Prospective follow up of children with anorectal malformations: Our experience with children up to 10 years-old

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Abstract

Introduction: the aim of our study was to perform a longitudinal follow-up in patients with anorectal malformations (ARMs) in order to determine the anorectal function problems and help to solve them.

Materials and methods: This study included 262 patients with ARM who were operated between 2006 until 2013. Children whose definitive reconstruction was performed at another hospital and underwent reoperation in our center were excluded. Patients who expired or did not come for follow-up were not included in the study either. Bowel function was prospectively assessed by using a questionnaire answered by the children's parents. supplementary bowel management with enemas, laxatives and the use of diapers were documented as well.

Results: Interviews were completed in 242 children, ages ranging from 3 to 10 years. More than thirty seven (37.7%) patients had constipation; 32.5% Grade 1 (controllable by altering in diet), 54.3% Grade 2 (requiring laxatives) and 13.2% Grade 3 (opposed to stool softeners and diet). Exactly 18.6% of patients had fecal soiling; 31.5% Grade 1: infrequently (once or twice for every week), 24% Grade 2: daily (no community difficulty) and 44.5% Grade 3: Constant (social problem).

Conclusion: In the present study several bowel function problems were found in ARM children. Physicians who perform the definitive operation on anorectal malformations should not loose follow up of patients as they grow up because they may develope great problems during their adolescence.

Keywords

- Anorectal malformation
- Postoperative complication
- Bowel functional outcome
- Fecal incontinence

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Introduction

Anorectal malformations are moderately frequent inherited anomalies. Constipation, soiling, and fecal incontinence influence Quality of life (QOL) in these group of patients. Near-to-normal QoL is an essential goal in the management and outcome of patients with anorectal malformations. The purpose of this review was to analyze and estimate the rate of bowel function problems in them.

Materials and methods

In this qualitative study we investigated the clinical queries and quality of life in patients with ARM operated in our center between 2006 until 2013. The medical records of 262 patients were reviewed. All patients were older than 3 years. This study was approved by our hospital institutional review board. The related sheet was filled with medical data and completed with information given by the children's parents in relation to the children's bowel function. Constipation was considered as having two or less defecations every week and/or history of throbbing or firm dification. Fecal soiling was defined as passing of changeable quantities of stool to the underwear one or more times a day. Patients were based on the International Krickenbeck agreement of classification and outcome in ARM (see **Tables 1**).[20]

Table 1: International classification (Krickenbeck) for postoperative results

International classification (Krickenbeck) for postoperative results[19]	
1. Voluntary bowel movements Feeling of urge, capacity to verbalize, hold the bowel movement	Yes/no
2. Soiling	Yes/no
Grade I	infrequently (once or twice for every week)
Grade II	daily, no community difficulty
Grade III	Constant, social problem
3. Constipation	Yes/no
Grade I	controllable by altering in diet
Grade II	Requires laxative
Grade III	opposed to stool softeners and diet

Results

From 2006 until 2013, 262 patients with ARM (47.3% females and 52.7% males) were operated in our center. Twenty cases were excluded because they were lost to follow up after discharge or expired. Mean age for definitive reconstruction was 5.15± 2.3 months (Range 2 days – 30 month). In this study the youngest patient was 3 years-old and the oldest was 10 years old. **Table 2** shows the classification of the malformations. The most common type was perineal fistula. More than half of the patients (52.2%) had associated malformations such as cardiac, renal, vertebral, genitalia and limb malformations **Table 3**. Five children had clear malformations on the spinal cord: an anterior lipomyelomeningocele in two patients and tethering of the cord in three. Definitive reconstruction was done by posterior sagittal anorectoplasty (PSARP) in 82%, anterior sagittal anorectoplasty in 2.4%, vaginal replacement with sigmoid colon in 1.2 %, cut back procedure in 3.2%, PSARP and abdominal approach in 7%, pull-through of proximal pouch of colostomy in 4.2%. Around half of the definitive procedures (53.7%) were done with magnification. Fifty-two percent of patients had short and long term complications. Short complications included: Wound infections (2%), wound dehiscence (1.2%), colostomy prolapsed (1.8%) and urethral damage (0.8%). Long term complications are depicted in **Table 4**. About 8.9% of patients suffered from mucosal prolapse and 35% had mega sigmoid. Stenosis of the neo-anus was seen in 10.7%, permanent neurogenic bladder dysfunction was found in 1.6%, recurrence of the urinary fistula in 1.6% of males and rectovaginal fistula in 1.2% of females. Ectopic neo-anus was seen in 3.3%. Thirty seven percent (37.7%) of children had constipation. **Table 5** shows constipation grades according to the Krickenbeck criteria. In our study we recorded

