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Bowel symptoms in children with anorectal malformation – a follow-up with a gender and age perspective



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ABSTRACT

Background: Gender specific outcome for children with anorectal malformations (ARM) is rarely reported although it is important for medical care and in parent counseling.

Purpose: To assess bowel function according to the Krackenbeck system in relation to ARM-subtype, gender and age.

Method: All children born with ARM in 1998–2008 and referred to two centers in two different countries were followed up. The bowel function in 50 girls and 71 boys, median age 8 years, was analyzed.

Results: Among those with a perineal fistula, incontinence occurred in 42% of the females and in 10% of the males ($p = 0.005$) whereas constipation occurred in 62% of the females and 35% of the males ($p < 0.001$). No bowel symptoms differed between the females with perineal and vestibular fistulas ($p > 0.3$ for every symptom). Sacral malformations were associated with incontinence only in males with rectourethral fistulas. Constipation among the males differed between the age groups: 58% versus 26% ($p = 0.013$). Bowel symptoms did not change with age among the females.

Conclusion: Gender differences in outcome for children with ARM must be considered. Males with perineal fistulas had less incontinence and constipation than the females with perineal fistulas. The females with perineal and vestibular fistulas had similar outcomes.

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Knowledge about the prognosis for bowel control in a newborn child with an anorectal malformation (ARM) is of great concern for the parents and important for planning the child's future medical care. Bowel function in children with ARM operated on with posterior sagittal anorectal plasty (PSARP) [1] has been described in many ways using different classification and follow-up systems [2]. This diversity has made it difficult to report the true outcome after PSARP and to compare results in different patient populations. Therefore, in 2005 the Krackenbeck classification system of ARM sub-groups and postoperative results was established [3]. This way of classifying the subtypes and outcome in children with ARM has been used in some recent reports [4–10], and facilitated prediction of the outcome and comparisons between groups. So far the subtype of ARM according to

the Krackenbeck classification seems to be one of the most important factors for predicting outcome after PSARP for a single child [4,6,7,9,10]. Interestingly, none of the previous reports have analyzed gender-specific outcome.

1. Aim

The main aim of the study was to describe bowel function according to the Krackenbeck follow-up system separately for females and males 4–12 years old with ARM. The secondary aim was to evaluate if fecal incontinence and constipation were less frequent in the older age group.

2. Materials and method

2.1. Children with anorectal malformation

All newborn children with ARM referred to two tertiary centers for pediatric surgery in two countries during the periods from January 1998 and January 2000, respectively, until December 2008 were included in the study. The eligible populations in the two countries

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Table 1

International classification Krickbeck postoperative results for anorectal malformation.

1. Voluntary bowel movements Feeling of urge, capacity to verbalize, hold the bowel movements	Yes/No
2. Soiling Grade 1 Grade 2 Grade 3	No Occasionally (1–2/week) Every day, no social problem Constant, social problem
3. Constipation Grade 1 Grade 2 Grade 3	No Manageable by changes in diet Requires laxative Resistant to laxative and diet

have similar socio-economic conditions with free health care for the inhabitants. There were always two surgeons performing the operations, and at least one of them was an experienced pediatric colorectal surgeon. In total, six colorectal pediatric surgeons were responsible for the operations included here.

The results were collected when the children were between 4 and 12 years of age. Excluded from the study were those with anal stenosis, rectal atresia and cloaca. Those operated on at other centers and those who had migrated were excluded. The studied group finally consisted of 60 and 77 children, respectively, from the two centers. These 137 children, 56 females and 81 males, fulfilled the criteria for inclusion. The children with ARM were subtyped according to the Krickbeck classification suggested by the International Conference for the Development of Standards for the treatment of Anorectal Malformations in 2005 [3].

2.2. Operation methods and follow-up

Both centers operate according to the PSARP-procedure, described by Dr. Pena [1]. The anal sphincter complex is defined by electromyostimulation and the mobilized rectum is placed in the center of

the sphincter complex. A diverting colostomy is used for males with rectourinary fistulas or no fistula, and for some of the females with vestibular fistulas and those with no fistula.

In two females and three males with perineal fistulas the skin incision was performed as a Y-V-plasty. Another two males with perineal fistulas were operated on with cut back where the centre of the anal channel was reached by an incision through the fistula without dividing the sphincter, followed by dilatations of the fistula.

All the children were examined under general anesthesia approximately two weeks after the operation, and then a dilatation program was started if needed. The majority of the children underwent regular dilatations.

The children were examined for associated anomalies with x-ray and ultrasound of the sacrum, and the urinary tract was examined with ultrasound and voiding cystourethrography. If there were signs of tethered cord or neurogenic bladder a MRI and/or urodynamic investigation was performed. Echocardiography of the heart was routinely performed.

2.3. Method

The study is a clinical follow-up and is both descriptive and comparative. Information about the subtypes of ARM, concomitant malformations and symptoms was collected from the regular annual follow-up of the patients. The registration of the bowel symptoms includes consequent registration in the medical records and in a prospectively maintained database, using the Krickbeck post-operative follow-up scheme [3] (Table 1). As suggested in the original article the outcome was measured before bowel management was introduced. Severe incontinence was defined as Krickbeck grade 2 and 3.

