Journal Pre-proof

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PII: S0022-3468(22)00180-4
DOI: https://doi.org/10.1016/j.jpedsurg.2022.02.015
Reference: YJPSU 60644

To appear in: *Journal of Pediatric Surgery*

Received date: 22 July 2021
Revised date: 31 January 2022
Accepted date: 8 February 2022

Please cite this article as: Hendrick J.J. van der Steeg , Iris A.L.M. van Rooij , Barbara D. Iacobelli , Cornelius E.J. Sloots , Anna Morandi , Paul M.A. Broens , Igor Makedonsky , Francesco Fascetti Leon , Eberhard Schmiedeke , Araceli García Vázquez , Marc Miserez , Gabriele Lisi , Paola Midrio , Eva E. Amerstorfer , Maria Fanjul , Johanna Ludwiczek , Pernilla Stenström , Alida F.W. van der Steeg , Ivo de Blaauw , on-behalf-of-the-ARM-Net-Consortium , Bowel function and associated risk factors at preschool and early childhood age in children with anorectal malformation type rectovestibular fistula: An ARM-Net consortium study, *Journal of Pediatric Surgery* (2022), doi: https://doi.org/10.1016/j.jpedsurg.2022.02.015

Bowel function and associated risk factors at preschool and early childhood age in children with anorectal malformation type rectovestibular fistula: An ARM-Net consortium study

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Abstract

Background Outcome of patients operated for anorectal malformation (ARM) type rectovestibular fistula (RVF) is generally considered to be good. However, large multi-center studies are scarce, mostly describing pooled outcome of different ARM-types, in adult patients. Therefore, counseling parents concerning the bowel function at early age is challenging. Aim of this study was to evaluate bowel function of RVF-patients at preschool/early childhood age and determine risk factors for poor functional outcome.

Methods A multi-center cohort study was performed. Patient characteristics, associated anomalies, sacral ratio, surgical procedures, post-reconstructive complications, one-year constipation, and Bowel Function Score (BFS) at 4-7 years of follow-up were registered. Groups with below normal (BFS < 17; subgroups 'poor' ≤ 11, and 'fair' 11 < BFS < 17) and good outcome (BFS ≥ 17) were formed. Univariable analyses were performed to detect risk factors for outcome.

Results The study included 111 RVF-patients. Median BFS was 16 (range 6-20). The 'below normal' group consisted of 61 patients (55.0%). Overall, we reported soiling, fecal accidents, and constipation

in 64.9%, 35.1% and 70.3%, respectively. Bowel management was performed in 23.4% of patients. Risk factors for poor outcome were tethered cord and low sacral ratio, while sacral anomalies, low sacral ratio, prior enterostomy, post-reconstructive complications, and one-year constipation were for being on bowel management .

Conclusions Although median BFS at 4-7 year follow-up is nearly normal, the majority of patients suffers from some degree of soiling and constipation, and almost 25% needs bowel management. Several factors were associated with poor bowel function outcome and bowel management.

Keywords

Anorectal malformation; Rectovestibular fistula; Bowel function score; Preschool age; Early childhood; ARM-Net, Type of Study, Observational Cohort-Study, Level of Evidence, Level III

1. Introduction

In females with an anorectal malformation (ARM), the rectovestibular fistula (RVF)-type is common[1-3] and, together with the rectoperineal fistula type, often referred to as a 'low' type ARM.[4-6] Although associated congenital anomalies can have a profound effect on functional results, these ARM-types are generally considered to have favorable outcome.[4,5,7-9] However, despite the fact that the type of fistula, the number and type of associated anomalies, and even the surgical procedures are different per ARM-type, overall conclusions on outcome are generally pooled into one ARM-group.[5-7,10,11] Additionally, most studies are single-center studies.[5-7,9,12-15] Furthermore, favorable results are often attained and described in adulthood[12,16], and again, mainly reported combined with data of other types of ARM.[6,7] Finally, surgical approach and perioperative care such as bowel preparation and antibiotic prophylaxis in RVF-patients are heterogeneous, even among specialized centers.[17] As a result, little specific data is available on the outcome of ARM type RVF and associated risk factors. The start of primary school (preschool/childhood age) is an important moment in time, when being potty trained is often obligatory. Thus, based on the described lack of specific data, counseling parents concerning the potential bowel function of their affected child at that age is challenging. The aim of this study is therefore to investigate the functional outcome at preschool and early childhood age (4-7 year) in RVF-patients, treated in European pediatric surgical centers joined in the ARM-Net consortium. In addition, we aim to identify factors that have a negative influence on this outcome. Our study is performed as a multi-center cohort study using collected data from the European ARM-Net registry.[18]

