ORIGINAL ARTICLE

Functional Outcomes in Anorectal Malformation Patients Following Definitive Surgery

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ABSTRACT

Introduction: Recently, management of anorectal malformation (ARM) emphasis on good intestinal functional outcomes after definitive procedure. This study analyzed the patients' outcomes following operation related with the predictive variables. **Methods:** We applied the Krickenbeck classification and Rintala scoring system to define ARM type and functional outcomes, respectively. **Results:** This study ascertained 72 patients: 38 males and 34 females. According to Rintala scoring system, 94.4%, 90.2%, 60%, 83.3%, and 60% patients showed no soiling, no constipation, ability to hold back defecation, defecation frequency of every other day to twice a day, and feels/reports the urge to defecate, respectively. In addition, none of patients had either accident or social problem. Rintala score of normal and good have been shown in 14 (19.4%) and 55 (76.4%) patients, respectively. Female patients had a 4.2-times higher risk for showing a more/less often frequency of defecation compared with male patients (95% confidence interval (CI)=1.03-17.1; p=0.035). **Conclusions:** ARM patients' functional outcomes after procedure in our institution are considered relatively good. In addition, the frequency of defecation in male patients after definitive surgery is better than female patients.

Keywords: Anorectal malformation; Functional outcomes; Gender; Prognostic factors; Scoring system

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INTRODUCTION

Anorectal malformation (ARM) is the most common congenital disorder in pediatric surgery. Its incidence is approximately 1:5,000 live birth (1). Currently, the Krickenbeck system has been used to classify ARM types based on the anatomical landmark of the rectal fistula (2).

Advances in surgical technique and neonatal care in the last decade have increased the survival of ARM patients. Therefore, Recently, management of ARM emphasis on good intestinal functional outcomes after definitive procedure (3-5). The prognosis of ARM can be affected by several factors (6-8). Functional outcomes after definitive surgery vary among scoring systems (3,4). In addition, the Rintala scoring system is the only questionnaire that was also validated on a healthy children population (4). Therefore, this study analyzed the ARM patients' outcomes following definitive surgery using the Rintala scoring system associated with the prognostic factors, consisting of gender, anoplasty approach (posterior or anterior sagittal anorectoplasty, PSARP/ASARP), one/three-stage repair, age at anoplasty, anoplasty dehiscence, and bougienage procedure.

MATERIALS AND METHODS

Patients

We ascertained children with ARM who underwent definitive repair in our institution from August 2012 to September 2016. We collected and reviewed 72 ARM patients' medical records retrospectively. The type of ARM was established by perineal inspection 24 hours after birth, followed by a cross-table lateral film for patients without clinically identified fistula to determine the level of distal gas shadow, or distal colostography before anoplasty to demonstrate the location of the blind rectum and the precise site of fistula (9). Krickenbeck classification was used to determine the ARM type (2). The Ethics Committee of our institution gave approval for this study (KE/FK/273/EC/2016).

Surgical and bougienage procedures

PSARP and ASARP were performed according to previous studies (10,11). One-stage repair was anoplasty without a colostomy, while three-stage repair consisted of colostomy at newborn, followed by anoplasty at three months of age, and stoma closure at three months after anoplasty (9).

Bougienage was conducted two weeks after anoplasty according to previous study (12), with the frequency of twice a day, tapered to once a day for 1 month, every third day for 1 month, twice a week for 1 month, once a week for 1 month and once a month for 3 months, after the dilator passes the anus easily without pain and no resistance. Moreover, the dilator size was increased every week until the anus gets the appropriate size according to the age of patients (12).

In addition, we determined anoplasty dehiscence as skin rupture (superficial) or subdermal structures involved (deep) up to 30 days postoperatively (13).

Rintala scoring system

ARM patients' functional outcomes following repair were analyzed using the Rintala scoring system. It consists of seven factors as follows: ability to hold back defecation, feels/reports the urge to defecate, soiling, frequency of defecation, constipation, accident, and social problems (14). The Rintala scoring system was determined as normal (score ≥18), good (12–17), fair (7–11) and poor (≤6) (14,15). We assessed the functional outcomes in patients ≥3 years old because the estimation of toilet training is within this age (16,17).

Statistical analysis

The data were shown as frequency and percentage. The association of prognostic factors (gender, anoplasty approach, one/three-stage repair, age at anoplasty, anoplasty dehiscence, and bougienage procedure) and functional outcomes in ARM patients after definitive repair was analyzed using Chi-square or Fischer Exact test. We determined $p<0.05$ as a significant level. For analysis of association between prognostic factors and frequency of defecation in ARM patients, we combined "more often" and "less often" categories into one group, i.e. more/less often group.