2.4. Statistical considerations

The exact Kruskal–Wallis test was used for ranks and the generalized Fisher's exact test was used for dichotomous outcomes

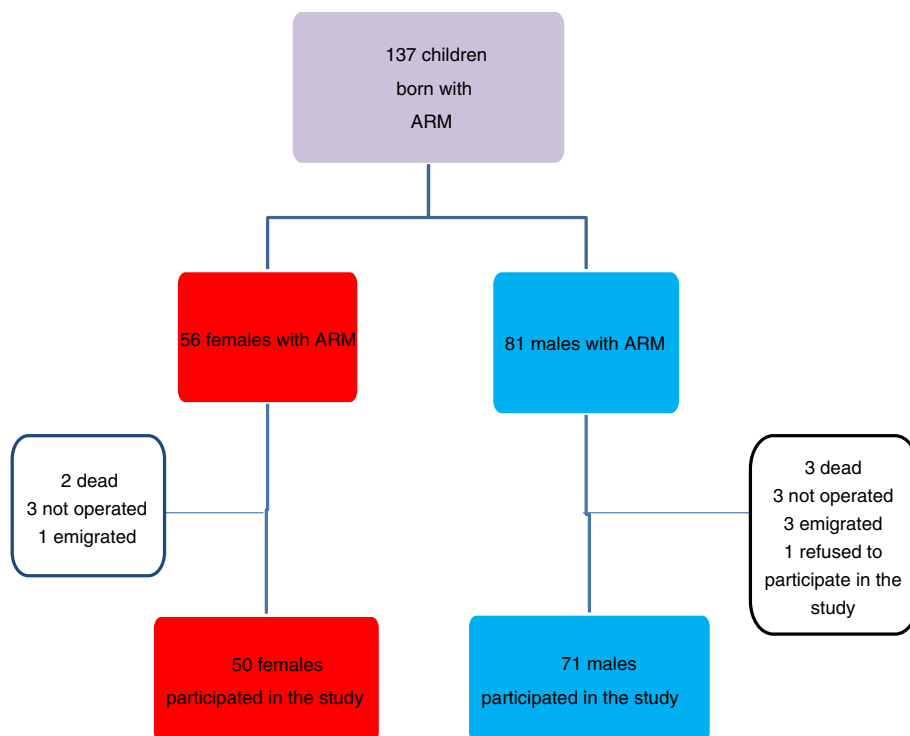


Fig. 1. Consort table illustrating the included children with anorectal malformation (ARM) and the reasons for drop out. n = number.

Table 2

Number (n) and per cent (%) of the children with anorectal malformation (ARM) and the distribution in the age groups 4–7 and 8–12 years, respectively.

Krackenbeck classification of the subtypes of ARM studied in this report	Females n 50			Males n 71			The distribution of the ARM-subtypes n 121 (%)
	All females			All males			
		4–7 years old n 21	8–12 years old n 29		4–7 years old n 24	8–12 years old n 47	
Perineal fistula females	24	10	14				20%
Vestibular fistula	22	9	13				18%
No fistula females	3	1	2				2.5%
Vaginal fistula	1	1	0				1%
Perineal fistula males				32	11	21	26%
Recto-urethral fistula (recto-bulbar and prostatic fistula)				29	11	18	24%
Recto-vesicular fistula				3	0	3	2.5%
No fistula males				7	2	5	6%

The distribution of the different subtypes of ARM in both age groups in both gender was similar ($p > 0.3$ for each group, Fisher's Exact Probability Test, two tailed).**Table 3**

Number (n) and (%) associated anomalies in the cohort of 121 children with anorectal malformation.

Associated malformations, anomalies and disability	Females Total	Females Perineal fistula	Females Vestibular fistula	Females Vaginal fistula	Females No fistula	Males Total	Males Perineal fistula	Males Recto-urethral fistula	Males Recto-vesicular fistula	Males No fistula
n	50	24	22	1	3	71	32	29	3	7
No concomitant malformation or disability	23 (46)	16 (67)	7 (32)	0	0	22 (31)	15 (47)	7 (24)	0	0
Syndrome, neurological disability	8 (16)	2 (8)	2 (9)	1	3 (100)	5 (7)	2 (6)	2 (7)	0	1
Sacral malformation	14 (28)	4 (17)	10 (45)	0	0	16 (23)	3 (9)	9 (31)	2 (67)	6 (86)
Cardiac malformation	12 (24)	4 (17)	5 (23)	1	2 (67)	10 (14)	1 (3)	8 (28)	0	1
Urinary tract anomaly, vesicourethral reflux, neurogenic bladder	20 (40)	4 (17)	13 (60)	1	2	27 (38)	8 (25)	14 (48)	3 (33)	2 (21)
Other malformations (genital, hypospadias, coloboma, ear-anomaly, cleft-lip-palate, esophageal atresia, tracheoesophageal fistula, skeletal malformation)	11	4	6	0	1	18	12	6	0	0

between all groups. If statistical difference was found, the results were analyzed with post hoc tests of joint ranks for ranks of symptoms or with the relevant 2-by-2 subtable Fisher's exact test when comparing binary symptoms. P-values are described between <0.3 and >0.001 . A p-value < 0.05 was considered significant. All statistical computations were performed by a statistician using the computer program R version 2.15.2 [11]. Multiple comparisons were adjusted using the false discovery rate procedure.