2. Methods

The ARM-Net consortium registry contains all ARM-patients treated in the involved pediatric surgical centers. The registry was opened in 2011, retrospectively including patients born from 2007 till 2011, and generally prospectively since 2011, depending on the moment of joining the consortium. Centers typically treat 5-25 new ARM-patients per year. Available data involve pseudo-anonymized background data, ARM-type, genetic and VACTERL-screening (VACTERL-association defined as ≥ 3 different VACTERL-anomalies[19]), surgical procedures, post-reconstructive complications (defined as minor and major according to Clavien-Dindo classification[20]), and one-year follow-up outcome parameters, e.g. constipation.[21] Measurements of sacral ratio, as a marker of sacral development and innervation, were documented, and a ratio in the antero-posterior plane of < 0.74 and/or in the lateral plane of < 0.77 was considered below normal (low).[1] While the registry does not always elucidate in which plane the measurement was done, a ratio < 0.74 was defined as being low. Constipation has been defined by the ARM-Net consortium as defecation requiring any form of intervention (e.g. change of diet, laxatives or enemas). Bowel management was defined as the need

for regular invasive rectal measures (enemas, wash outs) against otherwise untreatable chronic constipation and/or fecal incontinence. Specific criteria to initiate bowel management have not been defined within the consortium, and have been left to the discretion of the treating surgeon or multidisciplinary team.

The Bowel Function Score (BFS) as originally described by Rintala[22] is the internationally preferred score to document functional outcome in ARM-patients[7,9,13-15] and considered the standard of care within the ARM-Net consortium to evaluate bowel function of any ARM-patient. The BFS consists of 7 items[22]. Fecal incontinence is differentiated into 'soiling' (staining of the underwear or involuntary loss of small amounts of stool) and 'fecal accidents' (involuntary loss of large amounts of stool, requiring change of underwear). The score ranges from 0 to 20 points. Based on evidence that > 90% of healthy children of 4-7 years old have a BFS ≥17, suggesting this score to be normal for this age-range[23], outcome-selection and cut-off scores were determined as follows: below normal (<17) and good (≥17). Because the 'below normal' bowel function outcome group has a large range of BFS, we divided this group into two subgroups: 'poor' (BFS ≤ 11) and 'fair' (11 < BFS < 17). Patients on bowel management were grouped according to their actual BFS.

Bowel function in ARM-patients was evaluated in the timeframe of 4-7 years of age, while in the general population children are expected to be toilet trained at the age of 4[23,24], but not all patients are seen in the outpatient clinic at the age of 4. We identified those RVF-patients in the registry in whom preschool or early childhood functional outcome could be assessed before the age of 8. Patients with developmental delay were excluded, because achievement of continence may be significantly impaired by their developmental delay at this particular early age-frame. When assessed at the outpatient clinic, the questions were answered independently by the parents, as originally proposed by Rintala[25], or by the surgeon/nurse specialist while seeing the patient. If patients were already older and had not had an actual BFS filled in at the outpatient clinic at 4-7 years of age, the BFS was scored by the treating surgeon/nurse specialist based on the patient's file in that particular timeframe. Any missing data to complete a BFS led to exclusion from the study. Patients were assessed and enrolled in the study until September 2019.

Because BFS-data are presently not included in the ARM-Net registry, a separate Castor-database developed for our earlier study[17] was used, inviting all involved consortium members to extend data on BFS and urinary continence of their entered RVF-patients. The data from this prior study, including presence and timing of enterostomy, postoperative complications, type of complication (minor/major) and redo-surgery, were also used to identify risk factors for poor functional outcome.

The local Institutional Ethical Review Board of Radboudumc-Amalia Children's Hospital waived the study because all data were extracted through patient medical files, and additional evaluation of bowel function scores is considered standard care.

