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Can we expect a favorable outcome after surgical treatment for an anorectal malformation?

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SUMMARY

Background: The aim of this study was to retrospectively review the classification, surgical experience, and the functional outcome of anorectal malformations (ARMs) according the type of ARM.
Methods: A total of 311 children (M:F = 200:111) who underwent surgical treatment for ARM between 1990 and 2011 were reviewed. Functional outcomes were evaluated using the Krickenbeck classification. The mean follow-up period was 112.2 ± 76.7 months (range: 36.8–414.9 months).
Results: In the male patients, 90 (45%) had perineal fistulas, 60 (30%) had urethral fistulas, and 7 (3.5%) had rectovesical fistulas. There were 17 cases of ARM without a fistula (8.5%), and we could not determine the type of fistula in 26 boys (13%) because of follow-up losses and death. In the female patients, 34 (30.6%) had perineal fistulas, 71 (64%) had rectovestibular fistulas, and 2 (1.8%) had rectovaginal fistulas. Four patients did not have a fistula (3.6%). For 264 patients, we did anoplasty (121 cases), fistula transposition (14 cases), and posterior sagittal anorectoplasty (PSARP, 129 cases). We found that 224 (84.8%) patients showed voluntary bowel movements. The overall rate for constipation was 30.7% and for soiling was 6.5%. The continence outcome was good for 82.2% of children, fair for 2.7%, and poor for 15.2%. For rectovestibular fistulas, constipation was higher in the perineal operation group, but the continence outcome was similar.
Conclusion: Through a review of 20 years' experience, an accurate diagnosis based on the Krickenbeck classification and operations following the principles of PSARP are crucial to achieve a good functional outcome in children with an ARM.

Anorectal malformation (ARM) is a common congenital disease occurring in 1:5000 births. The development of ARM is believed to be caused by the lack of recanalization during the 9th week of gestation and ectopic positioning of the anal canal opening in the cloaca [1]. However, ARM comprises a wide spectrum of diseases with various presentations and associated anomalies. In 2005, an international meeting was conducted in Krickenbeck to agree on a new simplified classification for ARM. According to that classification, the major clinical groups include perineal fistula, rectourethral fistula (to the prostatic and bulbar urethra), rectovesical fistula, vestibular fistula, cloaca, no fistula, and anal stenosis. Rare/regional variants were classified as pouch colon, rectal atresia/stenosis, rectovaginal fistula, H-type fistula, and others [2]. After the introduction of posterior sagittal anorectoplasty (PSARP) in 1980, the procedure has become the predominant approach to correct ARMs, and their functional outcome has been much improved. Still, constipation and incontinence are major concerns that affect the quality of life of affected patients.

In this study, we retrospectively reviewed the ARM classification, surgical experience, and functional outcomes according the type of ARM in children based on our clinical experience over two decades.

1. Methods

1.1. Subjects

We retrospectively reviewed the medical records of children who were treated for ARM between January 1990 and December 2011 at Asan Medical Center, Seoul, Korea. We identified the type of fistula, the type of surgery, any associated anomalies, surgical complications, and the functional outcomes in each of these patients. We excluded 16 cases of persistent cloaca, 13 cases of Currarino syndrome, 4 cases of H-type fistula, and 2 cases of cloacal exstrophy because of the small number of these cases and a limited follow-up period.

1.2. Diagnosis and surgical method

After diagnosis of an ARM, all patients were evaluated to find associated anomalies. It included abdomino-pelvic ultrasonography, echocardiography, and spine ultrasonography. Magnetic resonance imaging (MRI) for the spine was required if anomalies were detected on

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Table 1
ARM classifications.

	Classsification	No.	%
Male (n = 200)	Perineal fistula	90	45.0
	Rectourethral fistula	63	31.5
	prostatic	5	2.5
	bulbar	58	29
	Rectovesical fistula	4	2
	No fistula	17	8.5
	Unknown	26	13.0
Female (n = 111)	Perineal	34	30.6
	Vestibular	71	64.0
	Rectovaginal	2	1.8
	No fistula	4	3.6

Table 3
Overall functional outcomes according to Krickenbeck classification.

Type of ARM	No. of Cases	VBM		Soiling				Constipation			
		Yes	No	No	Gr 1	Gr 2	Gr 3	No	Gr 1	Gr 2	Gr 3
Perineal	117	112	5	117	0	0	0	100	5	7	5
Rectourethral	64										
Bulbar	59	39	20	48	4	6	1	27	4	8	20
Prostatic	5	4	1	3	1	1	0	1	0	3	1
Vestibular	59	52	7	57	1	1	0	43	3	6	7
Vesical	3	1	2	1	1	0	1	0	0	1	2
No fistula	20	16	4	20	0	0	0	12	0	4	4
Vaginal	1	0	1	1	0	0	0	0	0	0	1
		84.8%	15.2%	93.5%	6.44%			69.3%	30.7%		

ultrasonography. When an ARM with a perineal fistula was noted, we performed an anoplasty within 3 days of birth. Anoplasty comprises cut-back anoplasty and Y-V anoplasty. If there was no meconium passage within 36 h or meconium passage via an external urethral orifice, we performed a three-staged operation, including a colostomy, PSARP, and colostomy closure. The fistula location was identified through distal cologrames via a colostomy. For a rectovestibular fistula there are three surgical options: a perineal operation including anoplasty and fistula transposition in the neonate, a one-stage PSARP after a widening of the rectovestibular fistula in infants, or a three-staged operation including a colostomy, PSARP, and colostomy closure [3].