2.5. Ethical considerations

The regional research ethics committee approved the study (registration number 2010/49) for one center and the institutional board at the hospital approved it at the other center. The data are presented in such a way that it is impossible to identify any single patient.

3. Results

Six females and 10 males dropped out from the study (Fig. 1). The studied group finally consisted of 121 children: 50 females and 71 males, each group with a median age of 8 years (4–12) (Fig. 1). The distribution between the different subgroups of ARM according to the Krackenbeck classification, gender and age group is shown in Table 2. The distribution of the ARM subtypes in the young and old age groups did not differ significantly ($p > 0.3$) for any of the largest subgroups (Table 2).

In total reoperations were performed in three females and two males. All had perineal fistulas.

The outcomes for the children with perineal fistulas operated on with Y-V-plasty (2 females and 3 males) and cut back (2 males) were gender-wise statistically compared with those operated on with PSARP. No statistical differences were found between the

Table 4

The frequency of voluntary bowel movements (feeling of urge, ability to verbalize and hold back) in 121 children with anorectal malformation (ARM). n = number (%).

Voluntary Bowel Movements	Females Total	Females Perineal fistula	Females Vestibular fistula	Females Vaginal fistula	Females No fistula	Males Total	Males Perineal fistula	Males Recto-urethral fistula	Males Recto-vesicular fistula	Males No fistula
n	50	24	22	1	3	71	32	29	3	7
Yes	35 (70)	19 (79)	16 (73)	0	0	41 (58)	29 (91)	10 (34)	0	2 (29)
No	15 (30)	5 (21)	6 (27)	1	3	30 (42)	3 (9)	19 (66)	3	5 (71)

Statistical analyses: Comparisons of the frequency of voluntary bowel movements between.

a) the total group of females and males, respectively ($p = 0.1864$).b) the females with perineal fistula and the rest of the females ($p = 0.224$).c) the males with perineal fistula and the rest of the males ($p < 0.0001$).

Gender specific statistical analyses regarding the ARM-subtypes are illustrated in Fig. 3.

* Fisher's Exact Probability Test, two-tailed.

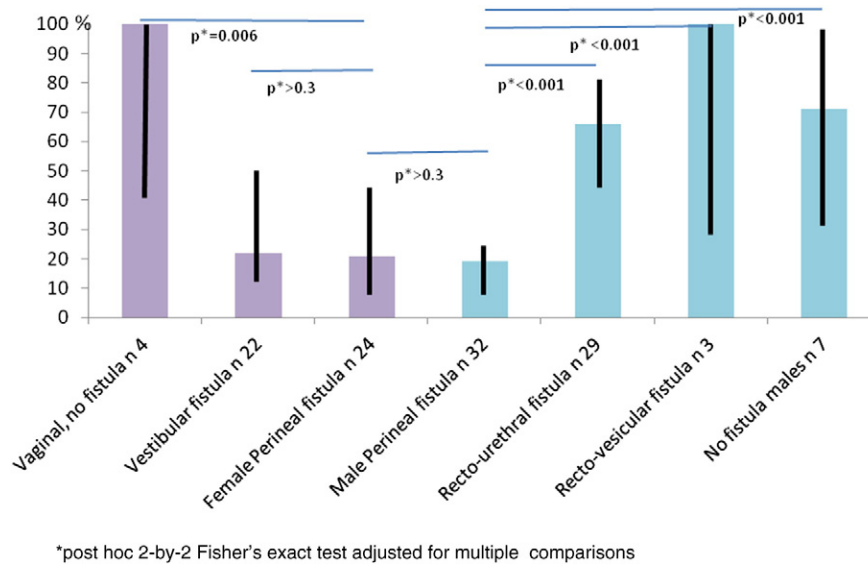


Fig. 2. Absent voluntary bowel movements among children with anorectal malformation (ARM). Comparison gender-wise between the perineal fistulas and the rest of the subtypes of ARM with 95% confidence interval. n = number.

groups concerning voluntary bowel movements ($p > 0.3$), fecal incontinence (>0.3) or constipation (>0.3) (Fisher's exact probability test, two tailed). Therefore all the patients with perineal fistulas were analyzed.

The frequency of associated anomalies in each gender and ARM-subtype is summarized in Table 3.

3.1. Voluntary bowel movements

Voluntary bowel movements were achieved statistically equally in the females (70%) and the males (58%). Among the children with perineal fistulas, 79% of the females reported voluntary bowel movements as opposed to 91% of the males. Almost 75% of the females with vestibular fistula had voluntary bowel movements, but only 33% of the males with rectourethral fistula (Table 4). The frequency of absent voluntary bowel movements was similar for the females with perineal fistula and for those with vestibular fistula,

while among the males with rectourethral fistula significantly more reported absent voluntary bowel movements than those with perineal fistulas (Fig. 2).

The loss of voluntary bowel movements did not differ between the lower and higher age groups for the females or for the males (Fig. 3).

3.2. Incontinence

The frequency of fecal incontinence was equal among the females (48%) and the males (42%). Females with perineal fistulas reported a higher frequency of incontinence than the males with perineal fistulas (42% and 10%, respectively) and similar to that of the females with vestibular fistulas (50%). The males with perineal fistulas had significantly less incontinence compared to the rest of the males with other subtypes of ARM (Table 5).

