Annals of Surgery Publish Ahead of Print
DOI:10.1097/SLA.0000000000006606

Patient-reported outcomes of children with an anorectal malformation

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Type of Study: Original article.

Declarations of Interest: none

Financial Support Statement: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors. Professor Sebastian King's position as an Academic Paediatric Surgeon is possible due to a generous grant from The Royal Children's Hospital Foundation [grant number 2016-728].

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Running head: Outcomes in anorectal malformations

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STRUCTURED ABSTRACT

Objective

We aimed to study the impact of anorectal malformation (ARM) type and sacral ratio on continence outcomes in children. We secondarily aimed to compare continence outcomes by age group and determine quality of life (QoL) with different bowel regimens.

Summary Background Data

Children with ARM experience dysfunctional stooling into adulthood. Little is known about how ARM type, sacral ratio, age, and bowel regimen affect continence and QoL.

Methods

We administered five validated survey measures on stooling habits and QoL to children aged 2-19 with ARM at two tertiary hospitals. Sacral ratio as a determinant of sacral hypodevelopment was defined as normal (≥0.7), moderate (0.4-0.0.69), or severe (≤0.39). Children not on an enema program were compared to those on an enema program to identify factors associated with achieving voluntary bowel movements defined as continence. All children were compared to identify factors associated with QoL.

Results

Of 910 patients, half (52.8%) had mild sacral hypodevelopment. In patients not receiving enemas managed solely at study hospitals, most were continent (69.2%). There were no differences in continence demonstrated by sacral ratio; 79.4% of those who were continent had normal/moderate sacral hypodevelopment and mild ARM.

Continence improved with age (50% ≤6 years old, 69.8% 6-12 years old, 82.1% >12 years old; *p*<0.001). Those on enemas and soiling had a QoL 19.6 points lower than those soiling on laxatives, and 20.7 points lower than those who were clean on enemas or continent with voluntary bowel movements without the need for enemas.

Conclusions

In patients on laxatives the type of malformation, but not sacral ratio, is associated with continence in patients with ARM. Continence improves with age; those on enemas with soiling have a worse QoL.

INTRODUCTION

Anorectal malformations (ARM) represent a spectrum of congenital conditions that affect the anorectal region, with variable involvement of the genitourinary systems. The conditions have a reported prevalence ranging from 3 - 5 per 10,000 live births, with a higher incidence in males, and a classification based on anatomical variants and complexity.[1–4] A significant minority of these children are also born with associated anomalies (including the VACTERL association) and/or chromosomal anomalies.

Surgical intervention is often required early in life, including in the first 24-48 hours, and both the ARM type and the associated anomalies influence the management plan.[5,6] Despite marked advances in the surgical approach to ARM, a large proportion of children will continue to experience functional problems well into adulthood.[7] These most commonly include constipation (41-77%) and fecal soiling (33-65%).[8–11] The effect of both of these long-term issues on patient quality of life (QoL) are well described.[12]

Due to the complex, multi-factorial nature of ARM, as well as the high likelihood of persistent functional problems, identification of predictive factors for treatment and long-term management success are required and are vital for clinician-parent discussions and for establishing expectations.[13] Proposed predictors of functional outcomes include ARM type, spine quality, sacral ratio, and the ARM index.[7,11,13,14] However, inconsistent findings throughout the literature suggest the need for further research to understand the relationship between these factors and patient outcomes.

After surgery, regular follow-up and measurement of functional outcomes is advised to evaluate the success of ongoing management strategies. Findings from these assessments may also lead to changes in management. Patient Reported Outcomes Measures (PROMs) (e.g. Baylor Continence scale, Cleveland Clinic Constipation Score, Vancouver Symptoms Score for Dysfunctional Elimination) have provided invaluable information that may be used to quantify and improve patient outcomes. Despite a growth in published research exploring the impact of ARM on patient QoL, evaluation of QoL is not routine in clinical practice. It is also unknown whether the QoL for ARM patients changes with increasing age, as they pass from early childhood and schooling through to adolescence and adulthood.[15–17]

There were a number of aims for the current study from two large specialized pediatric colorectal centers. We aimed to: (1) gain a greater understanding of the impact of the ARM type and sacral ratio on patient QoL outcomes; (2) compare the continence outcomes for ARM patients in different age groups; (3) determine the QoL outcomes achieved with different laxative and enema regimens; and; (4) compare clinical outcomes for patients operated upon in the two reference centers with those operated upon in outside hospitals.

