcome at the time of the follow-up in January 2018. Patients with poor outcomes were further evaluated by MRI and examination under anesthesia.

Results: Our retrospective study included 24 patients with an age range between 5 and 11 years at the time of follow up in January 2018. Sixteen patients had a recto-bulbar fistula, six patients had a recto-prostatic fistula, and only two patients had recto-bladder neck fistula. According to the continence outcome, 15 patients had a good outcome, all of them had a recto-bulbar fistula. Six patients had a fair outcome; 5 of them had a recto-prostatic fistula, and only one patient had a recto-bulbar fistula. The remaining three patients who had poor outcome included all patients with recto-bladder neck fistula (2 patients) and one patient with recto-prostatic fistula; this was statistically significant ($P < 0.001$). MRI and examination under anesthesia for the three patients with poor continence outcome revealed underdeveloped muscle complex and centrally positioned anus in all patients.

Conclusion: One stage neonatal PSARP for males with high anomalies needs special surgical experience, and it should be used only in selected cases. It has the advantage of avoiding colostomy complications, the cost of repeated surgeries and parents' psychological burden. Recto bladder neck fistula and poor sphincter muscle development are the main causes of poor functional outcome later on.

Key words: Male neonate, one stage PSARP, high anorectal malformation

Introduction

Anorectal malformations are birth defects in which there is an absent or malformed anus. These anomalies have a wide range of variations from simple to com-

plex types with different terminology. The general incidence is 1:5000 live births with an equal male: female ratio. However low anomalies are more common in females accounting for 90% of cases than in males occur-

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ring in 50% of cases. Anorectal malformations require rapid surgical intervention finding a pathway for stools to outside and according to the anomaly; surgery is either done by one stage perineal anoplasty or in staged form starting with colostomy followed by the definitive repair later on [1].

High anorectal malformations in males are usually associated with mild to moderate abdominal distension, non-bulging perineum and meconium may be detected on penile meatus. In a male newborn with high anorectal malformation; a diverting colostomy is the safest procedure for surgeons who do not have significant experience in surgery of anorectal malformation with the definitive repair postponed later on and lastly closure of this stoma [2].

Although the preliminary colostomy avoids wound infection and dehiscence, it is an invasive neonatal procedure carrying a high rate of complications including wound infection, bleeding, excoriation or dehiscence, stoma prolapse or stenosis, electrolyte and fluid imbalance which are poorly tolerated by the patients and their guardians [1].

Many authors studied the neonatal one stage posterior sagittal anorectoplasty for male patients with high anorectal malformation as a procedure that can avoid the high cost and morbidity associated with the three staged procedure especially in the developing countries. They also believe that the sterile meconium within the 1st week of life carries a low risk of wound infection in case of fecal contamination. Another point of view is that the earlier the repair of anorectal malformation, the earlier is the establishment of brain defecation reflex and hence better continence outcome [3].

The work aimed to evaluate the one stage posterior sagittal anorectoplasty as a surgical approach to treat male neonates with high anorectal malformations without a preliminary colostomy. The long-term functional and anatomical outcomes were evaluated as well as long-term complications if any.

Material and Methods

Our retrospective study was conducted at the Elshatby University Hospital, Alexandria, Egypt. We reviewed the records of male neonates with high anorectal malformation who underwent one-stage neonatal posterior sagittal anorectoplasty without prelimi-

nary colostomy in the period from 2006 to 2013 and evaluated the functional outcome of the procedure in January 2018.

These patients were selected pre-operatively according to certain criteria. These inclusion criteria which were very important involved patients with:

- Flat or mildly distended abdomen
- Cross table film showing that the rectal pouch reaching down to the fifth sacral segment
- Passage of meconium from the urethra.

