



# Evaluation of Long-term Results and Stool Incontinence of Patients with Operated Anorectal Malformations

## *Opere Anorektal Malformasyonlu Hastaların Uzun Dönem Sonuçlarının ve Fekal İnkontinanslarının Değerlendirilmesi*

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### ABSTRACT

**Objective:** Anorectal malformations (ARMs) are a group of anomalies with a wide spectrum, from simple types requiring minimal surgery to complex anomalies requiring complex surgical interventions. Constipation and inability to achieve stool continence are the leading problems in the late postoperative period for patients with ARMs.

**Method:** This study aimed to evaluate and compare the long-term results and stool continence status of the patients with ARMs who had undergone a definitive surgery in our clinic between January 1996 and December 2009, according to anal atresia types. The patients were evaluated in terms of anomaly type, additional organ anomaly, surgeries performed, and postoperative complications.

**Results:** A total of 68 patients including 40 (58.82%) male, and 28 (41.18%) female cases who had undergone definitive surgery were examined. Considering all patients, the most common anomaly was anocutaneous fistula detected in 20 patients (29.4%), and the least common one was rectovesical fistula detected in 1 (1.5%) patient. When the definitive surgeries performed on patients with ARM are evaluated according to the type of ARM, most frequently anoplasty (n=20) had been performed. The most common complaint of the patients was constipation (n=20). A statistically significant relationship was found between the type of anomaly, the type of the definitive surgery performed, constipation and stool incontinence ( $p<0.05$ ).

**Conclusion:** Patients should be followed up in the postoperative period for problems such as anal stenosis and constipation, and treatment should be initiated before formation of fecalith in patients with constipation.

**Keywords:** Anorectal malformation, constipation, incontinence

### ÖZ

**Amaç:** Anorektal malformasyonlar (ARM) minimal cerrahi gerektiren basit tipten, kompleks cerrahi girişim gerektiren karmaşık anomalilere kadar geniş bir spektruma sahip anomali grubudur. ARM'li hastaları bekleyen postoperatif geç dönem problemlerin başında konstipasyon ve gaita kontinansının sağlanamaması gelir.

**Yöntem:** Bu çalışma kliniğimizde Ocak 1996-Aralık 2009 yılları arasında definitif operasyonu tamamlanmış olan ARM'li hastaların uzun dönem sonuçlarının ve gaita kontinans durumlarının anal atrezi tiplerine göre değerlendirilip, karşılaştırılması amaç edinilmiştir. Hastalar anomali tipi, ek organ anomalisi, yapılan ameliyat ve postoperatif komplikasyonlar açısından değerlendirilmiştir.

**Bulgular:** Definitif operasyon uygulanan 68 hasta incelendiğinde 40 hastanın erkek (%58,82) ve 28 hastanın ise kız (%41,18) olduğu görülmüştür. Tüm hastalar ele alındığında en çok karşılaşılan anomalinin 20 hastada (%29,4) tespit edilen anokutanöz fistül olup en az karşılaşılan anomali 1 hastada (%1,5) tespit edilen rektovezikal fistül olmuştur. ARM'li hastalara uygulanan definitif operasyonlar ARM tipine göre değerlendirildiğinde, en çok yapılan ameliyat (n=20) anoplasti operasyonudur. Hastalar en çok kabızlıktan şikayet etmekteydi (n=20). Anomali tipi, uygulanan definitif ameliyat ile kabızlık ve gaita kaçırma arasında istatistiksel olarak anlamlı ilişki tespit edilmiştir ( $p<0,05$ ).

**Sonuç:** Hastalar postoperatif dönemde anal darlık ve kabızlık gibi problemler yönüyle takip edilmeli, kabızlık tespit edilen hastalarda dışkı taşlaşması oluşmadan tedavi başlanılmalıdır.

**Anahtar kelimeler:** Anorektal malformasyon, kabızlık, inkontinans

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## INTRODUCTION

Anorectal malformations (ARMs) are seen with a frequency of 1 in 4,000-5,000 live births. A male/female ratio of 1.4-1.6 has been reported<sup>(1-3)</sup>.

The majority of ARMs in boys are of the high type, while most ARMs in girls are of the low type<sup>(3)</sup>.