32.5% Grade 1, 54.3% Grade 2 and 13.2% Grade 3. Among patients older than 5 years 18.6% had fecal soiling, 31.5% Grade 1, 24% Grade 2 and 44.5% Grade 3, **Table 6**. Malone procedure was done in 1.6% of patients to correct fecal soiling.

Table 2: classification of the malformation

Classification of Anorectal Malformations	percentage
Perineal fistula	32.7%
Rectourethral fistula	11.6%
Vestibular fistula	22.7%
Persistent cloaca	2.5%
Imperforate anus without fistula	21.1%
Complex defects	0.8%
Rectal atresia	0.8%
Rectobladder neck fistula	7%
Rectovaginal fistula	0.8

Table 3: Anomalies Associated with anorectal malformations

Associated malformations	prcentage
Cardiovascular anomalies	21.7%
Tracheoesophageal abnormalities	5.1%
Symptomatic Lumbosacral anomalies	10%
Renal	13.9%
Hirschsprung disease	0.8
Hypospadias	4.8%
Limb	% 4.5
Down syndrome	1.2%

Table 4: Long-term complications

Long term complications	percentage
Stenosis of the neo-anus	10.7%
Mucosal prolapse	8.9%
Megasigmoid/megacolon	35%
Neurogenic bladder dysfunction	1.6%
Stenosis of the urethra	0.4%
Recurrence of the urinary fistula	1.6%
Ectopic neo-anus	3.3%
Constipation	37.7%
Fecal soiling	18.6%
Rectovaginal fistula	1.2%

Table 5: Constipation in patients according to Krickenbeck criteria

International classification Krickenbeck) for post-operativeresults[20]	
Constipation	37.7% of patients
Grade I: Controllable by altering diet	32.5%
Grade II: Requires laxative	54.3%
Grade III: Opposed to stool softeners and diet	13.2%

Table 6: Fecal soiling in patients according to Krickenbeck criteria

Table 1 International classification (Krickenbeck) for postoperative results [20]	
Soiling (children older than 5 years-old) total	18.6%
Grade I: Infrequently (once or twice for every week)	31.5%
Grade II: Daily, no community difficulty	24%
Grade III: Continuous, community difficulty	44.5%

Discussion

More complications are seen in high anorectal anomalies because of the need of multi-stage operations for correction. After the definitive procedure, anal dilatation is necessary to prevent stenosis of the neo-anus. However, and despite the absence of rectal stricture, several grades of dismotility are present. These cases require interventions, such as regular enemas, washouts, or antegrade colonic enemas for constipation or fecal incontinence control.[1]

The most frequent defect in males is imperforate anus with a rectourethral fistula. In females, it is the rectovestibular fistula. Imperforate anus without a fistula is rare, occurring in about 5% of the entire group of malformations, and is associated with Down syndrome.[1]

Constipation, soiling and fecal incontinence are common problems following surgery for anorectal malformation that may influenceon quality of

life (QoL). Quality of life includes physical, psychological and social domain.[3]

Borg[4] studied 41 patients with ARM, excluding perineal fistula. They assessed bowel function at ages 5, 10 and 15 years. A group of 52 healthy children was utilized as control. With age a successive development in soiling and constipation in patient group was observed. Continence was attained earlier in girls than boys (at 10 years: girls 80%, boys 36%). They reported psychosocial co-morbidity, neuropsychiatric diseases, developmental disorders and megarecto sigmoid as risk factors for poor functional results. In our study, the eldest patient was 10 years-old and our follow-up will have to continue for a long time in order to get proper information.