However, cases with complete sacral agenesis, major cord anomalies and patients with markedly distended abdomen -that could be due to a megarectum- were excluded from the selection

After obtaining a written consent; patients' data were collected by direct interview with them and their guardians and the following data were collected:

- A. Age at the follow-up
- B. Type of fistula
- C. Clinical examination: which included
 1. Location of the anus: Normal site or anterior displacement or posterior displacement or lateral displacement
 2. Size of the anus: Good size or stenotic anus
 3. The skin around the anus: Excoriated or not excoriated
 4. The muscle tone as felt by clinical examination: good tone or weak tone.
 5. Ectropion: Presence or absence of an ectropion
- F. Functional outcome measures: post-operative fecal continence was assessed using Rintala and Lindahl continence scoring system as in the following table [4].
- G. Investigations for cases having a poor continence score: Patients with poor continence score were subjected to:
 1. MRI.
 2. Examination under anesthesia: by Peña current muscle stimulator for the evaluation of the sphincter muscle complex.

According to the collected data; patients were divided into three groups: the first group represents the cases with good continence score, the second represents those with fair continence score and the third one represents those with poor continence score. The results of the three studied groups were compared to each

	3	2	1	0
Ability to hold back defecation	Always	Problems less than once/week	Weekly problems	No voluntary control
Feels/reports the urge to defecate	Always	Most of the time	Uncertain	Absent
The frequency of defecation		Every other day to twice a day	More/less often	
Soiling	Never	Staining less than 1/week, no change of underwear required	Frequent staining, change of underwear often required	Daily soiling, requires protective aids
Accidents	Never	Fewer than 1/week	Weekly accidents; often requires protective aids	Daily, requires protective aids during day and night
Constipation	No constipation	Manageable with diet	Manageable with laxatives	Manageable with enemas
Social problems	No social problems	Sometimes (foul odors)	Problems causing restrictions in social life	Severe social and/or psychic problems

Good continence: ≥ 18 , Fair continence: 9-17, Poor continence: 6-9

other regarding the type of fistula, anal stenosis, presence or absence of perianal excoriation, anal sphincter tone and presence or absence of ectropion.

Statistical analysis

Data were fed to the computer and analyzed using IBM SPSS software package version 20.0. (Armonk, NY: IBM Corp) [5]. Qualitative data were described using number and percent. The Kolmogorov-Smirnov test was used to verify the normality of distribution. Quantitative data were described using range (minimum and maximum), mean, standard deviation and median. The significance of the obtained results was judged at the 5% level of confidence

The used tests were

1. Chi-square test: For categorical variables, to compare between different groups.
2. Fisher's Exact or Monte Carlo correction: Correction for chi-square when more than 20% of the cells have expected count less than 5.
3. F-test (ANOVA): For normally distributed quantitative variables, to compare between more than two groups.

Results

Our study included 24 male patients with a history of high anorectal malformation, all of them had been repaired by one stage neonatal PSARP in the period from 2006 to 2013 on their first day of life. The procedure had been completed by this approach in 22 patients. However, conversion into abdominal exploration through left hockey stick incision was done in

two cases who were proved to have recto-bladder neck fistula and highly located rectal pouch. There were no reported cases of immediate intra-operative complications like the injury of the prostate or the urethra.

The patients' age at follow up was (5 - 11 years) with a median of 7.5 years. Also, according to the assessment of continence outcome as measured by Rintala and Lindahl continence scoring system; they were classified into three groups: good, fair and poor continence groups as shown in Table 1.

Review of patients' records revealed that there were different types of the fistula with recto-bulbar one in 16 patients, recto-prostatic one in 6 patients and the last two patients had recto bladder neck fistula. There was a statistically significant relationship between the type of fistula and the functional outcome as patients with recto-bladder neck type of fistula had poor outcome while patients with recto-bulbar and recto-prostatic fistula had a better outcome as shown in Table 2.

The clinical examination of the studied patients at follow up revealed different findings as shown in Table 3. There were no reported cases with retraction of the

Table 1. Distribution of the studied cases according to continence score (n = 24).