METHODS

This study presents data from two pediatric centers: Nationwide Children's Hospital (Columbus, Ohio, USA) and The Royal Children's Hospital (Melbourne, Victoria, Australia).

Participants

Parents/guardians of patients (2-19 years of age) who were diagnosed with an ARM and managed at the two centers were invited to participate in the study. Eligible patients were identified from the pre-existing colorectal patient databases held within the Department of Pediatric Colorectal and Pelvic Reconstructive Surgery (Nationwide Children's Hospital) and the Department of Paediatric Surgery (The Royal Children's Hospital). Participants of non-English speaking backgrounds were excluded. This was done as both institutions are large, tertiary referral centers that see patients from across their respective countries and internationally, resulting in many primary languages spoken. As the validated survey metrics were not available in all languages, the decision was made to include only English-speaking patients.

Recruitment

Participants from Nationwide Children's Hospital were consented at the time of enrollment as patients and added to the colorectal database, and their electronic medical records were reviewed. Clinically relevant data collected included their involvement in a bowel management program, their status of fecal control, and the scores obtained from several disease-specific and generic questionnaires. Participants from The Royal Children's Hospital were mailed recruitment packs containing a letter inviting them to take part in the study and a Participant Information and Consent Form. A follow-up telephone call was made two weeks after mailing to confirm that they had received the study information and to answer any questions. Participants who indicated a willingness to be involved were asked to provide written consent to take part. Once consent was obtained, a time was scheduled for the survey, which was administered via telephone by a member of the research team.

Definitions

The ARM subtype was defined based on guidelines for severity (Supplemental Table 1, Supplemental Digital Content 1, http://links.lww.com/SLA/F362). Mild ARM included rectoperineal fistula, rectovestibular fistula, rectobulbar fistula, H-type fistula, and stenosis of the anus or rectum. Moderate ARM included short-channel cloaca (<3 cm), rectovaginal fistula, and rectoprostatic fistula. Severe ARM included long-channel cloaca (>3 cm or unknown length), posterior cloacal variant, rectobladder neck fistula, rectovesical fistula, and cloacal exstrophy.

All others were categorized as unknown. Sacral ratio as a marker of sacral hypodevelopment was defined as normal (≥0.7), moderate (0.4-0.7), or severe (≤0.39).[18] Age was sub-grouped as less than 6 years old, 6-12 years old, and above 12 years old. The Rome IV criteria were used to define continence, with continent patients having one or less stooling accident per week while on laxatives or no bowel regimen. The term continence was only utilized for patients on oral medications or not on any medication and not for patient on an enema program. Continence analysis was performed only on patients on laxatives or no bowel regimen; children with enemas were only included for QoL analysis.

The Pediatric Quality of Life Inventory (PedsQL) Generic Core Scale was used to measure health-related quality of life. It is widely used, with demonstrated reliability and validity in children.[19,20] The PedsQL is a 23-25 item questionnaire that assesses health across four scales (physical, emotional, social, school functioning). Items are ranked on a five-point Likert scale, ranging from 0 (never) to 4 (almost always). Items are then reversed scored and linearly transformed to a 0-100 scale, so that high scores indicate a better quality of life. Scales are combined to create the Psychosocial Health Summary Score (the mean of emotional, social and school functioning scales) and the Physical Health Summary Score (the same as the physical functioning scale score). An overall Total Scale Score is calculated as the mean of all items. The PedsQL was completed via proxy-report or self-report. The version used was based on patient age and patient willingness to complete the PedsQL themselves.

Statistical Analysis

Descriptive statistics were computed, including frequencies and percentages for categorical variables, and medians and interquartile ranges for continuous variables. Various comparisons were performed between subgroups of the cohort. For comparisons, chi-square and

Fisher's exact tests were used for categorical variables, appropriately based on cell size. When stratified by age group with sequential ordinate categories, the Cochran-Armitage test and its extensions were used to compare for trend. PedsQL scores were compared by difference in score.