When comparing all grades (0–3) of incontinence in the females, there was no significant difference in the distribution of ranks of

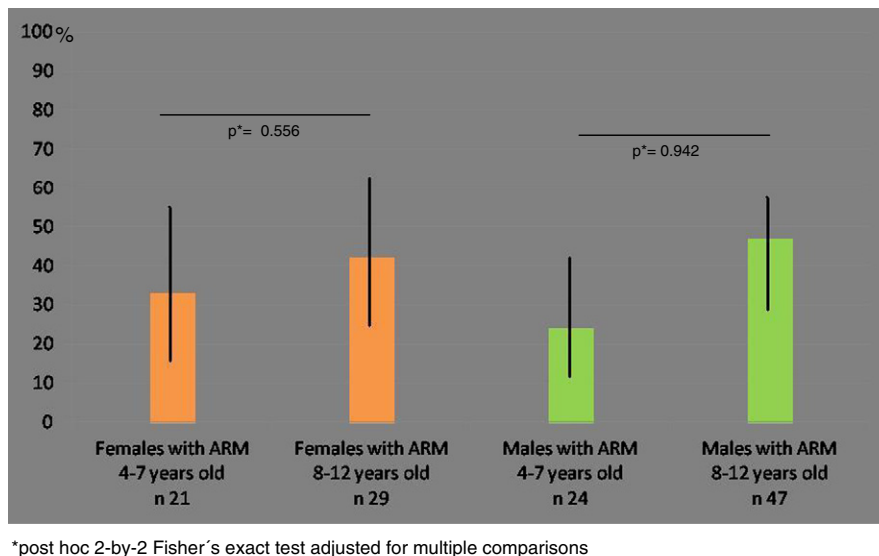


Fig. 3. Absent voluntary bowel movements among the children with anorectal malformation (ARM) in the age groups 4–7 and 8–12 years. Comparisons between the age groups for females and males, respectively, with all subtypes of ARM with 95% confidence interval. n = number.

Table 5

Fecal incontinence of different degrees according to the Krickenbeck follow-up in 121 children with anorectal malformations (ARM) n = number (%).

Fecal Incontinence	Females Total	Females Perineal fistula	Females Vestibular fistula	Females Vaginal fistula	Females No fistula	Males Total	Males Perineal fistula	Males Recto-urethral fistula	Males Recto-vesicular fistula	Males No fistula
n	50	24	22	1	3	71	32	29	3	7
No	26 (52)	14 (58)	11 (50)	0	1 (33)	41 (58)	29 (90)	9 (31)	2 (67)	1 (14)
Yes	24 (48)	10 (42)	11 (50)	1	2 (67)	30 (42)	3 (10)	20 (69)	1 (33)	6 (86)
Grade 1 (occasionally 1–2/week)	13 (26)	6 (25)	6 (27)	1	0	8 (11)	2 (6)	3 (10)	1	2 (29)
Grade 2 (every day, no social problem)	4 (8)	2 (8)	1 (5)	0	1	4 (6)	1 (3)	1 (3)	0	2 (29)
Grade 3 (constant, social problem)	7 (14)	2 (8)	4 (18)	0	1	18 (25)	0	16 (55)	0	2 (29)

Statistical analyses: Comparing fecal incontinence (yes/no) between.

a) the total group of females and the total group of males ($p > 0.3$).*b) the females with perineal fistula and the rest of the females respectively with vestibular fistula ($p = 0.282$ respectively $p > 0.3$).*c) the males with perineal fistula and the rest of the males respectively with rectourethral fistula ($p < 0.0001$ respectively $p < 0.0001$).*d) the females with perineal fistula with the males with perineal fistula ($p = 0.006$).*

* Fisher's Exact Probability Test, two-tailed.

incontinence when comparing perineal and vestibular fistula ($p > 0.3$, Kruskal–Wallis with post hoc test for rank data). Also severe incontinence (Krickenbeck grade 2 and 3) was equally reported in females with perineal and vestibular fistula (Fig. 4).

Comparison of all grades (0–3) of incontinence in males showed that those with perineal fistulas had a rank profile with less incontinence than the other male subtypes of ARM ($p < 0.001$, Kruskal–Wallis with post hoc test for rank data). In line with this, severe incontinence was significantly more common in the subgroups with recto-urethral fistula and no fistula compared with the males with perineal fistula (Fig. 4).

Among those with perineal fistula, severe incontinence was significantly more frequent among the females than among the males (Fig. 4).

The prevalence of severe incontinence did not differ between the lower and higher age groups, either for females or males with ARM (Fig. 5).

3.3. Constipation

The frequency of constipation (yes/no) was statistically higher among the females (62%) than among the males (35%). The frequency did not differ between the children with perineal fistulas and the rest

of the subtypes of ARM, either for females or males (Table 6). For those with perineal fistulas, constipation was more frequent among the females than among the males. The frequency of constipation was similar for the females with perineal fistulas and those with vestibular fistulas (Fig. 6).

The rank of the different degrees of constipation (Krickenbeck grades 0–3) did not differ between the perineal fistulas and the rest of the subtypes of ARM in either gender ($p > 0.3$ Kruskal–Wallis post hoc test for rank data). The prevalence of constipation grade 3 (not manageable with diet or laxatives) did not differ between females and males with perineal fistulas ($p > 0.3$, Fisher's exact post hoc test) or between those with perineal fistulas and the rest of the subgroups in each gender ($p > 0.3$ in each comparison, Fisher's exact post hoc test).