Outcome	Number	%
Good	15	62.5
Fair	6	25
Poor	3	12.5
Total	24	100

Table 2. Relation between the functional outcome and the type of fistula.

	Continence						χ ²	MC _p
	Good (N=15)		Fair (N=6)		Poor (N=3)			
	No	%	No	%	No	%		
Rectobulbar	15	100.0	1	16.7	0	0.0	24.261*	< 0.001*
Rectoprostatic	0	0.0	5	83.3	1	33.3		
Recto-bladder neck	0	0.0	0	0.0	2	66.7		

Table 3. Distribution of the studied cases according to clinical signs.

	Continenace						χ2	MC _p
	Good (N=15)		Fair (N=6)		Poor (N=3)			
	No	%	No	%	No	%		
Anal Stenosis								
Absent	15	100.0	5	83.3	2	66.7	4.508	0.130
Present	0	0.0	1	16.7	1	33.3		
Skin excoriation								
Absent	13	86.7	2	33.3	0	0.0	10.249*	0.001*
Present	2	13.3	4	66.7	3	100.0		
Sphincter tone								
Good	15	100.0	1	16.7	0	0.0	20.345*	<0.001*
Weak	0	0.0	5	83.3	3	100.0		
Ectropion								
Absent	10	66.7	4	66.7	1	33.3	1.345	0.694
Present	5	33.3	2	33.3	2	66.7		

χ^2 , p: Chi-square test, MC_p : Monte Carlo test, *: Statistically significant at $p \leq 0.0$

rectal stump nor post-operative urinary incontinence.

There were no cases of anal stenosis among those with good continence outcome. However, anal stenosis did not affect the functional outcome significantly ($P=0.130$) (Table 3).

All of the patients with poor continence had skin excoriation, this coincides with the poor outcome and had a statistically significant relationship ($P=0.001$) (Table 3).

Digital rectal examination demonstrated that the status of analysis sphincter tone correlates significantly with the functional outcome ($P=0.001$) (Table 3).

Ectropion was detected in 9 patients with similar frequency among patients of good and poor continence (about 33%) and higher frequency among patients with fair continence (66%). This difference in frequency of evolution among the studied groups did not affect the outcome significantly ($P=0.694$) (Table 3).

Analysis of the collected data revealed that there

is a statistically significant relationship between the anal sphincter tone and perianal skin excoriation and the functional outcome. On the other hand, there was no significant relationship between the functional outcome and the presence of anal stenosis and ectropion as shown in Table 3.

The three patients with poor continence outcome were further investigated by MRI and examination under anesthesia for assessment of the anatomy, development and the tone of anal sphincter with the following results in Table 4.