ARMs are still mainly evaluated as low type and high type. The diagnosis of low-type ARM can be made easily by physical examination, and its treatment gives better results. In the diagnosis of high-type malformations, additional examinations are needed as well as physical examination.

ARM may be suspected by the presence of dilated colon or intraluminal calcifications, or both, during routine prenatal diagnostic ultrasonographic controls of pregnant women, especially after the 25<sup>th</sup> gestational week. Diagnosis of ARM can be done by observing the fecal matter coming from the urethra or perineum in boys and from the perineum and vestibulum in girls in the immediate postpartum period. However in some cases extrusion of fecal matter can not be detected. In this case, as previously reported by Wangenstein and Rice an invertogram film is shot or as later on proposed a "cross- table lateral view" is obtained. In addition, computed tomography, magnetic resonance imaging technique, and fistulography can also be used for diagnosis and typing of ARMs<sup>(4,5)</sup>.

Patients with ARMs have attained near-normal living standards thanks to the latest developments in treatment. Dealing with the problems of these patients who are given a chance to survive after treatment, is at least as necessary as the treatment. Constipation and/or stool incontinence is one of the most critical problems and complaints awaiting patients with anal atresia who underwent definitive surgery. It is necessary to explain to the families the possibility of their children being unable to achieve stool control in the future in a realistic language without disappointing them. Likewise, families should give their children information about bowel control without causing them to have false expectations in the future.

Many additional organ anomalies may accompany high-type ARMs. Their treatment is more challenging with lower chances of continence. Posterior sagittal anorectoplasty technique (PSARP) was introduced by Peña and Devries<sup>(5)</sup> in 1982 and highly successful results started to be obtained. The treatment of ARMs aims to give the patients an opportunity to lead a near-normal

life. However, it is as important as the treatment to fight the problems that the surviving patients are confronting after their definitive treatment is completed. Constipation and/or stool incontinence are the leading problems for these patients. Families should be told about the possibility of their children being unable to control their defecation in the future in a realistic language without causing disappointment. Parents should also be able to provide their children with information about control of their bowel movements to the degree that they can meet their children's expectations in cooperation. This study aimed to evaluate and compare the stool continence of patients with an ARMs whose colostomies were closed.

## MATERIALS and METHOD

The ethical approval was obtained from Atatürk University Faculty of Medicine Clinical Research Ethics Committee (number: 78, date: 17.04.2009). After obtaining ethical committee approval, the archive records of the patients with an ARM hospitalized in our clinic, between January 1996 and December 2009, according to the type of atresia, were reviewed retrospectively.

The records of patients with ARMs were accessed, and then these patients were contacted by phone and invited to the hospital for re-evaluation. Data related to gender, age, anomaly type, additional organ anomaly, operation date, operation age, postoperative complaints, postoperative treatment methods, postoperative complications, dilatation, constipation history, incontinence, psychosocial problems, and additional operations of 68 patients who agreed to participate in the study were reviewed.

### Statistical Analysis

The obtained data were entered into the SPSS 20.0 program and analyzed using the chi-square test.

## RESULTS

The data of 68 patients diagnosed with ARM and had definitive surgery in our hospital were examined, and only 51 of these patients were evaluated in our clinic for long-term complications. A total of 68 patients with ARM including 40 (58.82%) male, and 28 (41.18%) female cases with a median age of 7.33 years (range: 2-14 years) had undergone definitive surgery with a male/female ratio of 1.42.

The most common anomaly was anocutaneous fistula (ACF) in 20 (29.4%), and rectovestibular fistula (RVF) in 16

patients (23.5%) followed by rectourethral fistula (RUF) and rectoperineal fistula in 13 (19.1%) patients. The least common anomaly was rectovesical fistula in one (1.5%) and perineal groove in another patient (1.5%) (Table 1).

ACF, which was present in 16 patients (23.5%), was the most common type of ARM in men, followed by RUF in 13 patients (19.1%). The most common type of ARM in girls was RVF in 16 (23.5%) patients followed by rectoperineal fistula in 6 patients.