The frequency of constipation did not differ significantly between the younger and older females with ARM. However, among the males constipation was less frequent in the higher than in the lower age group (Fig. 7).

3.4. Sacral malformation correlated to bowel symptoms

The frequency of sacral malformations did not differ between the females (28%) and males (23%), between the females and males with

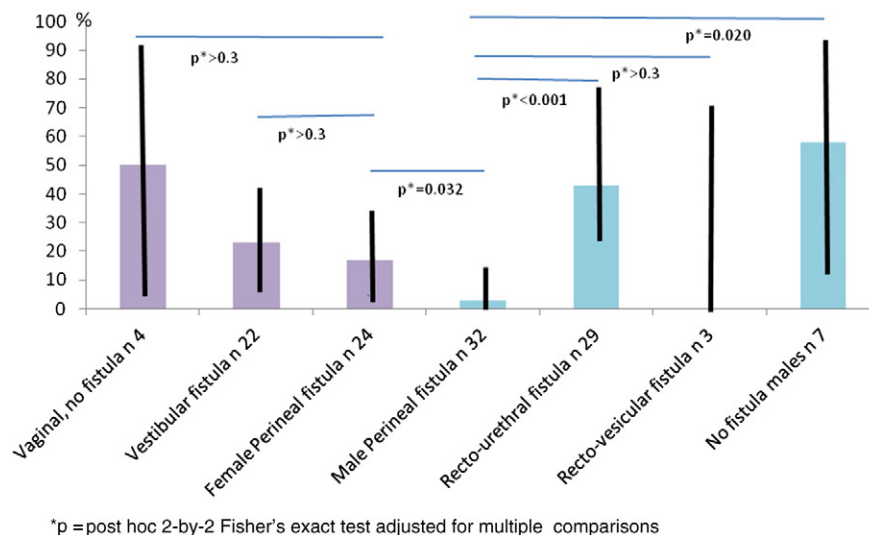


Fig. 4. Severe incontinence according to Krickenbeck grade 2 and 3 among children with anorectal malformation (ARM). Comparisons gender-wise between perineal fistulas and the rest of the subtypes of ARM with 95% confidence interval. n = number.

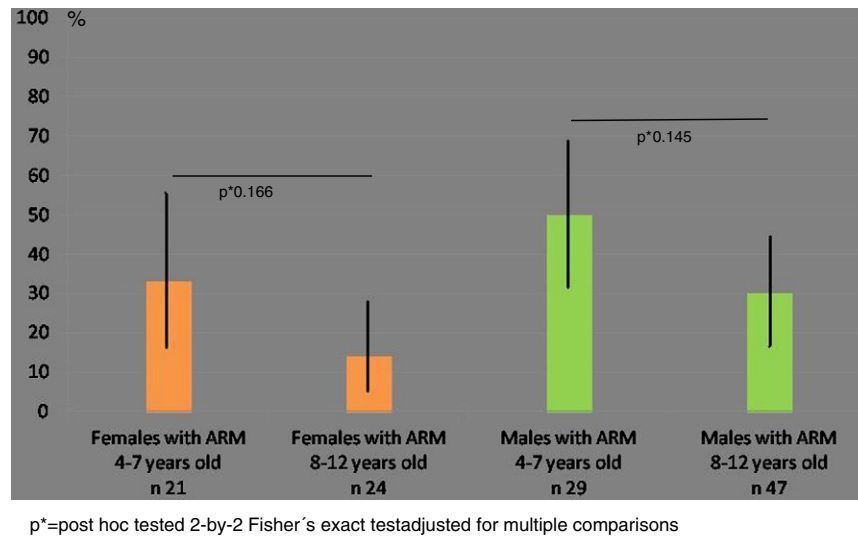


Fig. 5. Severe incontinence according to Krickenbeck grade 2 and 3 in the age groups 4–7 and 8–12 years. Comparison between the age groups for females and males with all subtypes of anorectal malformation (ARM) with 95% confidence interval. n = number.

Table 6

Constipation of different degrees according to the Krickenbeck follow-up in 121 children with anorectal malformations (ARM). n = number, (%).

Constipation	Females Total	Females Perineal fistula	Females Vestibular fistula	Females Vaginal fistula	Females No fistula	Males Total	Males Perineal fistula	Males Recto-urethral fistula	Males Recto-vesicular fistula	Males No fistula
n	50	24	22	1	3	71	32	29	3	7
No	19 (38)	9 (38)	10 (45)	0	0	46 (65)	24 (75)	17 (59)	2 (67)	3 (43)
Grade 1 (diet)	3 (6)	2 (8)	1 (5)	0	0	4 (5)	3 (9)	1 (3)	0	0
Grade 2 (laxatives)	21 (42)	11 (46)	7 (32)	1	2 (67)	14 (20)	4 (13)	6 (21)	1 (33)	4 (57)
Grade 3 (diet or laxatives are not enough)	7 (14)	2 (8)	4 (18)	0	1 (33)	7 (10)	1 (3)	5 (17)	(0)	(0)

Statistical analysis: Comparison of the frequency of constipation (yes/no) between.

a) the total group of females and males, respectively, with ARM ($p = 0.005$).

b) the females with perineal fistula and the rest of the females ($p > 0.3$).*

c) the males with perineal fistula and the rest of the males ($p = 0.136$).*

Gender specific statistical analyses regarding the ARM-subtypes are illustrated in Fig. 8.