RESULTS

Baseline characteristics

There were 72 ARM patients who had complete medical records to be analyzed further, consisting of 38 males and 34 females. Their clinical characteristics are described in Table I. Most patients underwent three-stage surgery (73.6%) and posterior sagittal anorectoplasty (66.7%). According to Krickenbeck classification, most male patients had ARM without fistula (44.7%), while most female patients showed ARM with vestibular fistula (52.9%) (Table I).

Associated anomalies

Eight patients were diagnosed with Down syndrome, followed by genitourinary (4) and cardiovascular (3) anomalies (Table I).

Functional outcomes using Rintala scoring system

Our study revealed that 94.4%, 90.2%, 60%, 83.3%, and 60% patients showed no soiling, no constipation, ability to hold back defecation, defecation frequency of every other day to twice a day, and feels/reports the urge to defecate, respectively. In addition, none of patients had either accident or social problem (Table II).

Table I: Clinical characteristics, types and anomalies/syndrome of ARM patients who underwent definitive surgery

Characteristics	n (%)
Gender	
▪ Male	38 (52.8)
▪ Female	34 (47.2)
Age at anoplasty	
▪ <6 months old	25 (34.7)
▪ ≥6 months old	47 (65.3)
Anoplasty approach	
▪ PSARP	48 (66.7)
▪ ASARP	24 (33.3)
Repair step	
▪ One-stage repair	19 (26.4)
▪ Three-stage repair	53 (73.6)
Anoplasty dehiscence	
▪ No	34 (47.2)
▪ Yes	38 (52.8)
Bougienage procedure	
▪ Not routinely performed	15 (20.8)
▪ Routinely performed	57 (79.2)

Sex	Group	Type	N (%)
Male (n=38)	Major	ARM without fistula	17 (44.7)
		ARM with perineal fistula	11 (28.9)
		ARM with rectourethral fistula	6 (15.8)
		ARM with rectovesical fistula	4 (10.5)
Female (n=34)	Major	ARM with vestibular fistula	18 (52.9)
		ARM without fistula	8 (23.5)
		ARM with perineal fistula	7 (20.6)
	Rare/ regional variant	ARM with rectovaginal fistula	1 (2.9)

Anomalies	n (%)
▪ Genitourinary system	4 (5.6)
▪ Cardiovascular system	3 (4.2)
▪ Central nervous system	1 (1.4)
▪ Musculoskeletal system	1 (1.4)
Syndrome	
▪ Down syndrome	8 (11.1)
▪ VACTERL	1 (1.4)

ARM, anorectal malformation; PSARP, posterior sagittal anorectoplasty; ASARP, anterior sagittal anorectoplasty; VACTERL, vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities

Total Rintala score in ARM patients

Rintala score of normal and good have been shown in 14 (19.4%) and 55 (76.4%) patients, respectively (Table II).

Prognostic factors and ARM patients' functional outcomes correlation

Next, we determined the prognostic factors and ARM patients' functional outcomes correlation. Female patients had a 4.2-times higher risk for showing a more/less often frequency of defecation compared with male

Table II: Functional outcomes and total of Rintala score for ARM patients following definitive surgery

Criteria	Score	%
Ability to hold back defecation		
▪ Always	3	36
▪ Problems less than 1/week	2	24
▪ Weekly problems	1	40
▪ No voluntary control	0	0
Feels/reports the urge to defecate		
▪ Always	3	36
▪ Most of the time	2	24
▪ Uncertain	1	40
▪ Absent	0	0
Frequency of defecation		
▪ Every other day to twice a day	2	83.4
▪ More often	1	8.3
▪ Less often	1	8.3
Soiling		
▪ Never	3	94.4
▪ Staining less than 1/week, no change underwear required	2	5.6
▪ Frequent staining, change of underwear often required	1	0
▪ Daily soiling, require protective aids	0	0
Accident		
▪ Never	3	100
▪ Fewer than 1/week	2	0
▪ Weekly accidents; often requires protective aids	1	0
▪ Daily, requires protective aids during day and night	0	0
Constipation		
▪ No constipation	3	90.2
▪ Manageable with diet	2	2.8
▪ Manageable with laxatives	1	4.2
▪ Manageable with enemas	0	2.8
Social problems		
▪ No social problems	3	100
▪ Sometime (foul odor)	2	0
▪ Problems causing restrictions in social life	1	0
▪ Severe social and/or mental problems	0	0

Total Score Classification	n (%)
Normal (≥18)	14 (19.4)
Good (12–17)	55 (76.4)
Fair (7–11)	3 (4.2)
Poor (≤6)	0

ARM, anorectal malformation

patients (95% confidence interval (CI)=1.03-17.1; p=0.035) (Table III).