For all continence outcomes, only children ≥3 years old with primary repair on laxatives or no bowel regimen were analyzed. Comparisons were performed first for children undergoing their full care at one of the study hospitals, and again including children referred to a study hospital after initial surgical care elsewhere. For PedsQL, all children including those undergoing redo repair and those on enemas were analyzed. Analyses were completed using SAS version 9.4 (SAS Institute, Cary, NC).

Ethics

Consent was obtained for all participants in the study. Ethics approval was granted by the Institutional Review Board at Nationwide Children's Hospital (IRB 14-00232) and The Royal Children's Hospital Human Research Ethics Committee (ID 36003).

RESULTS

Cohort Description

A total of 910 patients met the inclusion criteria, with 772 (84.8%) from Nationwide Children's Hospital and 138 (15.1%) from The Royal Children's Hospital (**Table 1**). The median age at the time of the study was 10 years [IQR: 7 – 14 years]. Almost half (45.9%) of the cohort was female. Most had a mild (56.1%) or moderate (24.5%) anorectal type; a small majority (52.8%) had a normal sacral ratio (greater than 0.7) and most (79.8%) had a normal spine.

Most patients (614/910, 67.4%) had a primary repair without need for a redo operation (**Figure 1**). Of these, about half (312/614, 50.8%) had their operative intervention and subsequent management at one of the two study hospitals. After excluding those who were using stomas, those who were underage, or those who did not have reportable data on continence or bowel management, most of this cohort (214/312, 68.6%) were on laxatives or no bowel management, of which the majority (126/182, 69.2%) were continent. Of those on retrograde or antegrade enemas (63/312, 20.2%), over half (38/63, 60.3%) were continent. In total, of the 312 patients managed solely at study hospitals of appropriate age with available information on continence and not using stomas, most were continent (164/242, 67.8%).

Patients on Oral Medications

We then analyzed patients at the study hospitals managed with either oral medications or on no bowel management (**Table 2**). When stratified by ARM subtype and sacral ratio, there was no difference in continence rates; we did not find a significant association between sacral ratio and continence. Of note, of those who were continent, 100/126 (79.4%) had a normal to moderate sacral ratio with a mild ARM subtype. Those with mild ARM subtypes and normal sacral ratios that were managed at study hospitals had higher rates of continence when compared with those managed at other hospitals (75.2% vs 57.9%, p=0.02) (**Table 3**).

When compared by age group, continence overall improved by age, with continence rates increasing from 50% in those less than 6 years old to 69.8% in those 6-12 years old and 82.1% in those greater than 12 years old (p<0.001) (**Table 4**). When stratified by ARM subtype, the same trend was observed in patients with a mild ARM subtype, with continence rates improving from 51.9% (<6 years old) to 76.2% (6-12 years old) to 87.0% (>12 years old, overall p-value=0.0012, **Table 5**). This trend was not demonstrated in those with moderate or severe ARM subtypes.

Quality of Life

There was no difference in QoL measures by PedsQL for clean/continent patients based on whether they utilized laxatives or enemas (**Table 6**). There was additionally no difference in PedsQL for those who were clean/continent or not on laxatives. However, those who were on enemas and soiling scored on average 19.6 points lower than those who were soiling on laxatives, and 20.7 points lower than those who were clean/continent on enemas.

DISCUSSION

In this study, we aimed to utilize prospectively collected data from two large pediatric hospitals with busy colorectal services to compare the outcomes of children with ARM. The specific focus was to assess those managed on oral medications in terms of their continence by ARM subtype severity, age, sacral ratio, and bowel management regimen. We have demonstrated that there was no difference in the continence rate when sacral ratio was assessed. An important observation was that when children aged, their rates of continence increased; however, this trend was observed only across those with mild ARM subtypes. Patients who were on enemas and soiling had significantly worse QoL outcomes when compared with those on laxatives and soiling, and those on enemas and clean/continent.