3. Statistical analyses

The distribution of characteristics between the 'below normal' and 'good' bowel function outcome groups was shown by providing frequencies and percentages per group and tested for statistical differences, using the Pearson chi-square or Fisher's exact test, as appropriate, to determine risk factors for a 'below normal' bowel function. In addition, for a detailed look into the 'below normal' bowel function outcome group we also identified differences between the 'poor' (BFS ≤ 11) and 'fair' (11 < BFS < 17) subgroups compared to the 'good' bowel function outcome group. We intended to perform multivariable logistic regression analyses to define independent risk factors for poor functional outcome, using those variables in the model that showed an association with poor bowel function outcome in the univariable analyses. Only two factors met this criterium, but we were not able to perform multivariable analyses with these factors, because of the absence of patients with both a good BFS and a low sacral ratio. As extra analysis, we searched for potential risk factors for being on bowel management by testing for statistical differences using the Pearson chi-square or Fisher's exact test, as appropriate. A P-value of <0.05 was considered statistically significant. Statistical analyses were performed using SPSS 25.0 for Windows (IBM SPSS, Chicago, IL, USA).

4. Results

At the end of the study (September 2019) 198 patients older than 4 years of age were available in the ARM-Net registry. Of these, 49 have not been entered in the Castor-database by our participating centers for further analyses (response rate 75%). Following the in- and exclusion criteria, 111 patients were eligible for statistical analyses (Figure 1). The 'below normal' bowel function outcome group consisted of 61 patients (55.0%), of whom 19 (17.1%) had a poor score (BFS ≤ 11) and 42 (37.8%) a fair score (11<BFS<17). Questionnaires to obtain the BFS were filled in by parents independently (13.5%), by the surgeon/nurse specialist while seeing the patient in clinic (41.4%), or by the surgeon based upon patient's file (38.7%). In 7 cases it was unknown who filled in the BFS questionnaire (6.3%). Statistical analysis showed no significant difference in the frequencies of both groups between different ways of acquiring BFS (data not shown).

A low birth weight (≤ 2500 gr) was seen in 27.4% of the participants (Table 1). VACTERL-association was present in 23.8%. Sacral anomalies and tethered cord were demonstrated in 23.0% and 11.7%, respectively. Data on sacral ratio were available in only 49 patients: 18.4% of these patients had a low sacral ratio. In 7.2% of the patients a syndrome of miscellaneous origin was diagnosed.

An enterostomy as initial treatment had been performed in 17 patients (15.7%) for different reasons, e.g. general practice of the specific center, a fistula too small to allow passage of stool sufficiently, or pre-/dysmaturity.[17] An additional 2 patients had an enterostomy performed during or after reconstruction (as prevention or treatment for wound complications). At time of assessment, all enterostomies had been closed. The preferred type of reconstruction was posterior sagittal anorectoplasty (PSARP: 53.2%), followed by anterior sagittal anorectoplasty (ASARP: 31.8%) and mini-PSARP (15.0%).

Post-reconstructive complications, such as wound infection/-dehiscence or anal stenosis, were recorded in 24 patients (22.2%), 11 of which (45.8%; 10.2% of total) were regarded as major. Redo-surgery was performed in 7 patients (6.5%). In the ARM-Net registry at one-year follow-up, constipation was documented in 44.9% of patients and partial prolapse of anal mucosa was seen in 4.5% of them.

The median BFS was 16 (range 6-20). Despite this, many patients suffered from soiling (64.9%; daily 5.4%), fecal accidents (35.1%; daily 2.7%), and constipation (70.3%; Table 2). Bowel management was needed in 26 patients (23.4%), and in this subgroup the median BFS was 11 (range 6-17). Urinary continence was documented in 87.3% of the patients, whereas only 1 patient was using clean intermittent catheterization.

When comparing the 'below normal' and 'good' bowel function outcome groups, only a low sacral ratio showed a significant difference between the groups (31.0% vs 0% respectively, P = 0.007). The 'fair' subgroup did not show any significant difference besides low sacral ratio compared to the

'good' group, and is therefore not further described (Table 3). Further focusing solely on the 'poor' subgroup, tethered cord was reported significantly more frequent than in the 'good' group (27.8% vs 4.9%, P = 0.02) (Table 3). In addition, the frequency of sacral anomalies was higher in the 'poor' than the 'good' group (37.5% vs 17.4%), although no statistical significance could be shown. The same was true for performing a prior enterostomy (21.1% vs 8.5%), and type of reconstruction, revealing higher frequency of ASARP in the 'poor' than in the 'good' group (57.9% vs 31.9%). Constipation at one-year follow-up was more often reported in the 'poor' group compared to the 'good' group (62.5% vs 39.5%), but also not statistically significant. No differences were seen in the distribution of low birth weight, presence of VACTERL-association, syndromic ARM, post-reconstructive complications, redo-surgery, or prolapse between 'below normal' and 'good' bowel function outcome groups, nor between 'poor' and 'good' bowel function outcome groups.

