Can Fecal Continence be Predicted in Patients Born with Anorectal Malformations?

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Abstract

Purpose

The purpose of this study was to identify factors associated with attaining fecal continence in children with anorectal malformations (ARM).

Methods

We performed a multi-institutional cohort study of children born with ARM in 2007-2011 who had spinal and sacral imaging. Questions from the Baylor Social Continence Scale were used to assess fecal continence at the age of ≥ 4 years. Factors present at birth that predicted continence were identified using multivariable logistic regression.

Results

Among 144 ARM patients with a median age of 7 years (IQR 6-8), 58 (40%) were continent. The rate of fecal continence varied by ARM subtype (p=0.002) with the highest rate of continence in patients with perineal fistula (60%). Spinal anomalies and the lateral sacral ratio were not associated with continence. On multivariable analysis, patients with less severe ARM subtypes (perineal fistula, recto-bulbar fistula, recto-vestibular fistula, no fistula, rectal stenosis) were more likely to be continent (OR = 7.4, p=0.001).

Conclusion

Type of ARM was the only factor that predicted fecal continence in children with ARM. The high degree of incontinence, even in the least severe subtypes, highlights that predicting fecal

continence is difficult at birth and supports the need for long-term follow-up and bowel management programs for children with ARM.

Keywords: Anorectal malformation; Continence; Predictors; Spine; Sacral ratio

Type of Study: Prospective Cohort Study

Level of Evidence: II

Introduction

Children born with an anorectal malformation (ARM) often have associated anomalies of the sacrum and spinal cord which impair the normal functioning of sensory innervation, sphincter control, and colonic motility needed for fecal continence. [1, 2] The arrest of the normal embryologic processes that results in the characteristic anomalies of the anus and rectum may also extend to the pelvic musculature and nerves essential for coordination of these mechanisms, leaving them rudimentary or absent altogether. Consequently, these children may frequently suffer from fecal incontinence later in childhood and throughout life. [1-4]

Parents or caregivers of newborns with an ARM often ask about the impact of their child's anomaly on their ability to be continent of stool later in life. At present, our ability to counsel families is imprecise and often based on personal experience and professional judgment. Anatomic features readily identifiable in the newborn period, including the type of ARM, degree of sacral development, and features of the spinal cord have been implicated in the potential for continence in these children, but these anomalies frequently occur simultaneously and the relative predictive value of each is not well understood. [5-7] Furthermore, most previous studies reporting on fecal continence in children with ARM did not use a standardized objective definition of continence.

The purpose of our study was to determine the association of the ARM subtype, sacral ratio, and presence of various spinal anomalies with fecal continence in children born with an ARM and treated across 10 children's hospitals.

Methods

Study Design and Patient Population

Patients born between January 2007 and December 2011 with a diagnosis of ARM who underwent corrective surgery at 10 children's hospitals participating in the Midwest Pediatric Surgery Consortium (www.mwpsc.org) were identified by retrospective chart review at each institution. This date range was selected to ensure that all patients were 4 years or older at the time of the prospective survey to determine continence. International Classification of Disease, 9th Edition, Clinical Modification (ICD-9-CM) diagnosis codes for ARM (565.1, 596.1, 619.1, 599.1, 602.8, 751.2, 751.5, 752.49, 753.8) were used to identify patients for inclusion. Each patient was classified according to the location of the rectal fistula as determined by imaging studies or the operative report. [8] Patients with a corresponding sacral x-ray and either ultrasound (if <6 months) or MRI of the spine were considered eligible for prospective determination of continence, while those missing either sacral or spinal imaging were excluded.

Data Collection

Demographic and clinical characteristics were recorded for each patient. Sacral x-rays were reviewed and sacral ratios calculated by pediatric radiologists at each institution using the ratio of the length of the sacroiliac joint to the coccyx to the length of iliac crest to the sacroiliac joint. In cases where both lateral and anteroposterior (AP) sacral images were available for review, the lateral sacral ratio was used for this calculation. Spinal ultrasound or MRI imaging reports were reviewed for the presence of a tethered cord, low conus, fatty filum, spinal dysraphism, myelomeningocele or spina bifida, presacral mass, hemivertebra, hemisacrum, or sacral agenesis. Radiologists were blind to the patient's clinical details at the time of sacral ratio

calculation, however any information available in the medical chart was available at the time of interpretation of the spinal imaging. Data was entered into a central Research Electronic Data Capture database. [9]