Any comorbidity was not found in 50 (73.53%) patients while additional organ anomalies were detected in 18 (26.46%) patients. Most frequently urinary system anomalies were seen (n=9) followed by cardiac (n=7), and extremity (n=2) anomalies

Urinary system anomalies were seen in 9 patients, including ureteropelvic junction stenosis (n=2), vesicourethral reflux (n=2), horseshoe kidney with accompanying hydronephrosis (n=1), hydronephrosis (n=1), nephrolithiasis (n=1), hypospadias (n=1), horseshoe kidney with accompanying vesicourethral reflux (n=1), and an isolated case of renal agenesis. Seven patients with cardiac anomalies were detected in indicated number of patients including atrial septal defect (ASD), patent ductus arteriosus (PDA), and patent foramen ovale (PFO) (n=1), ventricular septal defect (VSD), PFO (n=1), PDA, PFO (n=1), PDA, ASD (n=1), and ASD, VSD, PDA (n=1). One of the two patients with extremity anomalies had wrist and finger deformities, and the other one had bilateral club foot and duodenal atresia.

A definitive surgery was performed in a single session without opening a colostomy in 41 patients, while colostomies were created in 27 patients. The indicated number of patients had also undergone surgical interventions due to additional pathologies including ureteroneocystostomy performed with the diagnosis of vesicoureteric reflux (n=2), duodenojejunostomy due to

duodenal atresia (n=1), excision of Meckel’s diverticulum (n=1), anocutaneous junction revision due to prolapse of the anal mucosa (n=1), and stenosis of the sigmoid colon was dilated in one patient. In addition, two patients had an operation due to extremity anomaly (club foot and wrist deformity).

When the definitive operations performed according to the ARM type are evaluated, the most common surgical interventions was anoplasty in 20 (29.4%) patients, followed by anterior sagittal anorectoplasty (ASARP) in 15 (22.1%) and PSARP in 14 patients (20.6%)

Postoperative complications were present in 4 (5.8%) patients including recurrent RUF (n=1), anal stenosis (n=1), prolapse of the anal mucosa (n=1), and acute abdomen after colostomy repair (n=1).

Only 51 patients reached our clinic and were evaluated with face-to-face interviews. Constipation was detected in 22 (43.1%) of these 51 patients. In terms of stool continence, 36 patients (70.6%) had relevant complaints and 15 patients (29.4%) had stool incontinence. A statistically significant relationship was found between the type of anomaly and stool incontinence (p<0.05) (Table 2). There was no significant gender difference among children with stool incontinence (8 boys and 7 girls). No significant relationship was found between age and achievement of continence (p>0.05). Indicated number of ARM experienced stool control problems including cases with RUF (n=7), RVF (n=6), ACF (n=1) and rectoperineal fistula (n=1).

Colostomy was performed in 13 of 15 patients with stool continence problems, and colostomy was not performed in 2 patients. The relationship between stool incontinence and colostomy was statistically significant (p<0.01). Patients with ARM who had a colostomy were evaluated in terms of constipation, and any statistically significant correlation could not be found between

**Table 1. Distribution of ARM by gender**

		Types of ARM									Total
		ACF	RUF	PF	AEA	RPF	RVF	RVzF	Pgrv		
Gender	Boy	n	16	13	2	1	7	0	one	0	40
		%	23.5%	19.1%	2.9%	1.5%	10.3%	0.0%	1.5%	0.0%	58.8%
	Girl	n	4	0	0	1	6	16	0	1	28
		%	5.9%	0.0%	0.0%	1.5%	8.8%	23.5%	0.0%	1.5%	41.2%
Total		n	20	13	2	2	13	16	1	1	68
		%	29.4%	19.1%	2.9%	2.9%	19.1%	23.5%	1.5%	1.5%	100.0%

ARM: Anorectal malformations, ACF: Anocutaneous fistula, RUF: Rectourethral fistula, AVF: Anovestibular fistula, RPF: Rectoperineal fistula, RVF: Rectovestibular fistula, RVzF: Rectovesical fistula, Pgrv: Perineal groove, PF: Perineal fistula, AEA: Anterior ectopic anus

colostomy and constipation ( $p > 0.05$ ). Also, stool incontinence was detected in 11 of 24 patients with constipation, and constipation was detected in 11 of 15 patients with stool incontinence. Also a statistically significant correlation was found between stool incontinence and constipation ( $p < 0.05$ ).