Concerning bowel management, we identified sacral anomalies, a low sacral ratio, enterostomy prior to reconstruction, post-reconstructive complications, and constipation at one-year follow-up as significant risk factors for developing the need for bowel management at age 4-7 years (Table 4).

5. Discussion

To date, this is the largest study evaluating bowel function at preschool and early childhood age of ARM type RVF-patients. Overall, the bowel function as expressed by the BFS was near normal (median score 16). However, nearly a quarter of patients required bowel management. The study showed that a low sacral ratio, and tethered cord had a negative impact on bowel function. Sacral anomalies, enterostomy prior to reconstruction, and constipation at one-year follow-up also showed higher frequencies in the 'poor' compared to the 'good' bowel function outcome group, but these results did not reach statistical significance. Finally, sacral anomalies, a low sacral ratio, enterostomy prior to reconstruction, post-reconstructive complications, and constipation at one-year follow-up all showed to be a risk factor for the need of bowel management at 4-7 years of follow-up.

Although the overall bowel function score was near normal, certain items within the BFS (i.e. soiling and fecal accidents) were reported with a high frequency and deserve further attention because they are of utmost importance for wellbeing and certainly impact quality of life.[10,26]

Soiling was demonstrated in 64.9% of our patients, with daily soiling in 5.4% of patients. This corresponds with other reports of full continence of merely 33-47%.[27,28] Then again, Hassett et al. identified only 14% soiling in 14 RVF-patients. This was, however, without further details.[29] Others have mostly reported relatively favorable results, with soiling between 10-55%, but in these studies different ARM phenotypes with good prognosis were combined, and details concerning RVF specifically could not be identified.[5,9,11] Whether soiling is a sign of sphincteric dysfunction, inability to feel urge, suboptimal squeezing performance, a result of inadequate treatment of constipation[30], or just poor post-defecation perianal hygiene remains unclear. Furthermore, it is of importance to appreciate that in the general healthy population of the same age-group (4-7 years), 50% of children are reported to soil to a certain extent, with no sex difference.[23] Frequent staining requiring changes of underwear on the other hand is uncommon in the general pediatric population (1.2%).[23] Therefore, our study suggests that soiling is more than just confounding in this age group and might have been underreported in the previous smaller or combined studies. Fecal accidents were reported in 35.1%, with daily occurrence in 2.7%. Other studies reported on overall BFS or general ARM-populations, but specific data on fecal accidents in RVF-patients specifically are scarce.[27]

Bowel management was performed in 23.4% of our patients. Other series show lower frequencies of only 0-20%.[7,11,27,31] The relatively high percentage of bowel management is probably a result of the involved centers of expertise addressing intractable constipation or fecal incontinence as part of their referral function. We therefore treat larger numbers of patients in need of bowel management, and in addition have a relatively low threshold for initiating bowel management, based on positive results in these type of patients.[32,33] The relatively high number of patients being on bowel management has influenced the overall BFS by probably giving an overall better BFS as outcome, although their

median score is still lower than average. Whether a low BFS of an individual patient on bowel management is a reflection of the reason for initiating bowel management (i.e. recently started on bowel management) or a sign of ineffective bowel management, could not be discerned from our study.

Sacral anomalies and tethered cord are two of the most common vertebral or spinal anomalies associated with ARM[28,34,35], and both have been suggested to have a negative impact on bowel function[4,7,16,30], although not confirmed in every report.[28] In our series, tethered cord was indeed shown to be a risk factor for poor outcome.

The sacral ratio as a reflection of sacral hypoplasia and thus defective sacral spine innervation has been introduced as an objective measurement to predict potential continence in ARM-patients, defining the normal sacral ratio to be ≥ 0.74 in the anteroposterior plane and ≥ 0.77 in the lateral.[1] Different studies have, however, challenged its predictive value, because of a wide range of normal sacral ratio values[36,37], the influence of age on sacral ratio measurement[36,37], and the poor reliability of sacral ratio measurement in the lateral plane.[38] In addition, others showed no difference of sacral ratio between continent and incontinent ARM-patients.[39] Our study demonstrated a low sacral ratio to constitute a risk factor for poor outcome in general and for the need of bowel management, albeit from a small selection of patients. Further research is needed to determine the definite role of sacral ratio in predicting continence outcome in RVF-patients.