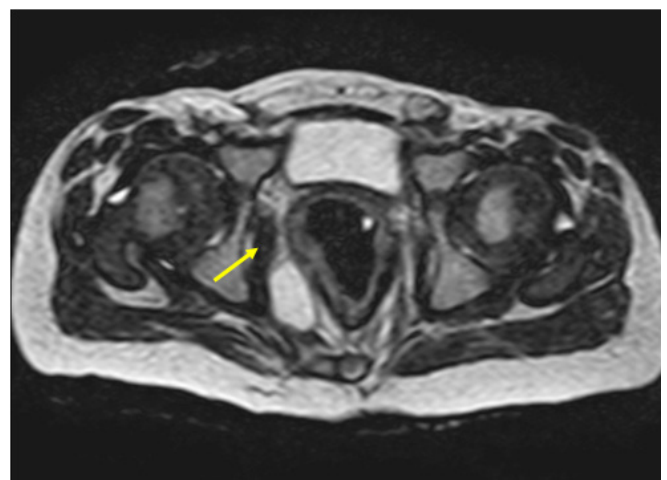
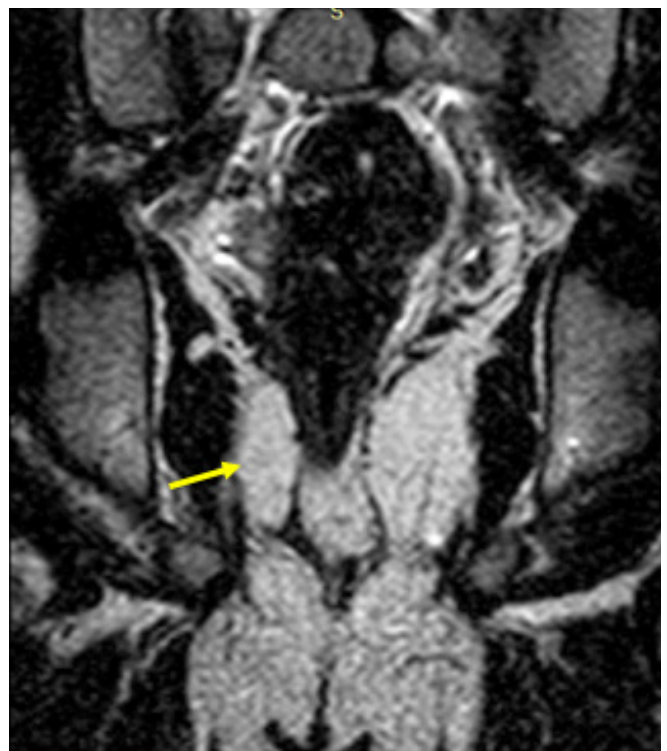
Discussion

The staged posterior sagittal anorectoplasty starting by a diverting descending colostomy followed by the definitive repair and lastly closure of the stoma, popularized by Devries and Pena, made a huge advance in the treatment of patients with anorectal anomalies [6].

Their contribution helped the pediatric surgeons to understand that there are other factors, beyond the

Table 4. Results of MRI and examination by Penna muscle stimulator in patients with poor continence score.

	Case 1	Case 2	Case 3
Sphincter muscle complex	Poorly developed especially at puborectalis muscle	Poorly developed especially at levator ani and external sphincter, being thinner on the right side	Poorly developed especially at levator ani and external sphincter on both sides
The position of pulled through bowel within muscle complex	Central	Central	Central
Associated findings	Partial sacral agenesis tethered spinal cord at level LV4-5, renal anomalies		Abnormal low sacral segments, absent coccyx
MRI	(Figure 1)	(Figure 2)	(Figure 3)
Penna muscle stimulator	Weak all around especially at 6 o'clock	Weak all around especially at 9 o'clock	Weak all around especially at 3 & 9 o'clock

**Figure 1.** Partial sacral agenesis in case 1 with poor continence.**Figure 2.** Poor development of puborectalis muscle especially on right side in case 2 with poor continence.**Figure 3.** Bowel centralized in levator ani which is poorly developed in case 3 with poor continence.

level of the rectum, which may influence the prognosis of the patients complaining of anorectal malformations like the anatomy of the sacrum and the degree of development of the muscle complex. This technique provided excellent exposure of the bowel end and the urinary fistula with the better mobilization of the bowel and safer identification and protection of urinary structures. It became the most common procedure performed for high anorectal malformations in males [6–8].

The traditional staged PSARP is not free from complications including:

- Co-morbidities associated with a colostomy [9].
- The high cost of the staged operations and the increased rate of non-compliance as most patients do

not return within 6-8 weeks for definitive procedure losing the chance of early restoration of defecation reflex, especially in developing countries [10].

In an attempt to reduce the morbidity associated with the classical staged posterior sagittal anorectoplasty, the neonatal one stage approach without preliminary colostomy was developed. This approach is similar to the classical one without protective colostomy. This should be performed by an experienced surgeon in well-selected patients [11].

The selection of the cases at the time of operation is very important for better outcome. This is usually based on multiple criteria such as flat or mildly distended abdomen, cross table film that showed the rectal pouch reaching down to the fifth sacral segment, and the passage of meconium from the urethra (it is unusual to have meconium passing per urethra except in exceptional wide fistula). However, cases with complete sacral agenesis, major cord anomalies and a markedly distended abdomen that could be due to a megarectum should be excluded from the selection [12].