** Fisher's Exact Probability Test, two-tailed.

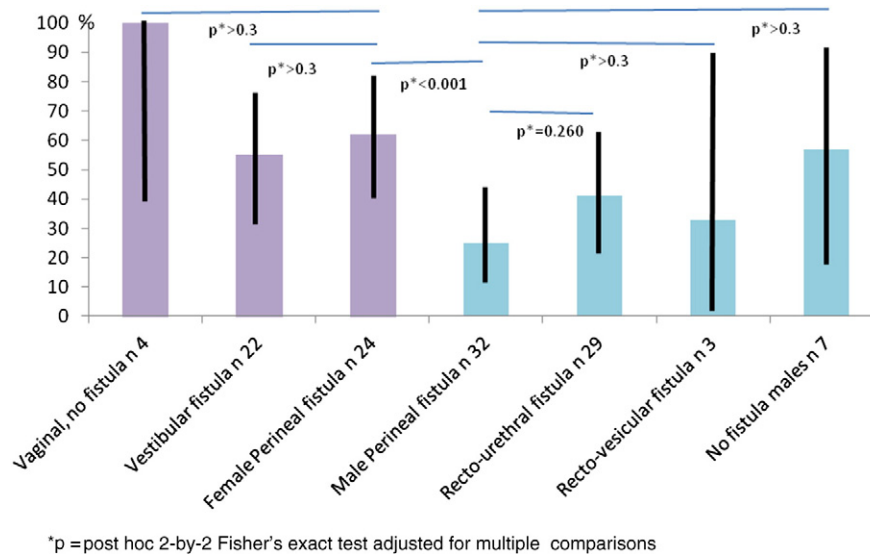


Fig. 6. Frequency of any grade of constipation (grades 1–3) according to Krickenbeck follow-up in children with anorectal malformation (ARM). Comparison between the perineal fistulas and the rest of the subtypes of ARM, gender-wise, with 95% confidence interval. n = number.

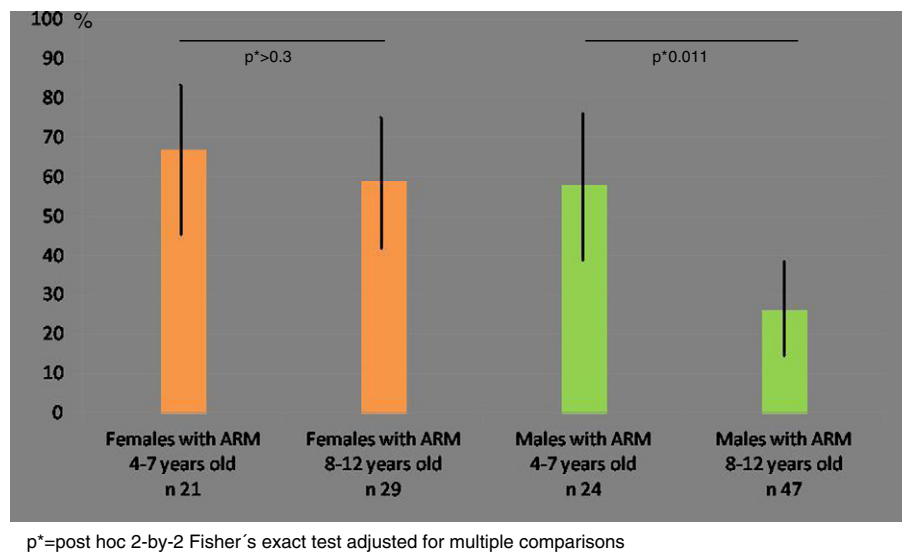


Fig. 7. Frequency of any grade of constipation (grades 1–3) according to Krickenbeck follow-up in the age groups 4–7 and 8–12 years for children with all types of anorectal malformation (ARM). Comparisons between the age groups, gender-wise, with 95% confidence interval. n = number.

perineal fistula (17% and 9%, respectively) or between the females with vestibular fistula (45%) and males with rectourethral fistula (31%) (Fig. 8).

Tables 7 and 8 show the correlation between bowel symptoms and sacral malformations in the gender specific subgroups of ARM. The only significant positive correlation found was that males with rectourethral fistulas and concomitant sacral malformations, had significantly less voluntary bowel movements (100%) and a higher frequency of fecal incontinence compared to those without sacral malformations (100% and 55%, respectively).