Low sacral ratio has been historically correlated with poor functional outcomes in children with ARM.[21] However, multiple recent studies have disputed this, with no evidence of a practical association between continence and sacral ratio.[14,22] The findings presented here, from the largest study of its kind, confirm the latter, demonstrating that in most patients, sacral ratio has no impact on rates of continence. However, in patients that are managed solely at a tertiary colorectal center, those with less severe malformations, such as mild ARM with normal to moderate sacral ratios, continence rates are higher compared with children initially managed at outside facilities. In children with complex colorectal malformations, there are lower rates of surgical interventions, anesthetics, intubations, hospital admissions and outpatient visits when children are operated on and managed in a multidisciplinary clinic.[23,24] Therefore, it appears consistent that children managed at two large multidisciplinary institutions with experienced surgical providers and access to excellent nursing care as well as urological, gynecological, and neurosurgical providers would have improved outcomes regarding continence. In our centers, children have access to additional resources not available to general pediatric surgeons, including single-center visits with multiple specialties, ancillary services such as experienced and specialized social workers, wound and ostomy nursing, and pelvic floor physiotherapy, and, most importantly, children are taken care of by experienced, high-volume surgeons. This may advocate for ubiquitous management of patients with ARM at tertiary and experienced care facilities to ensure best outcomes. Additionally, it is vital to note that in children with severe ARM subtypes, there are still children able to attain continence. It has been previously suggested that children with high or severe ARM will suffer from fecal incontinence.[25] While there are few patients seen with such severe malformations, it is encouraging that continence is possible in a subset.

Unsurprisingly, as patients' age increased, rates of continence also increased. As children grow older, the impact of fecal incontinence is more significant on their day-to-day life.[26] This results in adolescents and teenagers having enhanced motivation to play a major role in the development of their continence. As children become more participatory in their care, including with medication and therapy compliance, it is expected that their rates of continence should also increase, even in children with anatomical and functional challenges such as those with ARM. Therefore, it is encouraging that this trend was seen in our population. This trend was most pronounced in children with mild ARM subtypes, consistent with prior research demonstrating higher rates of continence in children with lower malformations.[27] This may provide valuable insight in counseling parents and families of children with mild ARM – if a child is unable to achieve continence at a young age, they may still have potential to acquire this later in life. Typical bowel management progresses along a standard pathway involving initiation of least invasive therapies, such as oral medications, to more aggressive therapies, such as rectal enemas, antegrade enemas, and other potential operative interventions. This improvement may also reflect the need for more aggressive therapy early in life, when children have lower rates of continence on laxatives, with subsequent transition to laxatives with age. This is an important additional opportunity for counseling of a patient and family.

It is vitally important to tailor bowel management to meet the patient and family needs, while still maintaining continence. Patients with fecal incontinence are known to have poor QoL.[28] However, the work required to maintain continence with enemas is much more significant when compared with laxatives.[29] Therefore, it is also important to consider whether functional outcomes, such as QoL, correlate with continence. In this study, we show that patients who are clean/continent have no difference in QoL metrics between laxatives and enemas. This suggests that families are willing to pursue aggressive measures with good QoL if their children are continent. However, when children are soiling, those on enema therapies have significantly worse QoL scores, both compared with those that are clean/continent on enemas and those who are soiling on laxatives. This may reflect underlying burden of enema therapy, which is poorly tolerated when the child is not conferred a benefit. This may advocate for earlier transition from enemas, if unsuccessful, to potential surgical interventions, including antegrade enemas in those in whom this has not been trialed, sacral nerve stimulation, or, in the case of severe and refractory incontinence, consideration of ostomy creation if within the patient's and family's goals of care.