1.3. Functional outcomes

To evaluate functional outcomes, we used the Krickenbeck classification [2]. We collected data related to the presence of voluntary bowel movements, frequency of soiling, and management of constipation. The mean follow-up period was 112.2 ± 76.7 months (range: 36.8–414.9 months). The patients were divided into three groups according to the age of evaluation: <6 years old, 6–12 years old, and >12 years old. Voluntary bowel movements (VBM) were defined as an urge to defecate, the capacity to verbalize this feeling, and the ability to hold the bowel movement. We defined constipation to be present if the patient had no bowel movement over 48 h. Pena defined overflow pseudoincontinence as a soiling in a constipated patient who, once treated adequately with laxatives, has voluntary bowel movements and stops soiling [4]. We recorded the frequency of soiling except overflow pseudoincontinence.

A good outcome was defined as a voluntary defecation with no/occasional soiling and no/light constipation (controlled with diet or laxatives). A fair outcome indicated voluntary bowel movements with soiling grade 1 or 2 and constipation grade 1 or 2. Poor results included patients without voluntary defecation, with soiling grade 2 or 3, and any degree of constipation [2,5]. The patients who underwent regular enemas were included in the "poor" group.

We described functional outcomes according to the type of fistula. We analyzed categorical variables using the chi-square test and Fisher's exact test with the statistical software SPSS (version 15 for Windows, SPSS Inc., Chicago, IL). Statistical significance was assigned to p-values < 0.05.

Table 2
Morbidity and mortality.

	No.	No.	Cause
Mortality	20	10	Congenital heart disease
	(6.4%)	5	Chronic lung disease
		2	Sepsis
		1	Neurologic anomaly
		1	Acute myeloid leukemia
		1	Smith Lemli Opitz syndrome
Morbidity	29	13	Mucosal prolapse
	(9.3%)		12: rectourethral fistula
			1: no fistula
		5	Recurrent fistula
			4: rectourethralfistula
			1: rectovaginal
		5	Adhesive ileus
			2: rectourethral fistula
			1: rectovesical fistula
			1: vestibular fistula
			1: no fistula
		3	Neoanus stenosis
			2: no fistula
			1: vestibular fistula
		1	Small bowel perforation
			1: no fistula
		1	Colostomy repair leakage
			1: rectourethral fistula
		1	Necrotizing enterocolitis
			1: vestibular fistula

2. Results

2.1. Patient characteristics

A series of 311 children was enrolled in this study, comprising 200 male and 111 female (M:F = 1.8:1) patients. Premature babies composed 13.2% (41) of these patients. Fifty-eight (18.6%) babies had a birth weight <2.5 kg, and 10 (3.2%) babies were <1.5 kg. Associated anomalies were present in 125 (40.2%) patients, and most associated anomalies were cardiovascular (39/125) or genitourinary (28/125). Sacral anomalies were found in 11 (4.1%) patients, and the VATER association of anomalies was present in 18 (5.8%) patients.

The type of ARM in the boys in our cohort included perineal fistulas in 90 (45%), rectourethral fistulas in 63 (31.5%, including 58 bulbar fistulas and 5 prostatic fistulas), and rectovesical fistulas in 4 (2%) patients. There were 17 cases of ARM in males without a fistula (8.5%), and we could not determine the type of fistula in 26 (13%) boys because of deaths and follow-up losses. In female cases, a perineal fistula was present in 34 (30.6%), a rectovestibular fistula was present in 71 (64%) cases, and a rectovaginal fistula was present in 2 (1.8%) patients. Four of the girls in our cohort did not have a fistula (3.6%; Table 1).

There were 20 (6.4%) deaths during the perioperative period. The cause of death was congenital heart disease in 10, chronic lung disease in 5, sepsis in 2, neurologic anomaly in 1, acute myeloid leukemia in 1, and Smith Lemli Opitz syndrome in 1 case, respectively (Table 2). Postoperative complications were related to PSARP in 29 (9.3%) patients, including 13 with mucosal prolapse, 5 with recurrent fistula (including 4 urethral fistulas and 1 vaginal fistula), 5 with adhesive ileus, 3 with anal stenosis, 1 with small bowel perforation, 1 with leakage after the stoma repair, and 1 with necrotizing enterocolitis. There were no complications with the colostomy in any of the children (Table 2).