3.5. Discussion

The most important and new finding in this study is that females with perineal fistula seem to have a less favorable outcome than males

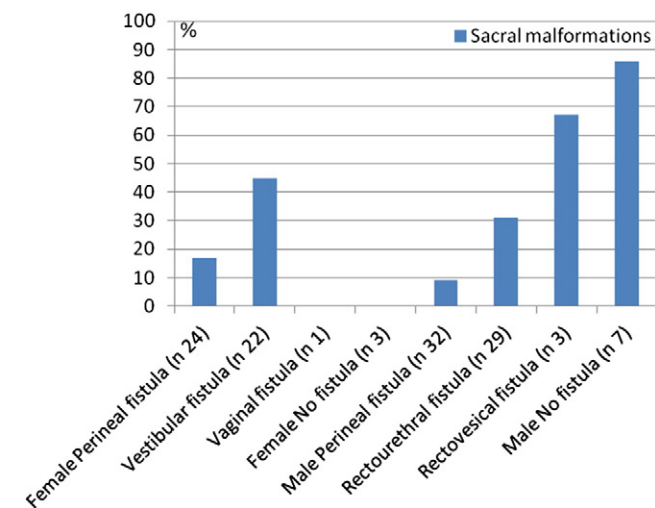


Fig. 8. Frequency (%) of sacral malformations in the different subgroups of anorectal malformations (ARM). n = number. Statistical analyses with comparison of the frequency of sacral malformations between: a) The total groups of females and males ($p = 0.672$)*. b) The females and males with perineal fistulas ($p = 0.686$)*. c) The females with perineal fistulas and vestibular fistulas ($p = 0.035$)*. d) The males with perineal fistulas and rectourethral fistulas ($p = 0.052$)*. e) The females with vestibular fistulas and males with rectourethral fistulas ($p = 0.384$)*. *Fisher's Exact Probability Test, two-tailed.

with the same malformation. In this patient population, females with perineal fistulas have the same outcome as those with vestibular fistulas. Consequently, the gender of a child with perineal fistula seems to be a factor that has to be taken into consideration when the prognosis for post operative bowel function is assessed.

There is a lack in the literature of gender specific analysis of the outcome after PSARP for perineal fistulas. Perineal fistulas are gathered into one gender mixed group, also in the Krickenbeck classification [3]. Contrary to our study, none of the other reports using Krickenbeck system [4,7,6] have evaluated females and males with perineal fistulas separately. As far as we are aware, similar gender specified results have not been demonstrated before.

Sacral anomalies have in some studies been associated with the outcome for bowel control in ARM-patients [12,13]. Interestingly, in the present study sacral malformation did not correlate with the outcome in the female ARM subtypes, the total group of females/males with ARM or perineal fistulas. The prevalence of syndromes was also the same for both genders with perineal fistulas. Therefore, the gender differences in functional outcome for perineal fistulas cannot be referred to either sacral malformations or an unequal frequency of syndromes.

It is difficult to speculate on the reasons for the gender different outcome. Anatomically there should not be any difference in the anorectal malformation. It would be unfortunate if the operation technique differed between the genders, but one could consider that a limited dissection of the rectum in females because of a fear of perforating the vagina, could lead to an incomplete rectoplasty. Another speculative reason for gender differences in bowel symptoms is that the anorectal malformation is less openly discussed with the females in their families [14]. Secondary to this, bowel management may fail, which in the long term could have a negative effect on the bowel control [15]. Other studies focusing on the gender differences in order to confirm or oppose our findings would be of value.

Since there are no previous studies separating females and males with perineal fistulas, it is not possible to make comparisons regarding the similar outcome for female perineal fistulas and vestibular fistulas. In the present study the frequency of concomitant malformations and sacral malformations was higher among the females with vestibular fistulas compared to the females with perineal fistulas. In spite of this, and that the anatomical features are supposed to differ between the subtypes, the outcome regarding incontinence

Table 7

Correlation between sacral malformations and symptoms, in females with.

Krackenbeck follow-up		Female Perineal fistula n 24			Vestibular fistula n 22		
		Sacral malformations		p-value ^a	Sacral malformations		p-value ^a
		Without n 20	With n 4		Without n 12	With n 12	
Voluntary Bowel Movements	Yes	17 (85)	2 (50)	p = 0.179	9 (75)	7 (70)	p > 0.3
	No	3 (15)	2 (50)		3 (25)	3 (30)	
Fecal incontinence	No	13 (65)	1 (25)	p = 0.272	8 (67)	3 (30)	p = 0.198
	Yes	7 (35)	3 (75)		4 (33)	7 (70)	
Occasionally	Grade 1	5	1	p = 0.259	1	5	p > 0.3
Every day, not a social problem	Grade 2	1	1		0	1	
Constantly, social problem	Grade 3	1	1		3	1	
Constipation	No	9 (45)	0	p = 0.259	6 (50)	4 (40)	p > 0.3
	Yes	11 (55)	4 (100)		6 (50)	6 (60)	
Diet controlled	Grade 1	2	0		1	0	
Laxative	Grade 2	8	3		4	3	
Diet or laxative is not enough	Grade 3	1	1		1	3	

^a Fisher's Exact Probability Test, two-tailed.

and constipation was similar for those females. Since the numbers of patients in the various subgroups are relatively small, further studies are needed to assess gender specific outcome.

Without separation of female and male perineal fistulas, there are some earlier published studies of the outcome after PSARP where the Krackenbeck postoperative follow-up system has been used. One is a multicenter study [4] and there are also some comparable single center studies [7,9,6]. A similar distribution of the subtypes of ARM, as in this study, was found in two of those reports [4,7]. However the subtypes recto-bulbar and recto-prostatic fistulas had to be gathered into “recto-urethral fistula” in the present report since the Krackenbeck subtyping was not used in the early years of the study, and a retrospective interpretation was considered as unreliable.