There is an on-going debate with regard to performing an enterostomy prior to reconstructive surgery in RVF-patients.[30,31,35,40] Many studies have reported on reconstructing a RVF without enterostomy, changing a three-stage repair into a one-stage procedure.[31,35,40] A recent systematic review and meta-analysis described similar prevalences of soiling and constipation at follow-up after one-stage procedure compared to a three-stage repair, despite increased risk of wound infection/dehiscence and anorectal stenosis.[41] Others have reported detrimental effects of post-reconstructive complications on outcome, and therefore remain apprehensive.[30] Our study revealed that an enterostomy prior to reconstruction seems to be associated with poor bowel function, and is indeed a risk factor for developing the need for bowel management. Whether the stoma itself led to worse outcome, or the underlying reason to perform an enterostomy (which was miscellaneous, such as general practice or fistulas too small to sufficiently allow passage of stool)[17] or other influencing factors not registered or accounted for in our registry, could not be discerned due to the limited information in our study.

Complications after reconstruction were encountered in 22.2%, but were not found to negatively impact bowel function outcome in general. Post-reconstructive complications were, however, shown to lead to the need for performing bowel management, independent of being a major or minor complication. As described in our earlier study[17], the term ‘major’ needs to be interpreted with care,

as taking back a patient to theatre for a few additional stitches in a small dehiscence is by the Clavien-Dindo classification regarded as a major complication by definition.[20] It could be hypothesized that any negative impact on healing after reconstruction, whether minor or major, results in less developed bowel control, due to increased fibrosis, limited control on urge, or more psychosomatically impaired defecatory function or behavior, and could thereby introduce the need for bowel management.

Although redo-surgery has been shown to lead to worse outcome than primary reconstructions[42,43], our study could not demonstrate an association, possibly due to limited numbers.

Early constipation, in our registry determined at one-year follow-up[17], affects the majority of patients, as other studies have also reported.[4,6,9,11,41] Since the BFS includes constipation as an item, it is a risk factor for poor outcome in general, and for bowel management in particular. Our study shows it to be most prevalent in the patients with poor bowel function scores. This suggests that early detection and aggressive timely treatment of constipation could improve outcome. Although pre-emptive measures to avoid constipation, such as individualized postoperative dilatation-schedule in order to prevent painful anorectal stimuli, dietary advise and early laxatives are already being used by surgeons involved in ARM-care, this result makes us aware of the actual impact of early constipation on the mid-term outcome. Preferably, this should at least lead to a more focused follow-up on constipation, and constipation related problems. One of the treatment-modalities of functional defecation disorders that has been implemented with success in several centers of the ARM-Net consortium, is the Multidisciplinary psycho- and physiotherapeutic Behavioral Treatment.[44,45] This is particularly true if painful stimuli to the pelvic floor (like perineal wound infection and dilatation-therapy) and/or stool withholding behavior is reported.

Even though this study is the largest to date addressing preschool and early childhood bowel function outcome of RVF-patients and identifying risk factors for poor bowel function outcome, limitations need to be addressed. Although data are collected largely prospectively, this study required additional information occasionally from medical files, introducing retrospectively collected data. This potentially introduced some degree of error, i.e. misclassifying or omitted data. In addition, assessing treatment results by the parents was done independently in only the minority of cases, allowing treating surgeons to receive socially desirable answers, or hear and report biased.[25] However, the percentage of poor/fair outcomes was not significantly different between different ways of obtaining BFS, allowing interpretation of data as being representative. Criteria for initiation of bowel management have not been protocolized, possibly introducing further bias concerning outcome, but unavoidable due to the observational nature of this study using many different clinical centers. Determining BFS < 17 as being below normal, was based on evidence that > 90% of healthy children of 4-7 years old have a BFS ≥17, suggesting this score to be normal for this age-range.[23]

Subdividing the 'below normal' group into subgroups with BFS ≤ 11 ('poor') and 11 < BFS < 17 ('fair') was performed based on other studies using the same cut-off scores.[7,15,27] Defining such arbitrary cut-off scores are necessary to perform analyses, but do not necessarily represent an overt change in bowel function. Finally, we were not able to perform multivariable logistic regression analyses to show the independence of the different risk factors. This was due to absence of patients in some categories. Therefore, the results should be interpreted with some caution and more studies with even larger samples sizes are needed to confirm our results.

