

## Original Article

# Long-term outcomes and quality of life of children following anorectoplasty: a single institutional review of 138 cases

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**Abstract:** Objective: This study aims to assess the long term functional outcomes and quality of life (QOL) of children after anorectoplasty. Methods: A retrospective study was conducted on 138 patients with ARMs, who underwent anorectoplasty between December 2000 and July 2013 in Xinhua Hospital. Functional outcomes were assessed using Kelly's Score (minimum = 0, maximum = 6) and the Japanese Study Group of Anorectal Anomalies (JSGA) score (minimum = 0, maximum = 8). QOL was assessed using the Pediatric QOL Inventory (PedsQL). Results: According to Kelly's Score, functional outcome was classified as good in 48 (34.8%) patients (score: 6), fair in 89 (64.5%) patients (score: 4-5), and poor in one (0.7%) patient (score: 2). According to the JSGA score, functional outcome was classified as good in 48 (34.8%) patients (score: 7-8), fair in 80 (58.0%) patients (score: 5-6), poor in nine (6.5%) patients (score: 3-4) and very poor in one (0.7%) patient (score: 0-2). Patients with associated sacral deformities were found to have poorer bowel functional outcome than those without sacral deformities. Furthermore, patients with low-type ARMs had better QOL compared to those with high-type ARMs. Conclusions: A number of children had fair functional outcome after anorectoplasty. The QOL of children after anorectoplasty depends on numerous factors. Various surgical techniques and different scoring systems could affect the results. Associated malformations, complications and higher level ARMs directly lead to negative outcomes. Postoperative functional outcome assessment anorectal functional training and psychological support are important for improving QOL.

**Keywords:** Anorectal malformation, anorectoplasty, quality of life, prognosis

## Introduction

Anorectal malformations (ARMs) are the most common pediatric alimentary tract anomalies, with an incidence in live births of 1:1,500-5,000 [1]. ARMs are classified using the Krickbeck and Wingspread classification into high-, intermediate-, and low-type; and are also grouped depending upon cloacal and rare malformations [2]. Since 1953, the surgical approach to correct ARMs has changed dramatically from sacroperineal to posterior sagittal anorectoplasty (PSARP) and from traditional staged repair to single stage repair [2-4]. The postoperative functional outcome remains variable, and depends not only on the anatomical position of the blind rectal pouch or associated urogenital and sacral deformities, but also on the preservation of pelvic nerves and mus-

cles during dissection. With the development of surgical techniques, the postoperative functional outcome and quality of life (QOL) of ARM patients is not low. 1. A retrospective study of 1192 patients with anorectal malformations, which were operated using PSARP revealed that 75% of them had voluntary bowel movements, and about 37.5% of them were totally continent, while 25% of all patients suffered from fecal incontinence, but their quality of life was significantly improved after participating a bowel management program [3]. Another study showed that in 48 patients with high imperforate anus and undergoing PSARP, 77% had good and fair stooling outcome, and the percentage of those with good and fair quality of life (QOL) reached 75% [12]. In another study, in 32 perineal fistula patients who underwent single-stage repair, all sixteen pa-

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**Table 1.** Demographic data of the patients

Classification	Malformations	Gender		Average age when anorectoplasty was performed (5.7 ± 3.5)				Types of surgery			
		Female (56, 40.6%)	Male (82, 59.4%)	2-4 years old	5-7 years old	8-12 years old	13-18 years old	PSARP	Perineal anoplasty	Incision and dilatation	Laparoscopic assisted anorectoplasty and cloaca combined with PSARP
High position (21, 15.1%)	Anorectal agenesis (4, 2.9%)	0	4 (2.9%)	73 (53%)	34 (24%)	24 (17%)	7 (6%)	77 (56.1%)	45 (32.4%)	10 (7.2%)	6 (3.6%)
	Rectal atresia (17, 12.3%)	4 (2.9%)	13 (9.4%)								
Intermediate (70, 48.9%)	Rectovestibular fistula (26, 18.8%)	26 (18.8%)	0								
	Rectourethral fistula (26, 18.8%)	0	26 (18.8%)								
	Anal dysplasia, No fistula (18, 13.0%)	2 (1.4%)	16 (11.6%)								
Low position (47, 34.6%)	Rectovestibular fistula (12, 8.7%)	12 (8.7%)	0								
	Rectum to skin fistula (8, 5.8%)	8 (5.8%)	0								
	Anal skin fistula (21, 15.2%)	0	21 (15.2%)								
	Archostenosis (6, 4.3%)	4 (2.9%)	2 (1.4%)								

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**Table 2.** Functional outcomes of the patients

Functional outcomes	Defecation frequency				Soiling	
	Constipation	1-2 timesperday	3-4 timesperday	5-6 timesperday	No soiling	1-2 timesper week
Patient number (percentage)	13 (9.4%)	76 (55.1%)	36 (26.1%)	11 (8.0%)	79 (57.6%)	59 (42.4%)