The results from this study demonstrate a considerably poorer outcome compared with results from the single center studies [7,6] but in line with those from the multicenter study [4]. Severe incontinence (Krackenbeck grade 2 and 3) was reported by 24% in the present study and by 30% in the multicenter study. The frequency of severe constipation (Krackenbeck grade 3) was similar in the multicenter and our study, 15% and 12% respectively [4]. An even worse outcome regarding voluntary bowel movements was found in the present study where 37% did not have any voluntary bowel

control compared with 24% in the multicenter study [4]. In contrast, better results were reported in our study than in the multicenter study [4] regarding “no incontinence” and “no constipation”. The similarities and differences could be true or reflect the difficulties in interpreting the outcome according to the Krackenbeck follow-up and definitions of soiling and incontinence. The results could also reflect that the answers in our study were based only on those of the families and not on any objective examination of constipation as in the multicenter study [4].

Regarding evaluation of the outcome for the small groups of males with recto-vesicular fistula and females with no/vaginal fistula, we have too few patients to make any conclusion.

The outcome of the males with perineal fistulas in this report differed from a follow-up study of males with perineal fistulas, median age 9 years, where all the patients had voluntary bowel movements and mainly suffered from light soiling and light constipation [16,17]. Since the follow-up schedules were different, it is difficult to make a trustworthy comparison, especially regarding the severity of symptoms. One reason for the difference may be that the age of the males in the two studies differs. As the result in the present study suggests, constipation becomes less frequent as males with ARM grow older.

Table 8

Correlation between sacral malformations and symptoms in 71 males with anorectal malformation (ARM).

Krackenbeck Follow-up		Male Perineal fistula n 32			Rectourethral fistula n 29			Rectovesical fistula n 3		Male No fistula n 7		
		Sacral malformations		p-value ^a	Sacral malformations		p-value ^a	Sacral malformations		Sacral malformations		p-value ^a
		Without n 29	With n 3		Without n 20	With n 9		Without n 1	With n 2	Without n 1	With n 6	
Voluntary Bowel Movements	Yes	26 (90)	3 (100)	>0.3	10 (50)	0	= 0.011	0	2	1	1	= 0.287
	No	3 (10)	0		10 (50)	9 (100)		1	0	0	5 (83)	
Fecal incontinence	No	26 (90)	3 (100)	>0.3	9 (45)	0	= 0.027	1	1	0	1	>0.3
	Yes	3 (10)	0		11 (55)	9 (100)		0	1	1	5 (83)	
Occasionally	Grade 1	2	0		3	0		0	1	1	1	
Every day, not a social problem	Grade 2	1	0		0	1		0	0	0	2	
Constant, social problem	Grade 3	0	0		8	8		0	0	0	2	
Constipation	No	21 (72)	3 (100)	>0.3	11 (55)	6 (67)	>0.3	1	1	0	3 (50)	>0.3
	Yes	8 (28)	0		9 (45)	3 (33)		0	1	1	3 (50)	
Diet-controlled	Grade 1	3	0		1	0		0	0	0	0	
Laxative	Grade 2	4	0		5	1		0	0	1	3	
Diet or laxatives is not enough	Grade 3	1	0		3	2		0	1	0	0	

The number of males with rectovesical fistulas was too small for statistical comparison. n = number (%).

^a Fisher's Exact Probability test, two tailed.

One potential bias in the present study was the perineal fistulas operated with cut-back and Y-V-plasty. However, in comparison of outcome there were no statistical differences in either gender.

Bowel function reported by the ARM patients in this study was compared with bowel symptoms reported in a Finnish population of normal children aged 4–12 years assessed by the Rintala scale [18]. Since the evaluation differed from the Krickbeck follow-up scheme, the comparisons of the severity of symptoms had to be approximated. In the present study fecal incontinence occurred in 48%/42% of all females/males with ARM and severe incontinence in 36%/37%. In the normal population 43% had some incontinence but only 1% had severe problems. Thus, children with ARM suffer significantly more from severe incontinence than healthy children. Also constipation was reported more frequently among the patients than the normal population. 11% of the healthy children reported constipation compared to 62% females and 35% males with ARM in this study. The lowest prevalence of constipation was found among the males with perineal fistulas (25%), but still this figure was twice that of the normal population. The information that light soiling and constipation are normal phenomena among children in general is important when initiating bowel management for patients with ARM. The goals should not be set too high but instead aimed to reach “social continence” [19].

The secondary aim of the study was to map if bowel symptoms differed between the younger and older age group. Improvement of bowel related symptoms over time among children with ARM has been suggested [20] and also shown in some few patients in a longitudinal follow-up until 15 years of age [10]. The result from this study did not show less incontinence in the older age group or less constipation for older females with ARM. Still the oldest children in the study were only 12 years old, and there may be improvements later on in adolescence [14].

Weaknesses with the present age-related comparisons are that this was not a longitudinal study and that all subtypes of ARM were gathered together. Caution is therefore advisable when drawing conclusions about the symptom development over time in the different ARM-subtypes.

In conclusion, the results from this study suggest that bowel symptoms in a child with ARM depend on both gender and subtype of ARM. Females and males with perineal fistulas should not be expected to have a similar prognosis, while females with perineal and vestibular fistulas have a similar outcome.

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