Prospective Survey

Parents or guardians of each patient were then contacted by study staff at each institution and offered voluntary participation in a prospective survey, administered either by phone or e-mail, to assess fecal continence. To maximize the response rate, study staff at each institution made two attempts to reach parents or guardians by mail and, if unsuccessful, three attempts were made to reach family by telephone. In addition, a $10 incentive was offered for completion of the survey. Continence was determined using the Rome III diagnostic criteria for fecal continence, defined as ≤ 1 stool accident per week. [10] This was reported as part of the Baylor Social Continence Scale, which asks about the number of times in the last week the child experienced any leakage of stool at night, during the day, or both at night and during the day. [11] The survey also asked if the child currently had a colostomy or ileostomy, and it included questions on currently used bowel management techniques, such as whether and how frequently laxatives, enemas, and cecostomy flushes were used, and whether the child had ever attended a dedicated bowel management program. Children with a colostomy or ileostomy at the time of follow-up were considered incontinent.

Statistical Methods

Baseline patient characteristics were described using frequencies and percentages for categorical variables and medians and interquartile ranges for continuous variables. Pearson chi

square, Fisher exact, or Mann Whitney U tests as appropriate were used to compare characteristics between patients who were and were not fecally continent at follow-up. Multivariable logistic regression models including the severity of ARM and particular spinal cord findings of interest were then fit for fecal continence. Due to the relatively small number of patients with a measurement of their lateral sacral ratio, we fit a separate logistic regression model for fecal continence that included only the severity of ARM and the lateral sacral ratio as a continuous variable. Finally, using chi square and Fisher exact tests, we evaluated whether particular bowel management techniques used at follow-up were associated with continence. These factors were not considered for inclusion in the multivariable regression models since they were assessed at follow-up. SAS Enterprise Guide version 7.15 (SAS Institute Inc., Cary, NC) was used for all statistical analyses. P values less than 0.05 were considered statistically significant. Each participating institution received either approval to rely on the lead institution's institutional review board (IRB) or independent approval for this study from their IRB. A waiver of consent was approved by the IRBs.

Results

Among 341 eligible ARM patients with sacral and spinal imaging, 144 (42%) completed the survey assessing fecal continence. There were no significant differences between responders and non-responders to surveys in terms of age, gender, type of ARM, severity of ARM, sacral ratio, or findings on spinal imaging (data not shown). A summary of the demographic and clinical characteristics of the 144 patients included in our analyses is shown in Table 1. The median patient age at the time of the determination of fecal continence was 7 years (IQR 6-8).

Table 1

There was a near even gender distribution, the majority of patients were white, and the median age at the time of the first ARM-related surgery was 3 days. Perineal fistula (37%) was the most common ARM subtype, followed by rectovestibular fistula (12%) and imperforate anus without fistula (10%).

Table 2

The results of univariable analyses of clinical characteristics and fecal continence are shown in Table 2. Overall, 58 patients (40%) were continent of stool. The rate of continence varied by ARM subtype (p=0.002), with the highest rate seen in patients with perineal fistula (60%) and lowest in cloacal exstrophy (0%). There was an inverse relationship between sacral ratio in the AP position and continence rates, with continence seen in 31% of patients with a sacral ratio ≥0.70, 53% with a sacral ratio of 0.40-0.69, and 60% in those with a sacral ratio of <0.40 (p=0.04). The lateral sacral ratio was not significantly associated with fecal continence, with continence rates of patients with lateral sacral ratio ≥0.70, 0.40-0.69, and <0.40 of 39%, 45%, and 50%, respectively (p=0.87). Findings on spinal imaging were not associated with fecal continence, with patients with a tethered cord or low conus continent at a rate of 43% (p=0.69), fatty filum 43% (p=0.79), myelomeningocele/spina bifida 30% (p=0.74), and normal imaging 41% (p=1.00).

Table 3

Table 3 details the use of bowel management and their association with fecal continence. Laxative use and the use of cecostomy flushes were not associated with continence. However, both enema use and history of participation in a bowel management program were significantly more common in patients who were incontinent as compared to those who were continent.

Table 4

Table 4 demonstrates the results of our multivariable modeling to identify factors independently associated with fecal continence. Patients with less severe ARM subtypes

(perineal fistula, recto-bulbar fistula, recto-vestibular fistula, no fistula, or rectal stenosis) were more likely to be continent (OR = 7.4, p=0.001) than patients with the most severe ARM subtypes (cloacal exstrophy, cloaca >3 cm, rectovesical fistula). Findings on spinal cord imaging, including tethered cord or low conus (OR = 1.6, p=0.32), fatty filum (OR = 1.2, p=0.77), or myelomeningocele/spina bifida (OR = 2.3, p=0.36) were not independently predictive of fecal continence. Similarly, when tethered cord was evaluated independently instead of tethered cord/low conus, patients with the lowest severity ARM were statistically more likely to be continent (OR = 6.7, 95% CI 1.7-26.1, p=0.002), while the presence of tethered spinal cord (OR 0.9, 95% CI 0.3-2.4, p=0.80), fatty filum (OR 1.5, 95% CI 0.5-4.6, p=0.50), or myelomeningocele/spina bifida (OR 2.8, 95% CI 0.4-17.6, p=0.27), were not statistically significant.