Limitations

This study is vulnerable to multiple limitations. First, patients were included only if they received management at any point at one of two major pediatric hospitals. There may be patients managed at outside facilities who have smooth courses who do not require transfer to tertiary care; therefore, the failures in the tertiary care group may represent significantly challenging patient pathologies or those with ability to seek care at other institutions. This may have artificially impacted the low continence rate in those initially operated upon outside of our facilities. Second, due to the small number of children managed annually with severe ARM and severe spinal status, this subgroup are unable to be statistically compared. Additionally, due to the rarity of each individual ARM subtype, grouping was necessary to provide meaningful comparisons. Each individual type of ARM was unable to be studied on its own. However, the two participating institutions represent two of the largest referral centers internationally, with many patients. We have described these cohorts descriptively, but further multicenter studies may provide additional insight. Finally, given the study's retrospective design, we are unable to assess factors that impacted whether children were started on enemas or laxatives, or to mitigate some lost-to-follow-up bias. However, patients were contacted to provide additional details as necessary for clinical care and, therefore, this represents a robust and highly accurate dataset.

CONCLUSION

In conclusion, in a large, multi-institutional sample, we demonstrate that type of malformation, but not sacral ratio is significantly associated with the development of continence in children with ARM. Continence improves with age, especially in children with mild ARM subtypes. While QoL does not differ by bowel management type in children who are clean/continent, those who are soiling on enemas have significantly worse QoL compared with all others. Overall, this provides important discussion points when counseling patients and families and may indicate areas for future study.

ACKNOWLEDGEMENTS

We wish to thank the patients and families who generously donated their time to this research project.

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Figure 1. The primary continence assessment was composed of patients undergoing primary (non-redo) repair without the use of enemas at a study hospital. Secondary continence assessment also included patients referred for care. Quality of life assessment included all patients, including those requiring redo, those on enemas, those with a stoma or unknown continence, or those underage (defined as <3 years old).

