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ORIGINAL ARTICLE

The importance of dedicated colorectal team participation in the management of spina bifida and spinal cord injury patients

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Abstract

Purpose In September 2020, the colorectal team of the International Center for Colorectal and Urogenital Care joined the spina bifida and spinal cord injury multidisciplinary clinic at Children's Hospital Colorado. Many important lessons were learned.

Methods A retrospective review of patients seen in the spina bifida and spinal cord injury multidisciplinary clinic from September 2020 to May 2021 was conducted. Data collected included demographics, diagnosis, pre or post-natal repair for those with myelomeningocele, whether the patient was previously seen by the colorectal team, wheelchair usage, voluntary bowel control vs. fecal incontinence, urinary control vs. clean intermittent catheterization, characteristics of contrast enema, and our proposed intervention.

Results Overall, 189 children were seen during the study period, ranging from 3 months to 20 years of age (average = 9.5 years). One hundred and two were males and 87 were females. Diagnosis included myelomeningocele ($n = 153$), spinal cord injury ($n = 18$), transverse myelitis ($n = 7$), sacral agenesis ($n = 5$), diastematomyelia ($n = 2$), spinal stenosis ($n = 2$), and tethered cord with lipoma ($n = 2$). Fifteen patients with myelomeningocele were repaired in-utero. One hundred and sixty patients were new to the colorectal team. Eighty-one patients were wheelchair users. One hundred and twenty-three patients suffered from fecal incontinence and needed enemas to be artificially clean for stool and thirty-eight patients had voluntary bowel movements and were clean with laxatives, suppository, or rectal stimulations. Twenty-eight patients were younger than three years of age and still in diapers. Despite a non-dilated colon on contrast enema, this population has a hypomotile colon. One hundred and twenty-eight patients required clean intermittent catheterization.

Conclusion Joining the spina bifida and spinal cord injury multidisciplinary clinic allowed us to better serve this population and gave us enormous satisfaction to contribute to improve the quality of life of the patients and their parents.

Level of evidence III

Keywords Spina bifida · Spinal cord injury · Pediatric colorectal

Introduction

Spina bifida (SB) is the most common congenital neural tube defect, and it occurs during gestational days 24 and 26 [1, 2]. Spina bifida results in the loss of normal motor and sensory control in the gastrointestinal tract and anorectal dysfunction. Fecal incontinence is a significant, life-altering sequela of SB and studies demonstrate that over 50% of SB patients in the United States suffer from bowel dysfunction [3]. While due to different pathophysiologic mechanisms, bowel function is often significantly altered in patients suffering from spinal cord injury (SCI). Over 98% of individuals with SCI report at least one bowel problem related to their SCI,

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and 40–60% state that bowel dysfunction adversely impacts day-to-day activities, lifestyle, and quality of life [2–6]. Managing bowel function in SB and SCI patients is one of the most important challenges to improve quality of life and promote social well-being in this patient population.

Beyond constipation and incontinence, bowel dysfunction in SB and SCI patients often results in secondary complications including worsening of urinary incontinence, urinary tract infections (UTI), shunt malfunction, skin breakdown, hemorrhoids/anal fissures, social discomfort, and loss of work activities [2, 7–9, 9, 13]. Reports from the first 10 clinics in the National Spina Bifida Patient Registry (NSBPR) identified males, non-Hispanic blacks, patients with higher lesion levels, and those with public insurance to be less likely to have bowel control. There is a need for a multidiscipline, proactive, systematic approach to bowel management in patients with SB and SCI.

Current bowel management guidelines from the Spina Bifida Society include education, dietary management (fiber and fluids), pharmalogical adjuncts (sennoside) and/or rectal stimulants (glycerin, docusate sodium, or bisacodyl suppositories) to manage constipation. However, there is no mention about utilization of abdominal radiographs to evaluate for stool load, no differentiation between constipation with bowel control and constipation with fecal incontinence, nor recommendations for finding an individualized enema solution that keeps the patient clean for stool prior to recommending an antegrade enema procedure (ACE).

Our pediatric colorectal team has had great success implementing bowel management in patients who suffer from constipation with bowel control [14] or fecal incontinence (with or without constipation) in patients with anorectal malformations and Hirschsprung's disease [10, 16, 17]. Due to the success of the program, patients with SB started to also benefit from it. Thus, the goal of this study was to evaluate the impact of dedicated pediatric colorectal team participation in the management of pediatric patients with SB and SCI at Children's Hospital Colorado. In September 2020, the pediatric colorectal team of the International Center for Colorectal and Urogenital care joined the spina bifida and spinal cord injury multidisciplinary clinic at Children's Hospital Colorado. The significant findings, lessons learned, and proposed interventions are detailed in the current manuscript.