Discussion

This study investigated the association of ARM subtype, sacral development, and spinal cord anomalies with obtaining fecal continence later in life in children with ARMs across 10 institutions. Using the definition of fecal continence as the ability to have voluntary bowel movements with no more than one “soiling” event per week based on the Rome III Diagnostic Criteria, our results suggest that the ARM subtype is the only independent predictor of fecal continence later in life.

The association between ARM subtype and continence has been previously described, with most studies demonstrating decreasing continence rates with increasing complexity of malformation. [1, 3, 4] More complex ARMs result from an earlier arrest of normal embryologic development, and repair of these anomalies requires a more extensive procedure. [12-15] Our

study demonstrates that when grouped together by complexity of the malformation, those patients with simpler anomalies had a higher rate of continence (49%) compared to those with moderate (26%) or more complex (18%) malformations (p=0.005). However, the presence of associated sacral and spinal anomalies may also be contributing to impaired continence, as the rates of both have been demonstrated to be higher in more complex anomalies. [2, 5] In this larger study, we performed multi-variable modeling to evaluate the association of each of these factors. ARM subtype was the only independent predictor of continence.

Neither sacral hypoplasia nor spinal anomalies were found to be significant predictors of continence in our study. Sacral hypodevelopment is common in children born with ARMs, which suggests that the arrest that leads to the anorectal malformations often extends regionally to other developing pelvic structures. [6, 16] The sacral ratio was proposed in 1995 as a quantifiable measure of pelvic development, which serves as a marker of development of the pelvic musculature and nerves important to normal bowel control. [16] It has been shown that the sacral ratio correlates to continence rates, and patients sacral ratios of >0.70, 0.40-0.70, and <0.40 have described as having excellent, moderate, and poor continence potential, respectively. [17, 18] However, the effect of sacral ratio has not been independently assessed. For example, Torre et al. demonstrated that 48% of patients with a sacral ratio ≥0.52 were continent, compared to only 17% of patients with a sacral ratio <0.52. Although they identified a higher rate of spinal anomalies in the cohort of patients with a lower sacral ratio (75% vs. 32%), the effect of these findings were not independently assessed. And while imaging in both the AP and lateral views are important for an overall assessment of associated anomalies in patients with ARM, the former is an unreliable plane from which to calculate a sacral ratio, as the tilt of the pelvis may falsely affect this ratio. [2, 16] In our study, the sacral imaging technique was not standardized,

and thus many patients had sacral imaging in the AP plane but only 46 patients had measurements of a lateral sacral ratio available. In addition, many patients did not have dedicated sacral x-rays; therefore the sacral ratios were calculated from abdominal x-rays in which the sacral bones were included. The lateral sacral ratio was not associated with continence in our study; therefore, it is possible that our study lacked the power to detect an association between lateral sacral ratio and continence. The AP sacral ratio was associated with fecal continence on univariable analysis ($p=0.04$); however, the relationship was in the opposite direction of what has been previously reported. Furthermore, the AP sacral ratio did not remain predictive of continence in multivariable modeling. This suggests that ARM subtype either overshadows or accounts for the effect of sacral ratio on continence. These results suggest that routine sacral imaging for the purpose of counseling may be of limited value. However, given the small sample size and limitations of the present study, definitive recommendations as to whether sacral ratios should be measured routinely cannot be made based on this study alone. A larger study evaluating the additional value of sacral ratios calculated using dedicated sacral films for predicting continence is needed and is currently underway.

Similarly, spinal anomalies were not predictive of fecal continence in our study. Congenital anomalies of the caudal spinal cord are also seen in a high rate in patients with ARM and increase in incidence along with the severity of ARM. [5, 19] The S2-S4 spinal nerve roots which originate from this region of the spinal cord provide innervation to the distal bowel. In contrast to sacral anomalies, spinal lesions such as tethered cord present an opportunity for surgical intervention, although at present there is not good evidence to support detethering for the purpose of bowel function. [19, 20] Some studies suggest worse clinical outcomes in patients with associated spinal lesions, but these studies did not independently investigate whether the

spinal lesions themselves were associated with worse continence or if they are instead a marker of more abnormal development in the region which may also be captured by the type of ARM. [7] Our data suggests the latter, as patients with tethered cord, tethered cord or low conus, fatty filum, myelomeningocele, or normal spinal imaging had similar continence rates. Furthermore, in multivariable analysis that included the type of ARM, spinal anomalies were not independently associated with continence. This data suggests that like the sacral ratio, the influence of spinal anomalies on continence is either overshadowed or accounted for by the type of ARM.