Furthermore, none of prognostic factors correlated to another Rintala scoring system, involving feel/report the urge to defecate, soiling, ability to hold back defecation, soiling, and constipation (p>0.05).

Association between prognostic factors and Rintala scoring system

Subsequently, we analyzed whether the prognostic factors affected the Rintala scoring system. Here, we showed that male subjects were more likely to have a Rintala score of ≥18 (better outcome) than female subjects, but it did not get a significant level (p=0.12) (Table IV).

DISCUSSION

We are able to show that our ARM patients' functional outcomes after procedure are considered relatively good, in regard to the ability to feel the urge to defecate, to hold back defecation, frequency of defecation, soiling, accidents, constipation and social problems. In addition, we found that female patients show worse frequency of defecation than male patients (Table III).

Table III: Association between prognostic factors and frequency of defecation in ARM patients

Characteristics	More/ less often (n, %)	Every other day to twice a day (n, %)	p	OR (95% CI)
Gender				
▪ Male	3 (7.9)	35 (92.1)	0.035*	4.2 (1.03-17.1)
▪ Female	9 (26.5)	25 (73.5)		
Age at anoplasty				
▪ <6 months old	3 (12)	22 (88)	0.52	0.5 (0.1-2.0)
▪ ≥6 months old	9 (19.1)	38 (80.9)		
Anoplasty approach				
▪ PSARP	8 (16.7)	40 (83.3)	1.0	1.0 (0.3-3.7)
▪ ASARP	4 (16.7)	20 (83.3)		
Repair step				
▪ One-stage repair	4 (21.1)	15 (78.9)	0.72	1.5 (0.4-5.7)
▪ Three-stage repair	8 (15.1)	45 (84.9)		
Anoplasty dehiscence				
▪ No	5 (14.7)	29 (85.3)	0.67	0.8 (0.2-2.7)
▪ Yes	7 (18.4)	31 (81.6)		
Bougienage procedure				
▪ Not routinely performed	1 (6.7)	14 (93.3)	0.44	0.3 (0.04-2.5)
▪ Routinely performed	11 (19.3)	46 (80.7)		

*, p<0.05; ARM, anorectal malformation; ASARP, anterior sagittal anorectoplasty; CI, Confidence Interval; OR, Odds Ratio; PSARP, posterior sagittal anorectoplasty

Table IV: Association between prognostic factors and Rintala scoring system in ARM patients

Characteristics	Score ≥18 (n, %)	Score <18 (n, %)	p	OR (95% CI)
Gender				
▪ Male	10 (26.3)	28 (73.7)	0.12	2.7 (0.8-9.5)
▪ Female	4 (11.8)	30 (88.2)		
Age at anoplasty				
▪ <6 months old	5 (20)	20 (80)	1.0	1.1 (0.3-3.6)
▪ ≥6 months old	9 (19.1)	38 (80.9)		
Anoplasty approach				
▪ PSARP	8 (16.7)	40 (83.3)	0.53	1.7 (0.5-5.5)
▪ ASARP	6 (25)	18 (75)		
Repair step				
▪ One-stage repair	4 (20)	15 (80)	1.0	0.9 (0.2-3.2)
▪ Three-stage repair	10 (18.9)	43 (81.1)		
Anoplasty dehiscence				
▪ No	9 (26.5)	25 (73.5)	0.15	0.4 (0.1-1.4)
▪ Yes	5 (13.2)	33 (86.8)		
Bougienage procedure				
▪ Not routinely performed	5 (33.3)	10 (66.7)	0.15	0.4 (0.1-1.4)
▪ Routinely performed	9 (15.8)	48 (84.2)		

ARM, anorectal malformation; ASARP, anterior sagittal anorectoplasty; CI, Confidence Interval; OR, Odds Ratio; PSARP, posterior sagittal anorectoplasty