## DISCUSSION

ARMs are congenital anomalies presenting with very different clinical manifestations. They constitute the majority of pediatric gastrointestinal system anomalies. They are seen in a wide spectrum, from near-normal appearance to complex ARMs and syndromes. When all ARMs are evaluated collectively, the incidence of one or more additional anomalies varies between 25-75%<sup>(3,6)</sup>. Although many theories have been put forward related to the formation of ARM, its exact pathogenetic mechanism has not been revealed yet

The type of ARM present determines the long-term results of the patients, as well as the surgical technique and skill applied in the definitive surgery. If there is a malformation in the group with poor prognosis, these children will inevitably experience problems with stool control.

Although incontinence due to sphincter insufficiency, which is seen as the most critical problem after surgical treatment of ARM, is still a major problem, problems such as constipation, overflow incontinence due to constipation, and bowel motility disorders have come to the fore despite the advantages of surgical methods developed and applied over time<sup>(6)</sup>.

Although significant advances have been made in understanding and treating ARMs, problems such as constipation, panty-soiling, and incontinence after surgery persist and can be seen at a rate of 30-70%. More than half of the patients experience bowel

disorders that cause physical, psychological, and social problems<sup>(7,8)</sup>.

Contrary to the literature data, in our series, most frequently low-type malformations were detected in boys, and intermediate-type RVFs in girls. Most common low-type anomaly in male patients was ACF, followed by a RUF. Among the rectourinary fistulas, rectobulbar fistula was detected. Rectoprostatic fistula was detected in only one patient. These findings are also contrary to the literature data<sup>(9,10)</sup>. In female patients, the most common anomaly type was RVF consistent with statistical data cited in the literature<sup>(11)</sup>.

Accompanying additional organ anomalies are more frequently seen in cases with high-type anomalies<sup>(3,12)</sup>. Comorbidities seen in ARM patients are genitourinary (49%), musculoskeletal (43%), craniofacial (34%), cardiovascular (27%), and gastrointestinal anomalies (18%)<sup>(12,13)</sup>. Most frequently urogenital system anomalies were seen followed by cardiac anomalies<sup>(3)</sup>. Apart from these, extremity anomalies were also detected. When these findings are compared with the literature data, vertebral (especially sacral) and urinary anomalies are seen frequently, and in our country, urinary anomalies ranked on top followed by cardiac anomalies.

ASARP was performed mostly in female, and anoplasty in male patients. Compared with the literature, Peña and Hong<sup>(11)</sup> recommends opening a colostomy in all patients with high-type ARM and PSARP as definitive surgical intervention without gender discrimination. In our series, on the other hand, our colostomy rates are in line with Peña's recommendation, and the difference in our preferences for definitive surgery is remarkable.

Peña and Hong<sup>(11)</sup> advocated that all ARMs, except cloacal malformations, can be corrected using the PSARP method. However, in our clinical series, ASARP was the primary surgical technique of choice in girls

**Table 2. Comparison of stool incontinence and anomaly types**

			Types of ARM							Total
			ACF	RUF	PF	AEA	RPF	RVF	Pgrv	
Stool incontinence	Yes	n	one	7	0	0	one	6	0	15
		%	2.0%	13.7%	0.0%	0.0%	2.0%	11.8%	0.0%	29.4%
	No	n	13	5	2	2	7	6	1	36
		%	25.5%	9.8%	3.9%	3.9%	13.7%	11.8%	2.0%	70.6%
Total	n	14	12	2	2	8	12	1	51	
	%	27.5%	23.5%	3.9%	3.9%	15.7%	23.5%	2.0%	100.0%	

ARM: Anorectal malformations, ACF: Anocutaneous fistula, RUF: Rectourethral fistula, AVF: Anovestibular fistula, RPF: Rectoperineal fistula, RVF: Rectovestibular fistula, Pgrv: Perineal groove, PF: Perineal fistula, AEA: Anterior ectopic anus

with anovestibular fistula. One of the interesting results of our clinical series is that no cloacal malformation was encountered in girls. This finding is inconsistent with the literature data.