**Table 3.** Distribution of Kelly score and JSGA score

Score	Kelly score	JSGA score
1	NA	0
2	1 (0.7%)	1 (0.7)
3	0 (0.0%)	1 (0.7%)
4	29 (21.0%)	8 (5.8%)
5	60 (43.5%)	37 (26.8%)
6	48 (34.8%)	43 (31.2%)
7	NA	32 (23.2%)
8	NA	16 (11.6%)

NA: not applicable.

tients older than five years of age can voluntarily control bowel movements, and no patient had constipation [4]. However, the assessment method of postoperative functional outcome of patients varies between studies, leading to ambiguity in this subject. So in this study, using two score systems and Pediatric Quality of Life Inventory™ 4.0 (PedsQL™), we retrospectively reviewed the postoperative functional outcomes and QOL of 138 pediatric patients who underwent anorectoplasty to correct ARMs during a time span of 13 years in our center. These findings may be useful to pediatric surgeons and parents to evaluate the potential outcomes of children after the anorectoplasty and improve the QOL of these patients.

### Materials and methods

#### Subjects

From December 2000 to July 2013, 367 patients with ARMs were treated in Xinhua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine. After excluding patients with incomplete medical records, 138 patients were included in our analysis. This study was conducted in accordance with the declaration of Helsinki. The methodology was approved by the Human Ethics Committee of Xinhua Hospital at Shanghai Jiao Tong University School of Medicine. Written informed consent was obtained from all participants.

#### Retrospective review

Analyses of clinical data including demographics, ARM type, surgical techniques and post-operative morbidities were performed. Post-operative functional outcomes were assessed using Kelly's Score (minimum = 0, maximum = 6; severity of accidents 0-2, severity of staining 0-2, and strength of sphincter squeeze 0-2) and the Japanese Study Group of Anorectal Anomalies (JSGA) score (minimum = 0, maximum = 8; presence of urge to defecate 0-2, severity of incontinence 0-4, severity of constipation 0-4, and severity of soiling 0-2). QOL was assessed using the Pediatric QOL 4 Inventory (PedsQL) [5].

#### Statistical analysis

Values were reported as mean  $\pm$  standard deviation. Continuous variables were compared using the Mann-Whitney *U*-test. Discrete variables were compared using the Kruskal-Wallis test. All statistics were analyzed using SPSS version 19.0 for windows. A *P*-value  $<$  0.05 was considered statistically significant.

### Results

A total of 138 patients (82 males and 56 females) with ARMs were included in the study. The mean birth weight was  $3,219.20 \pm 457.10$  g (range: 2,000-4,500 g). The average age of these patients when anorectoplasty was performed was  $5.7 \pm 3.5$  years. Furthermore, 73 patients were 2-4 years old (53%), 34 patients were 5-7 years old (24%), 24 patients were 8-12 years old (17%), and seven patients were 13-18 years old (6%) (**Table 1**).

These patients were divided into three groups according to the Krickenbeck and Wingspread classification of anomalies: high-type (21 patients, 15.1%), intermediate-type (70 patients, 48.9%), and low-type (47 patients, 34.6%). Furthermore, 39 patients had associated malformations (28.8%) including 21 patients with sacrococcygeal and spinal cord malformations, eight patients with urinary tract

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**Table 4.** Distribution of deferent ways chosen of surgery

	First stage operation (N, percentage)	Two stage operation (N, percentage)
High position	1 (1.0%)	20 (50.0%)
Intermediate	50 (51.0%)	20 (50.0%)
Low-position	47 (48.0%)	0 (0.0%)

**Table 5.** Compared PedQL 4.0 Scores by classification

Rank	Classification	N	Rank mean
PedQL Scores	Low	47	88.35
	Intermediate	70	66.09
	High	21	38.69
	Total	138	
Test statistics <sup>a,b</sup>			
		PedQL Scores	
Chi-Squar		23.434	
df		2	
Asymptotic significance		.001	

a. Kruskal Wallis test; b. Grouping variables: classification.

**Table 6.** Compared PedQL 4.0 scores by kelly scores

Rank	Kelly Scores	N	Rank mean
PedQL Scores	4.00	29	39.69
	5.00	59	59.64
	6.00	49	97.62
	Total	137	
Test statistics <sup>a,b</sup>			
		PedQL Scores	
Chi-Squar		44.586	
df		2	
Asymptotic significance		.000	

a. Kruskal Wallis test; b. Grouping variables: Kelly Scores.

malformations, six patients with congenital heart disease, three patients with esophageal atresia, and one patient with hydrocephalus (**Table 1**). Moreover, 98 patients underwent a single-stage operation (70.5%), while 40 patients underwent a two-stage operation (28.8%) (**Table 4**).