6. Conclusion

Although median BFS at preschool and early childhood age is near normal in ARM type RVF patients, the majority suffers from some degree of soiling and constipation. Furthermore, almost 25% of the patients need bowel management. Associated risk factors for poor outcome are low sacral ratio, and tethered cord. Bowel management at 4-7 years of follow-up was seen significantly more often in patients with sacral anomalies, low sacral ratio, enterostomy prior to reconstruction, post-reconstructive complications, and constipation at one-year follow-up. The results of this study are helpful to adequately council parents, realistically manage expectations concerning potential bowel function, and benchmark treatment strategies. Further research with larger study populations is needed to definitively establish the role of low sacral ratio, tethered cord, and other risk factors in functional bowel outcome at a later age. Additionally, obtaining BFS solely by patients/parents, independent of treating surgeons or involved researchers as recommended, will strengthen any future results. Finally, follow-up is needed to determine whether aggressive treatment of constipation, soiling or fecal incontinence at an early age in these patients in fact leads to improved bowel function.

Funding
No sources of funding were used

Declarations of interest
none

Acknowledgements
We would like to thank the following pediatric surgeons as members of the ARM-Net consortium, for their work in keeping the registry updated. Their data have not been included in the present study: Dr. Hakan Çavuşoğlu, Gazi University Faculty of Medicine, Ankara (Turkey); Dr. Emre Divarci, Ege University Medical School, Izmir (Turkey); Dr. Carlos Giné, Hospital Vall d'Hebron, Barcelona (Spain); Dr. Stefano Giuliani, St George's Hospital and University, London (United Kingdom); Dr. Sabine Grasshof-Derr, Bürgerhospital and Clementine Children's Hospital, Frankfurt (Germany); Dr. Martin Lacher, Universitätsklinikum Leipzig, Leipzig (Germany); Dr. Alessio Pini Prato, Children's Hospital "Cesare Arrigo", Alessandria (Italy); Dr. Carlos Reck, Medizinische Universität Wien, Vienna (Austria); Dr. Stephan Rohleder, Universität Medizin Mainz, Mainz (Germany); Dr. Inbal Samuk, Schneider

Children's Medical Center, Petach Tikva (Israel); Dr. Alejandra Vilanova-Sánchez, University Hospital la Paz, Madrid (Spain); Dr. Patrick Volk, Universitätsklinik Heidelberg, Heidelberg (Germany).

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Table 1. General, surgical and one-year follow-up characteristics in 'below normal' and 'good' bowel function outcome group in 111 patients with anorectal malformation type rectovestibular fistula

Characteristic	BFS <17 (N = 61)	BFS ≥ 17 (N = 50)	Overall %	P-value
birthweight ≤ 2500 gr	16/58 (27.6%)	13/48 (27.1%)	27.4%	0.95[1]
VACTERL-association	12/42 (28.6%)	7/38 (18.4%)	23.8%	0.29[1]
sacral anomaly	15/54 (27.8%)	8/46 (17.4%)	23.0%	0.22[1]
tethered cord	9/53 (17.0%)	2/41 (4.9%)	11.7%	0.11[2]
low sacral ratio (< 0.74)	**9/29 (31.0%)**	**0/20 (0%)**	18.4%	**0.007[2]**
syndromic ARM	4/61 (6.6%)	4/50 (8.0%)	7.2%	1.00[2]
enterostomy prior to reconstruction	13/61 (21.3%)	4/47 (8.5%)	15.7%	0.07[1]
type of reconstruction:				
- ASARP	19/60 (31.7%)	15/47 (31.9%)	31.8%	0.53[1]
- Mini-PSARP	7/60 (11.7%)	9/47 (19.1%)	15.0%	
- PSARP	34/60 (56.7%)	23/47 (48.9%)	53.3%	
post-reconstructive complications	14/61 (23.0%)	10/47 (21.3%)	22.2%	0.84[1]
- If yes, major[3]?	5/14 (35.7%)	6/10 (60.0%)	45.8%	0.41[2]
redo-surgery	2/61 (3.3%)	5/47 (10.6%)	6.5%	0.24[2]
constipation (1 year follow-up)	27/55 (49.1%)	17/43 (39.5%)	44.9%	0.35[1]
circumferential/partial prolapse of anal mucosa	4/48 (8.3%)	0/40 (0%)	4.5%	0.12[2]