Attaining fecal continence in ARM patients is important as it has been shown that incontinence adversely affects quality of life. [21-24] Notably, 40% of patients with perineal fistula in our study, regarded as the most minor ARM subtype with the highest continence potential, were incontinent. These results underscore the need for continued assessment of stooling behavior by those who care for these children. Our results suggest that fecal continence rates in ARM patients may be lower than previously reported. Children with ARM will likely benefit from long term follow-up and many may need additional bowel management to obtain continence.

After surgical repair of an ARM, patients often require significant medical management to achieve continence. In severe cases, this may take the form of a formal bowel management program (BMP) to find an adequate enema or laxative regimen to become socially continent. [25-28] While it is clear that anatomic features are important predictors of continence, it has been shown that aggressive bowel management can improve continence in these patients. The high rate of incontinence in our study supports the continued investigation of modifiable adjuncts to

achieve continence, including tailored enema or laxative regimens that empty the colon once daily and keep the patient clean of stool between discrete bowel movements.

We recognize several limitations to our study. First, we had a 42% response rate to follow up questionnaires, which may introduce selection bias into our study. Despite appearing low, this response rate for a survey study is not unexpected considering that the families being surveyed had their child treated a minimum of 4 years prior. Thus, we lacked current contact information for many families. The low response rate coupled with the lower than expected rate of continence supports the need for routine longitudinal follow-up of the patients as part of clinical practice. Second, this study is limited by the variable surgical and bowel management of patients with ARM across participating institutions. Our study did not include features of bowel management, which is institution-dependent and may play a significant role in helping children with ARM achieve continence. We also did not asses the effects of the type of pull-through or hospital volumes on continence. Third, the assessment of sacral ratio as a factor associated with continence was limited by a lack of dedicated sacral x-rays to calculate sacral ratios. The inter-rater reliability of calculating sacral ratios across a variety of x-ray types (sacral x-ray, babygram, abdominal x-ray) is unknown and is currently being investigated. Fourth, the association of laxative use and bowel management on continence may be influenced by selection bias due to the potential that families using these may have been more likely to respond to the surveys. However, we believe the likelihood of this is low given that there were no significant differences in important clinical characteristics between responders and non-responders to our survey. Lastly, we chose to use the Rome III definition of fecal incontinence which classifies patients as simply “continent” or “incontinent” based on a single stooling accident per week. We acknowledge that other definitions of continence can be used; however we do advocate that an

objective and consistent definition of continence should be used for the follow-up of children with ARM.

Conclusion

The type of ARM was the only factor identified early in life that predicted fecal continence in children born with an ARM. The high degree of incontinence, even in the least severe subtypes, highlights that predicting fecal continence is difficult at birth and supports the need for long-term follow-up and bowel management programs for children with ARM.

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Table Legends

Table 1: Cohort demographics and clinical characteristics.

Characteristic	Median (IQR) or N(%)
Age in days at first ARM-related surgery, median (Q1, Q3)	3 (1-98)
Gender, n (%)	
Male	81 (56)
Female	63 (44)
Race, n (%)	
White	111 (77)
Black/African American	18 (13)
Asian	4 (3)
Native Hawaiian or Other Pacific Islander	1 (1)
American Indian or Alaskan Native	1 (1)
Bi-racial/Multi-racial	3 (2)
Unknown	6 (4)
Type of ARM, n (%)	
Perineal fistula	53 (37)
Recto-prostatic fistula	8 (6)
Recto-bulbar fistula	9 (6)
Recto-vesical fistula	9 (6)
Recto-vestibular fistula	17 (12)
Cloaca < 3 cm channel	7 (5)
Cloaca > 3 cm common channel	9 (6)
Cloacal exstrophy	10 (7)
Imperforate anus without fistula/anal atresia	15 (10)
Pouch colon/rectal atresia	0 (0)
Rectal stenosis	3 (2)
Rectovaginal fistula	4 (3)
Number of patients per institution, n (%)	
Hospital 1	29 (20)
Hospital 2	23 (16)
Hospital 3	20 (14)
Hospital 4	17 (12)
Hospital 5	16 (11)
Hospital 6	14 (10)
Hospital 7	8 (6)
Hospital 8	8 (6)
Hospital 9	5 (3)
Hospital 10	4 (3)

Table 2: Continence rate by various clinical features.