Seventy-seven patients underwent PSARP (56.1%), in which 45 patients had perineal ano- plasty (32.4%), 10 patients had incision and dilatation (7.2%), five patients underwent lapa- roscopic assisted anorectoplasty (3.6%), and

one patient with a cloaca underwent com- bined laparoscopic surgery and PSARP (**Table 1**).

### Functional outcomes

Thirteen patients had mild constipation (9.4%), and required occasional fleet enemas. A total of 76 (55.1%) patients defecated 1-2 times per day, in which 36 (26.1%) patients defecated 3-4 times per day and 11 (8.0%) patients defecated 5-6 times per day. Fifty-nine patients experi- enced soiling 1-2 times per week (42.4%) (**Table 2**), while the other patients did not suffer from soiling.

The average Kelly's Score was  $5.1 \pm 0.8$ . Furthermore, 48 (34.8%) patients scored 6, 60 (43.5%) patients scored 5, 29 (21.0%) patients scored 4, and one (0.7%) patient scored 2. Moreover, functional out- come was good (score 6) in 48 (34.8%) patients, fair (score 4 to 5) in 89 (64.5%) patients, and poor (score 2 to 3) in one (0.7%) patient (**Table 3**).

The Kelly's Score in 21 patients with as- sociated sacrococcygeal and spinal cord mal- formations was significantly lower than that in patients without associated spinal deformities ( $4.4 \pm 0.9$  vs.  $5.2 \pm 0.7$ ,  $P = 0.0021$ ).

The mean JSGA score was  $6.0 \pm 1.2$ . Fur- thermore, 16 (11.6%) patients scored 8, 32 (23.2%) patients scored 7, 43 (31.2%) patients scored 6, 37 (26.8%) patients scored 5, eight (5.8%) patients scored 4, one (0.7%) patient scored 3, and one (0.7%) patient scored 2. Moreover, functional outcome was good (sco- re 7-8) in 48 (34.8%) patients, fair (score 5-6) in 80 (58.0%) patients, poor (score 3-4) in nine (6.5%) patients, and very poor (score 0-2) in one (0.72%) patient (**Table 3**).

### Quality of life

The mean score of the PedsQL 4 Inventory was  $67.5 \pm 8.5$ . Physical functioning score was  $68.1 \pm 8.9$ , emotional functioning score was  $66.5 \pm 9.5$ , social functioning scored was  $67.5 \pm 10.0$ , and school functioning score was  $66.3 \pm 11.9$ . The lowest score was 35.0. The ana- tomical position of the blind rectal pouch was found to be significantly correlated with postop- erative QOL. The higher the level of the ARM, the worse the QOL became ( $X^2 = 23.43$ ,  $P =$

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**Table 7.** Compared kelly score by deformity accompanied

Combined malformation	N	Kelly score	Wilcoxon Rank-sum test
Spine and caudal deformity	21	Average value 4.42 ± 0.94	Statistic S = 707.00, Z = -3.04, P = 0.0021 (bilateral)
No spine and caudal deformity	117	5.24 ± 0.70	

0.001) (Table 5). Functional outcomes also correlated with QOL. The lower the Kelly's Score, the worse the QOL became ( $X^2 = 44.59$ ,  $P < 0.001$ ) (Table 6). Patients with associated malformations were found to have significantly lower PedsQL scores ( $Z = -3.04$ ,  $P = 0.0021$ ) (Table 7).

### Discussion

ARMs are the most common pediatric alimentary tract anomalies, which impact slightly more males than females. However, its underlying embryology and pathophysiology remains not fully understood. Despite the use of various surgical techniques in correcting malformations, its postoperative functional outcome remains variable.

PSARP, advocated by Pena in 1982, marked an important evolution in the management of ARMs [6]. The principle of PSARP is to dissect the muscles of continence in a midline plane through a posterior approach without damaging the nerve supply. With advances in laparoscopic techniques, Georgeson advocated for laparoscopically assisted anorectal pull-through in 2000, allowing clearer anatomical delineation and minimizing damage to the pelvic musculature and nerves during dissection [7]. Even in patients with high-type ARMs, which usually associate with other malformations that may impact surgical outcomes, laparoscopic operation has been shown to be safe with rapid postoperative recovery and positive functional outcomes. Using three-dimensional computed tomography in patients with low-type ARMs, Watanabe *et al.* found deformed hypoplastic anal sphincters in many of these cases, suggesting that the choice of surgical technique is important in neo-anus reconstruction at the center of the hypoplastic sphincter [8].

In the present study, the patient with a cloaca and associated sacrococcygeal and spinal cord malformation underwent combined laparoscopic surgery and PSARP. Postoperative complications with persistent incontinence required long-term colostomy decompression for that patient.