[1] Pearson chi-square test

[2] Fisher's exact test

[3] according to Clavien-Dindo classification of complications: Grade 3 and 4 are considered major complications

ARM, anorectal malformation; ASARP, Anterior Sagittal AnoRectoPlasty; BFS, bowel function score; PSARP, Posterior Sagittal AnoRectoPlasty

Table 2. Bowel Function Score[22] (total and separate items) in the study population of 111 patients with anorectal malformation type rectovestibular fistula

Bowel Function Score, Median (range)	16 (6-20)
Ability to hold back:	
always	59.5%
weekly problems	23.4%
< weekly problem	9.9%
no voluntary control	7.2%
Feels/reports urge to defecate:	
always	53.2%
most of the time	20.7%
uncertain	18.9%
absent	7.2%
Frequency of defecation:	
every other day to twice daily	82.0%
less or more often	18.0%
Soiling:never	35.1%
occasional, < 1/week	35.1%
frequent, > 1/week	24.3%
daily	5.4%
Accidents:never	64.9%
occasional, < 1/week	23.4%
frequent, > 1/week	9.0%
daily	2.7%
Constipation:no	29.7%
manageable by diet	11.7%
manageable by laxatives	35.1%
manageable by enemas (bowel management)	23.4%
Social problems:	
no social problems	83.8%
sometimes (foul odors)	13.5%
problems causing social life' restrictions	1.8%
severe social and/or psychological problems	0.9%

Table 3. General, surgical and one-year follow-up characteristics in 'poor' and 'good' bowel function outcome group in 111 patients with anorectal malformation type rectovestibular fistula

Characteristic	BFS ≤ 11 (N = 19)	BFS ≥ 17 (N = 50)	P-value
birthweight ≤ 2500 gr	6/16 (37.5%)	13/48 (27.1%)	0.53[1]
VACTERL association	2/12 (16.7%)	7/38 (18.4%)	1.00[1]
sacral anomaly	6/16 (37.5%)	8/46 (17.4%)	0.16[1]
tethered cord	**5/18 (27.8%)**	**2/41 (4.9%)**	**0.02[1]**
low sacral ratio (< 0.74)	**5/12 (41.7%)**	**0/20 (0%)**	**0.004[1]**
syndromic ARM	3/19 (15.8%)	4/50 (8.0%)	0.38[1]
enterostomy prior to reconstruction	4/19 (21.1%)	4/47 (8.5%)	0.21[1]
type of reconstruction:			
- ASARP	11/19(57.9%)	15/47 (31.9%)	0.11[1]
- Mini-PSARP	1/19 (36.8%)	9/47 (19.1%)	
- PSARP	7/19 (5.3%)	23/47 (48.9%)	
post-reconstructive complications	5/19 (26.3%)	10/47 (21.3%)	0.75[1]
- If yes, major[3]?	2/5 (40.0%)	6/10 (60.0%)	1.00[1]
redo-surgery	2/19 (10.5%)	5/47 (10.6%)	0.99[1]
constipation (1 year follow-up)	10/16 (62.5%)	17/43 (39.5%)	0.12[2]
circumferential/partial prolapse of anal mucosa	1/15 (6.7%)	0/40 (0%)	0.27[1]

[1] Fisher's exact test

[2] Pearson chi-square test

[3] according to Clavien-Dindo classification of complications: Grade 3 and 4 are considered major complications

ARM, anorectal malformation; ASARP, Anterior Sagittal AnoRectoPlasty; BFS, bowel function score; PSARP, Posterior Sagittal AnoRectoPlasty

Table 4. Risk factors for the need of bowel management (BM) at 4-7 years of follow-up in 111 patients with anorectal malformation type rectovestibular fistula

Characteristic	BM (N=26)	No BM (N=85)	P-value
sacral anomaly	10/23 (43.5%)	13/77 (16.9%)	0.008[1]
low sacral ratio (< 0.74)	7/14 (50.0%)	2/35 (5.7%)	0.001[2]
enterostomy prior to reconstruction	8/26 (30.8%)	9/82 (11.0%)	0.03[2]
post-reconstructive complications	11/26 (42.3%)	13/82 (15.9%)	0.005[1]
constipation (one-year follow-up)	15/24 (62.5%)	29/74 (39.2%)	0.05[1]

[1] Pearson chi-square test

[2] Fisher's exact test

Figure 1. Flow-chart showing inclusion and exclusion of patients with anorectal malformation type rectovestibular fistula