The patients were evaluated for stool control after the definitive surgery, based on the disease history, physical examination findings, and sphincter activities detected during the definitive operations. The anal tone of the patients and the presence of anal stenosis were checked with digital rectal examination. Sphincter activities were evaluated by looking at the contraction of sphincter structures on digital rectal examination. No patient with weak anal tone was detected in this examination.

The complaints of the patients and whether they received treatment for these complaints or those with these complaints faced psychosocial problems were questioned. Only 15 of the 51 evaluated patients had stained underwear and were considered in the group with stool incontinence. All but one of the 15 patients who stained underwear had a complaint of constipation, and stool incontinence was in the form of overflow incontinence. A statistically significant relationship was found between constipation and stool incontinence.

Colostomy was performed in 13 of 15 patients with stool incontinence, and these patients had high-type ARMs. Two patients were operated on in a single stage and had no colostomy history. A statistically significant correlation was detected between anomaly type, operation technique and stool incontinence, and also between colostomy opening and stool incontinence.

Constipation or staining underwear after bowel movements were not observed only in patients with high-type malformations but also such complaints were also observed in cases with low-type malformations. Almost none of the patients who had undergone definitive surgeries attended control visits at regular intervals in the long term. Patients with stool incontinence did not come for control after definitive surgery, even though they complained of staining underwear and constipation, and very few patients were treated for constipation.

The reason why the patients did not come to the control was due to the fact that they did not experience any social, psychological, or environmental problems. Half of the patients (50%) were in school, and none had social, psychological, and school-related problems. Medical treatment for constipation was started, and

regular follow-ups were recommended for these patients.

In the long term, bowel training programs, enemas, or even secondary surgeries may be required in patients with ARMs, especially in patients with high-type malformations, for problems such as stool incontinence and fecal contamination<sup>(11,14)</sup>.

None of the patients had enough complaints to restrict their social life. The patients did not need a second operation after the definitive surgeries performed and did not receive any bowel training program or supportive medical treatment. Indeed, they continued their lives without experiencing psychosocial problems.

The type of anomaly predicts the long-term results of the patients with ARM and the surgery performed. No matter how successful the operation is, it should be known that patients who belong to the poor prognostic group or in other words patients who have high-type ARM may have a low chance of achieving bowel control. Patients with low-type malformations are more fortunate in this regard.

In our evaluations, bowel control after definitive surgeries performed for low-type ARMs was satisfactory in more than 90% of patients, but it was not desirable in cases with high and intermediate-type ARMs.

There was no significant difference between our results and the results given in the literature, in terms of the sphincter examination performed with a nerve stimulator during the operation and digital rectal examination of the anal sphincter we performed in the postoperative period, as well as the Krickenbeck criteria used in the evaluation of the treatment results<sup>(15)</sup>.

### Study Limitations

This study has some limitations including its retrospective design. Besides, the patients were not evaluated using a nerve stimulator under anesthesia. Future studies can be planned prospectively to fill these deficiencies.

### CONCLUSION

Since the problems detected in the patients are mainly constipation and overflow incontinence due to constipation, patients should be followed up for problems such as anal stenosis and constipation in the postoperative period, and families should be informed about getting the patients accustomed to defecation at regular intervals and at certain times. In patients

with constipation, treatment should be started before formation of fecaliths, and if necessary, the bowels should be emptied with enemas, and the patients should be fed with regular and appropriate meals to regulate their bowel movements.

### Ethics

**Ethics Committee Approval:** The ethical approval was obtained from Atatürk University Faculty of Medicine Clinical Research Ethics Committee (number: 78, date: 17.04.2009).

**Informed Consent:** Retrospective study.

**Peer-review:** Externally and internally peer reviewed.

### Author Contributions

Surgical and Medical Practices: B.F., A.B.S., Concept: B.F., A.B.S., Design: B.F., A.B.S., Data Collection or Processing: B.F., A.B.S., Analysis or Interpretation: B.F., A.B.S., Literature Search: B.F., A.B.S., Writing: B.F., A.B.S.

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