Witvliet et al[5] reviewed thirty articles in relation to quality of life in children with imperforate anus. Around 83% of the reports had not used confirmed QoL sheets. QoL were frequently bottom on functional outcomes, while physical, psychological, and social and environment problems were not considered. Our study focused only on surgical complications and bowel function outcome. Schmidt[6] investigated on 55 patients with history of anorectal malformation with ages ranging from 18 to 56 years. Cases of mucosal prolapse, mega sigmoid and stenosis were observed more frequently than in our study. They also found higher rates of bladder dysfunction and 70% of patients needed a reoperation. These reports show challenges in treatment of adults with history of ARM. Rectal prolapse is common in patients with week sphincters. To keep away from this complication, it is better to fix the posterior rectal wall to the posterior rim of the muscle complex and establish anoplasty under minor tension.[7] Many factors influence on fecal continence. ARM children with neurogenic bladder dysfunction (NBD) and spinalcord malformations have the most horrible outcome regarding both bladder and intestinal function.[8] As we know, condition of the sacrum is a main factor of functional outcome in these children.[9] There are different reports about prognosis of boys with prostatic and bulbar urethral fistula.[10]

Alberto Pena[11] investigated 1806 patients with imperforate anus, 212 were reoperated on after an unsuccessful surgery at a different hospital. Complications led to reoperation were narrowing or acquired atresia of the rectum, aberrant rectum, repeated urinary fistula, continual urogenital sinus, rectal prolapse, narrowing of the vagina, stricture of the urethra, and persistent cloaca. In our study around 20% of patients had to be reoperated because of similar complications. Urodynamics should be done in the majority of cases of cloaca malformation. A. E. Archibong[12] reported fecal incontinence (13%), anal stenosis (11.1%), constipation (7.4%) and colostomy prolapse (5.6%) in their study. Constipation is one of most common long term co morbidities of this malformation. In our present study on 242 patients, 37.7% had some degree of constipation. Management of constipation can improve growth and development of these patients. Chao and coworkers[13] reported that healthy children with sufficient handling of functional constipation gainedmore height and weight than the non-responsive constipated children.[13]

Eberhard Schmiedeke[14] assessed 297 children with ARM. Total continence was seen in 27%, perineal fistula in 40%,rectourethral/vesical in 10%, vestibular in 24%, cloaca in 0%. In our study 18.6% of patients older than 5 years had fecal soiling, Naomi Iwai et al[15] studied 29 cases with ARM, with age between 20 to 40 year. approximate 30% of adult patients with high or intermediate type anomalies had a number of troubles in bowel function. Fecal soiling troubled their job related life. A majority of them had normal urinary and sexual functions if they did not have related genitourinary anomalies.[16] Konuma et al[16] accounted that sexual troubles were widespread in male cases with high or intermediate- kind, particularly with sacral problems. For adolescents with anorectal malformation, the transition into adulthood is particularly difficult.[17, 18] We propose future widespread research. Many factors explain the outcome of bowel function in children with anorectal malformations, but criteria used to assess long-term outcome have been fairly variable. Pediatric surgeons who do the definitive operation on imperforate anus lose contact with patients as they become adults .These patients have many troubles in adolescence including epididymitis, sexual function and social problems. Helpful mind should focus on growth, dietary management, and motor growth of these children.

Conclusion

In this study ARM patients demonstrated refractory constipation and significant fecal incontinenccy. Parents should be convinced that follow-up is essential. Long term assessment by a multidisciplinary team, including a pediatric surgeon, a pediatrician, a physical therapist, a dietician, psychologist, nephrologist, and adult urologist is advocated throughout childhood and adolescence. In this way, favorable growth and development can be attained.

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