Characteristic	Continent (N=58) N (%)	Incontinent (N=86) N (%)	**P**
Type of ARM			0.002
Perineal fistula	32 (60)	21 (40)	
Recto-prostatic fistula	1 (13)	7 (87)	
Recto-bulbar fistula	2 (22)	7 (78)	
Recto-vesical fistula	1 (11)	8 (89)	
Recto-vestibular fistula	8 (47)	9 (53)	
Cloaca < 3 cm channel	2 (29)	5 (71)	
Cloaca > 3 cm common channel	4 (44)	5 (56)	
Cloacal exstrophy	0 (0)	10 (100)	
Imperforate anus without fistula/anal atresia	6 (40)	9 (60)	
Rectal stenosis	0 (0)	3 (100)	
Rectovaginal fistula	2 (50)	2 (50)	
Severity of ARM			0.005
Highest risk (>3cm cloaca, cloacal exstrophy, rectovesical fistula)	5 (18)	23 (82)	
Moderate (recto-prostatic fistula, rectovaginal fistula, <3cm cloaca)	5 (26)	14 (74)	
Lowest risk (perineal fistula, recto-bulbar, recto-vestibular, imperforate anus without fistula/anal atresia, rectal stenosis)	48 (49)	49 (51)	
AP sacral ratio categories			0.04
≥0.7	19 (31)	43 (69)	
0.4-0.69	24 (53)	21 (47)	
<0.4	3 (60)	2 (40)	
Lateral sacral ratio categories			0.87
≥0.7	13 (39)	20 (61)	
0.4-0.69	5 (45)	6 (55)	
<0.4	1 (50)	1 (50)	
Spinal cord/Spine/Sacral findings			
Tethered cord or low conus	18 (43)	24 (57)	0.69
Tethered cord (not low conus)	11 (35)	20 (65)	0.54
Fatty filum	9 (43)	12 (57)	0.79
Myelomeningocele/spina bifida	3 (30)	7 (70)	0.74
Presacral mass	0 (0)	3 (100)	0.27
Spinal hemivertebra	3 (25)	9 (75)	0.36
Sacral dysplasia/hemisacrum	10 (34)	19 (66)	0.48
Sacral agenesis	3 (75)	1 (25)	0.30
None of these findings on either imaging exam	27 (41)	39 (59)	1.00

Table 3. Bowel management techniques used and their association with fecal continence.

Variable	Continent N (row %)	Incontinent N (row %)	P
Laxative use (N=127*)			0.48
Never	28 (49)	29 (51)	
Any (infrequent, every day, 1-2 times/day, > 2 times/day)	30 (43)	40 (57)	
Laxative use (N=127*)			0.53
Never/Infrequent	36 (48)	39 (52)	
All others (every day, 1-2 times/day, > 2 times/day)	22 (42)	30 (58)	
Enema use (N=127*)			<0.001
Never	47 (62)	29 (38)	
Any (infrequent, every week, every day, > 1 time/day)	11 (22)	40 (78)	
Enema use (N=127*)			<0.001
Never/Infrequent	52 (54)	43 (45)	
All others (every week, every day, > 1 time/day)	6 (19)	26 (81)	
Cecostomy flushes (N=125* as 2 patients did not answer)			0.11
No	54 (50)	55 (50)	
Yes	4 (25.0)	12 (75.0)	
Previous bowel management program participation (N=127*)			0.005
No	51 (53)	46 (47)	
Yes	7 (23)	23 (77)	

*The 17 patients with an ileostomy or colostomy at the time of the survey were excluded because they did complete the Baylor Scale which contains the answers to these questions.

Table 4: Multivariable model controlling for the severity of ARM and spinal cord findings on continence.

Variable	Odds Ratio (95% CI)	P-value
Severity of ARM (ARM subtypes)		
Highest risk (>3cm cloaca, cloacal exstrophy, rectovesical fistula)	ref	
Moderate (recto-prostatic fistula, rectovaginal fistula, <3cm cloaca)	2.5 (0.5-12.2)	0.87
Lowest risk (all others)	7.4 (1.9-28.7)	0.001
Spinal cord findings		
Tethered cord or low conus	1.6 (0.7-3.7)	0.32
Fatty filum	1.2 (0.4-3.4)	0.77
Myelomeningocele/spina bifida	2.3 (0.4-14.0)	0.36