Functional outcomes assessment after the surgical correction of ARMs should be performed to evaluate overall neurophysiological function. Most postoperative functional outcomes assessment tools assess the frequency, amount and consistency of stools. Different scoring systems have been compared for validity and reliability [9]. According to these references, Ochi *et al.* found that continence in males with high-type ARMs was not consistently assessed when comparing four assessment protocols [9]. Holschneider *et al.* suggested the assessment of postoperative outcomes based on surgical techniques instead of using any existing scoring system or classification of postoperative continence [10].

Kelly's Score and JSGA scores were used for this study. The JSGA score appeared to be more sensitive than Kelly's Score, with a lower overall score among the patients included in the present study. The functional outcomes of patients with high-type ARMs were worse than those with intermediate- and low-type ARMs, and patients with low-type ARMs had the best functional outcomes among the three groups. The functional outcomes of patients with low-type ARMs are considered to be better than those with high-type ARMs, and the patients with high ARMs have the worst outcomes of faecal control [21].

Associated malformations involving gastrointestinal, cardiac, urological and musculoskeletal systems were found in 28%-72% of patients with ARMs, and were associated with worse functional outcomes than in patients without associated malformations. In the present study, patients with one or more associated malformations, particularly patients with sacrococcygeal and spinal cord malformations, had the worse postoperative functional outcomes. Borg *et al.* studied 37 children with ARMs, and found a high correlation between spinal cord malformation and postoperative bladder dysfunction [11]. Clinicians should assess these associated malformations, in order to better predict postoperative functional outcomes.

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Our study revealed that the QOL of patients with low-type ARMs was very close to normal, and their bowel function went well; whereas the QOL was significantly reduced in children with severe incontinence compared to normal children. The present study revealed that the higher the level of the ARM, the worse the QOL became ( $X^2 = 23.434$  [23.43],  $P = 0.001$ ).

Although surgeries can offer the anatomical correction of ARMs in neonates or infants, low postoperative functional outcomes or complications may cause long-term psychological or social impairments in these patients. It is thereby important to assess QOL after surgery in children with ARMs. Fecal and urinary incontinence and constipation are the main contributors to reducing overall QOL postoperatively in children with ARMs [12-14].

Abnormal defecation in children may lead to psychological and social problems and poor QOL. Negative feelings such as despair, anxiety and depression were common in incontinence patients. Children with incontinence needed long-term anal or hydrotherapy, as well as anus perineal area exposure [15-17]. Furthermore, they experienced low self-confidence and reported of being dissatisfied. Our findings indicate that in children with ARMs, children with normal bowel function had a higher QOL compared to children without normal bowel function. The friends and classmates of children with incontinence might avoid contact with them, and even discriminate against them, leading to the patients feeling frustrated and fearing attendance at public activities. Furthermore, isolation resulted in low QOL scores.

Patient QOL might be associated with the education level and economic situation of the parents. In children with the same level of defecation function, QOL increased with an increase in the mothers' education level [18]. In the present study, poor economic conditions did not associate with poor QOL. However, other reports have shown that a poor family's economic situation negatively affected the children's postoperative recovery [19, 20]. Long-term research on this subject is still needed.

Postoperative outcomes can affect a child's entire future. Therefore, it is necessary to recommend postoperative rehabilitation therapy and work toward continuous improvement in

surgical treatments. Postoperative rehabilitation must receive financial and mental support from society and family. Long-term follow-up care should improve the QOL of children with ARMs. Various training methods can be explored during follow-up care, depending on the type of defecation dysfunction, with treatment approaches including anti-peristalsis agents, laxatives, feces forming agents, or dietary adjustments, in order to change the stool and achieve regular daily bowel movements. During follow-up care, doctors can also provide psychological guidance, helping to minimize the negative impact of bowel dysfunction on a child's psychological development.

ARM patients with associated spinal cord and caudal deformities exhibited poor postoperative bowel function and poor QOL. Score differences existed in four aspects of the postoperative QOL assessment: physiological function, emotional function, social function, and role function.

The limitations of this study include the following: among the 367 patients with ARMs treated in Xinhua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine, only 138 patients were included into the study due to loss of follow-up, and therefore, a comprehensive study could not be conducted; the evaluation of postoperative bowel function was not sufficiently detailed; and the QOL assessment utilized was not comprehensive.

In conclusion, a number of children had fair functional outcomes after anorectoplasty, and that functional outcome and QOL could be improved. The QOL of children after anorectoplasty depends on a number of factors. Various surgical techniques could change functional outcomes. Different scoring systems could also affect these results. Associated malformations, complications and higher level of the ARMs directly lead to negative outcomes. Postoperative functional outcome assessment, anorectal functional training, and psychological support are important for improving QOL.

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### Disclosure of conflict of interest

None.

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