Recent studies concluded that the neuronal continence framework is present since birth but complete development requires early training so Moore highlighted that early restoration of gut continuity in cases of anorectal malformations would establish brain defecation reflex early and then better long-term functional outcome [10,13].

Neonatal repair of anorectal malformation had been advised by Albanese et al., who advocated that early restoration of gut continuity in this deformity would train the pelvic musculature early and then better functional outcome on the long run. Better rectal dissection and tapering –if needed- is noticed in neonatal PSARP due to the absence of pouchitis and fibrosis noticed in a long-term unused rectal pouch in delayed repair [14].

Another advantage of the one stage neonatal repair is noticed in some cases of anorectal malformations which are associated with esophageal atresia requiring colonic esophageal replacement. One stage neonatal PSARP without preliminary colostomy saves the colon for this future procedure like what happened in one of our cases who had a good functional outcome on follow up.

Our study included 24 patients; Mirshemirani et al., [14] included 30 patients however fewer patients were included in Nagdeve et al., [15] study involving only 12 patients.

The age of our patients ranged between 5 and 11 years with a median age of 7.5 years in comparison to the study of Pena et al. in which the median age was 4.5 years [16,17].

There were 2 cases in whom the procedure was not completed through perineal approach with conversion into abdominal assisted approach through left hockey stick incision. These patients were proved to have recto-bladder neck fistula-which is treated through primary abdominal approach-. This conversion was attributed to the absence of preoperative distal loop-gram in those non-colostomized patients.

The fecal continence of our patients was assessed using the Rintala and Lindahl, [4] scoring system which showed higher simplicity in comparison to other scoring systems like Holschneider et al., [17] scoring system and Pena [2] scoring system.

In our study 62.5% of patients had good continence score; this approaches the results of Nagdeve et al., [15] study in which good continence score was detected in 75% of cases. Similarly, Mirshemirani et al., [14] reported that overall 83% of their patients had good continence results.

On the contrary, in their comparative study between a group of patients that underwent a staged procedure and another group that underwent a one-stage procedure, Liu et al., reported that the rate of excellent and good continence was 53.8% (35 of 65 cases) in the one stage group compared to a rate of 58.3% in the 3 stages group with no significant difference between the 2 groups [18].

In our study, there was no statistically significant relationship between the functional outcome and the presence of ectropion or anal stenosis. This is similar to the reported results of Rintala et al. study in which neither the presence of anal stenosis nor ectropion had a relationship with the continence of the studied patients [19].

On the contrary, the skin excoriation around the anus was significantly higher in patients with poor continence; this is due to increased defecation frequency. This was approved by the study of Elhalaby et al. [20].

The anal sphincter tone as being felt clinically by per rectal examination was significantly associated with fecal incontinence in our study. This is similar to the study conducted by Hallan et al. [21].

Regarding the result of MRI examination of the 3 cases with poor continence score; it showed underdevelopment of the sphincter muscle complex; this was concordant to the results of the examination by the muscle stimulator. These observations emphasize the fact that the continence result is multifactorial and depends mostly on the degree of development of the sphincter muscle complex; this was also demonstrated in the study conducted by Ros Mendoza et al. [22].

Our study concluded that primary repair of high anorectal malformation in male neonates that are properly selected is technically feasible and safe. It avoids the risk of complications related to colostomy, the risk of repeated anesthesia and reduces the economic and psychological burden on the family.

A multi-institutional study with a large number of patients and long-term anorectal function follow up is recommended to determine whether the one stage procedure is recommended rather than the classical three staged procedure. Also widening the selection criteria to include patients with associated congenital anomalies like meningocele, GIT anomalies, etc. to save these patients from co-morbidities of the staged procedure.

Conflict of interest statement

The authors have no conflicts of interest to declare.

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