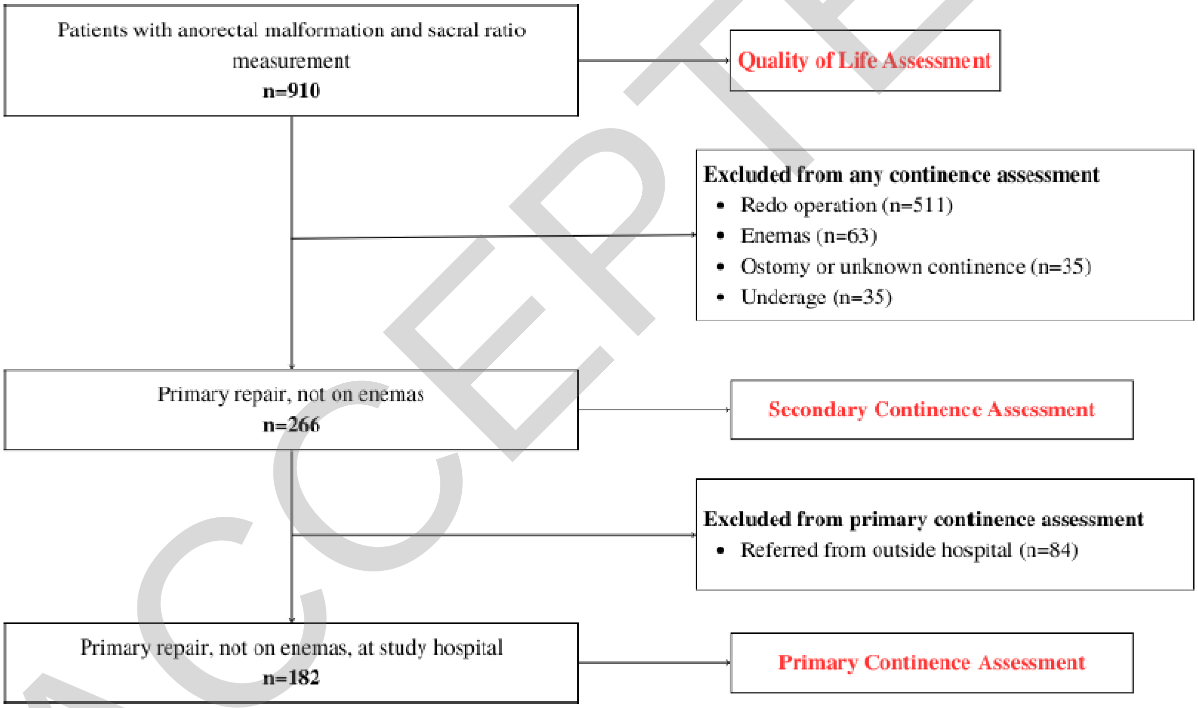

Table 1. Cohort description

Characteristic	n (%) or median [interquartile range] (n = 910)
Current age, years	10 [7-14]
Female	418 (45.9)
ARM subtypes	
Mild	466 (56.1)
Moderate	204 (24.5)
Severe	161 (19.4)
Unknown/Missing	79
Sacral ratio	
Mild (0.7 or greater)	428 (52.5)
Moderate (0.4-0.69)	293 (35.9)
Severe (0.39 or less)	95 (11.6)
Unknown/Missing	94 (10.3)
Spinal status	
Normal	726 (79.8)
Tethered cord release	161 (17.7)
Myelomeningocele	23 (2.5)

Table 2. Continence data for all patients on laxatives or without bowel management (n=182) managed solely at the study hospitals. Fisher's exact tests used for statistical comparison.

ARM subtype	**Sacral ratio**	**Continent** n=126 (69.2%)	**Incontinent** n=56 (30.8%)	**p-value**
Mild	Normal/moderate	100 (79.4%)	33 (58.9%)	>0.999
	Severe	2 (1.6%)	1 (1.8%)	
Moderate	Normal/moderate	19 (15.1%)	10 (30.3%)	N/A
	Severe	0 (0.0%)	0 (0.0%)	
Severe	Normal/moderate	5 (4.0%)	10 (30.3%)	>0.999
	Severe	0 (0.0%)	2 (3.6%)	

Table 3. Comparative outcomes of continence on laxatives or no bowel management in patients based on management by a study hospital and outside of the study hospital. Fisher's p-values. Percentages are cell percentages. Bold indicates statistical significance.

ARM subtype	Sacral ratio	Study hospitals	Other hospitals	p-value
Mild	Normal/moderate	100/133 (75.2%)	33/57 (57.9%)	**0.02**
	Severe	2/3 (66.7%)	1/1 (100.0%)	N/A
Moderate	Normal/moderate	19/29 (65.5%)	11/17 (64.7%)	N/A
	Severe	-	-	
Severe	Normal/moderate	5/15 (33.3%)	7/9 (77.8%)	N/A
	Severe	0/2 (0.0%)	-	

Table 4. Continence information for all ARM subtypes, all sacral ratios, not on enemas, stratified solely by age group. Cochran-Armitage test for trend. Bold indicates statistical significance.

Age Range (years)	**Continent** n=126 (69.2%)	**Incontinent** n=56 (30.8%)	**Continence %**	**p-value**
<6	20 (15.9%)	20 (35.7%)	50%	**0.0009**
6-12	60 (47.6%)	26 (46.4%)	69.8%	
>12	46 (36.5%)	10 (17.9%)	82.1%	

Table 5. Continence data based on age for all patients not on enemas (n=182) managed solely at the study hospitals. Cochran-Armitage tests used for trend, and row percentages are reported. Bold indicates statistical significance.

ARM subtype	**Age Range** (years)	**Continent**	**Incontinent**	**p-value**
Mild	<6	14 (51.9%)	13 (48.1%)	**0.0012**
	6-12	48 (76.2%)	15 (23.8%)	
	>12	40 (87.0%)	6 (13.0%)	
Moderate	<6	4 (66.7%)	2 (33.3%)	0.8608
	6-12	10 (66.7%)	5 (33.3%)	
	>12	5 (62.5%)	3 (37.5%)	
Severe	<6	2 (28.6%)	5 (71.4%)	0.7066
	6-12	2 (25.0%)	6 (75.0%)	
	>12	1 (50.0%)	1 (50.0%)	

Table 6. PedsQL scores for all children in the study with comparison between columns or rows. Bold indicates significant as >18 point difference

	Clean/Continent	**Soiling**	
Laxatives	85.7 [77.3 – 93.5]	83.7 [73.7 – 92.3]	2.0
Enemas	84.8 [73.8 – 95.2]	64.1 [52.5 – 85.1]	**20.7**
	0.9	**19.6**	