Interestingly, although not statistically significant, female patients also tend to have a poorer bowel functional outcome (Rintala score of <18) than male patients (Table IV). Our finding confirmed previous report that showed males with perineal fistula have a better outcome than females with the same ARM type (27). These outcomes differences might not be affected by either the anatomy, sacral abnormalities or presence of syndrome (27). They proposed some hypotheses to explain this gender different outcome: 1) inappropriate anoplasty in female patients due to a limited dissection of the rectum because of a fear of perforating the vagina; and 2) female patients less openly discussed with their families regarding their bowel function, resulting in failure of bowel management (27-29). However, our findings should be interpreted carefully since none of the scoring methods to determine functional outcomes in ARM patients have been generally established to date

(4). Moreover, previous study revealed that gender does not affect the bowel functional outcomes in an Italian population (15). This discrepancy might be due to the difference of the populations analyzed, i.e. all ARM types (our study) vs. ARM with good prognosis, i.e. some types of ARM with prominent midline groove and normal sacrum (15).

We assessed the functional outcomes in patients ≥3 years old because the estimation of toilet training is within this age (16,17), however, we did not separate the patients according to their age (i.e. young children and adolescents) during the analysis. Notably, the functional outcome usually improves as the ARM patients reach adolescence (18).

In this study, we utilized the Rintala scoring system to determine the functional intestinal outcomes in ARM patients after procedure. The advantage of the Rintala scoring system over other different scores is that it is the only questionnaire that was also validted on a healthy children population (4). In addition, all scoring systems determine functional outcomes using various parameters, resulting in different answering choices and final scores among scoring systems (4).

Most of our patients had Rintala scores of 12-17 (76.4%) and ≥18 (19.4%) that are considered as good and normal outcomes, which might be associated with the relative high frequency of low ARM (perineal fistula, 25%) in our cohort. It has been shown that low ARM patients have good bowel function and do not differ much from healthy peers (19). Further study is necessary to determine the association between ARM type and functional outcomes in our patients. Almost half of our patients showed ARM without fistula. One of the possible explanations for this is the relative high incidence of Down syndrome in our study (11.1%) as noted by previous study (20).

Moreover, more than half of our patients suffered an anoplasty dehiscence. Previous study showed that one-stage repair has a higher risk for dehiscence than multi-stage repair (13). Further study is important to determine the risk factors, including one/multi-stage repair, age at anoplasty, gender, and presence of a genitourinary anomaly (13,21), for development of dehiscence in our cohort patients. Anoplasty dehiscence might cause stricture, resulting in constipation (22). Our study showed that almost all patients had no constipation. This finding might relate to the fact that there was no correlation found between prognostic factors and constipation in our subjects.

Bougienage that is routinely performed after definitive surgery did not have an association with our patients' functional outcomes. In contrast, previous report described that long-term anal bougienage is needed to decrease morbidity and provide good result (22), because it will ease the stool passage, and prevent the constipation and stricture (12).

We did not find any association between anoplasty approach (PSARP vs. ASARP) and patients' functional outcomes (Table II). However, one study from India clearly showed that ASARP is preferable for female ARM patients due to better cosmetics and good outcomes (23). In addition, PSARP might cause constipation, especially in female patients with rectovestibular fistula, because of untreated anal stricture or due to colonic motility disorder (5,24). We also did not find any significant effect between one-stage and three-stage repair for functional outcomes, although several studies have successfully shown the benefit of one-stage over three-stage repair (25,26).

Age at anoplasty is also still controversial. Early anoplasty leads to early anal dilatation (bougienage) and anal dilatation is easier to be performed in infant age than older age (22). In contrast, early anoplasty has a higher risk of organ injury because the organ identification in neonates during surgery is not easy. Our study is compatible with previous study that found age at anoplasty did not influence functional outcomes (15). This study was a retrospective design, therefore, some data were incomplete, such as other prognostic factors that might affect the functional outcomes, e.g. type of ARM, sacral ratio and abnormalities of spine; becoming a weakness of our report (30). We also evaluated all male cases with recto-urethral bulbar fistula and recto-urethral prostatic fistula together under the label of "recto-urethral fistula". These facts should be considered during interpretation of our findings since they might have a difference in prognosis (7,30). Moreover, Rintala scoring is designed to be completed by patient/parent independently or they answer the questions presented by a person who is not involved in the management of the patients (14).

CONCLUSION

ARM patients' functional outcomes after procedure in our institution are considered relatively good. In addition, the frequency of defecation in male patients after definitive surgery is better than female patients.

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