Methods

Study setting

We conducted a retrospective review of all patients seen in the spina bifida and spinal cord injury multidisciplinary clinic at Children's Hospital Colorado from September 6, 2020 to May 18, 2021. Patients were included if they presented to the multidisciplinary clinic during the study timeframe. This study was approved by the Colorado Multiple Institutional Review Board (COMIRB).

Data collection and statistical analysis

The electronic medical record (EMR) was reviewed for each patient who presented to the spina bifida and spinal cord injury multidisciplinary clinic during the study period. Demographics and clinical data obtained included: age, gender, disease diagnosis, pre or post-natal repair for patients with myelomeningocele, wheelchair usage, voluntary bowel control vs. fecal incontinence, urinary control vs. clean intermittent catheterization, and whether antegrade enema procedure (ACE) or antegrade channel for bladder catheterization (Mitrofanoff) with or without bladder reconstruction was performed. If the patient had a contrast enema, it was reviewed by members of the research study team and its characteristics were documented. Data on whether the patient had been previously seen by the pediatric colorectal team were also recorded.

Specific outcomes of interest included: (A) Incidence of bowel continence and the type of lesion associated with patients who displayed bowel continence, (B) Incidence of new patients to the pediatric colorectal team, (C) Incidence of in-utero repair of myelomeningocele, (D) Incidence of wheelchair users, (E) Characteristic findings demonstrated on contrast enema (dilated colon vs. normal caliber colon), and (F) Incidence of urinary continence. Finally, we report our proposed intervention for bowel management for this patient cohort. Demographics and clinical data are summarized using means with standard deviation (SD) and median with first and third quartiles.

Results

A total of 189 children were evaluated in the spina bifida and spinal cord injury multidisciplinary clinic during the study period. The age of patients at the time of presentation ranged from 3 months to 20 years, with an average age of 9.5 years. There were 102 male patients and 87 female patients. The most common diagnosis seen in clinic was myelomeningocele ($n=153$) following by spinal cord injury ($n=18$), transverse myelitis ($n=7$), sacral agenesis ($n=5$), diastematomyelia ($n=2$), spinal stenosis ($n=2$), and tethered cord with lipoma ($n=2$).

Overall, the majority of patients in the spina bifida and spinal cord injury multidisciplinary clinic (84.7%, 160/189) were new to the pediatric colorectal team. When evaluating bowel function, 76.4% (123/161) suffered from fecal incontinence and required regular enemas to be

artificially clean for stool. Thirty-eight patients had voluntary bowel movements or behaved as having voluntary bowel movements, and were clean with either laxatives, suppositories, or rectal stimulation. Twenty-eight patients were younger than 3 years of age and still in diapers, therefore they were not able to be evaluated for bowel control. The level of the lesion for those with voluntary bowel control ranged from C1 to the sacrum, including six cervical lesions, one thoracic lesion, and thirty-one lumbar and/or sacral lesions. Of those with voluntary bowel control, ten patients were wheelchair users and twelve required clean intermittent catheterization to be dry of urine. Demographics and clinical characteristics for patients with voluntary bowel control are summarized in Table 1.

Of patients that were referred to our bowel management week after the colorectal team joined the spina bifida and spinal cord injury multidisciplinary clinic, 78.9% were

Table 1 Demographics and clinical data for spina bifida and spinal cord injury patients with voluntary bowel control

Patient	Gender	Age (yrs)	Level of the lesion	Wheelchair use	Urinary control
1	M	8.3	Meningocele	N	Y
2	F	19.2	Lipomeningocele	N	N
3	F	15.5	C4 Transverse myelitis	Y	N
4	F	14.4	ASIA spinal cord injury	N	Y
5	M	15.6	Cauda equina syndrome	N	Y
6	F	6.1	Lumbar myelomeningocele	N	Unknown
7	F	10.4	Diastematomyelia	N	Y
8	M	12.4	L5–S1 myelomeningocele	N	Y
9	F	8.7	L1–2 myelomeningocele	Y	N
10	M	10.9	Lumbosacral lipomyelomeningocele	N	Y
11	M	17.2	L3 Myelomeningocele	N	Y
12	F	16.3	Lipomeningocele	N	Y
13	F	8.1	L3 myelomeningocele	N	Y
14	M	14.5	L2–L3 myelomeningocele	Y	N
15	M	5.8	Cervical spinal cord injury	N	Y
16	F	15.7	C1 congenital stenosis	Y	Y
17	F	3.6	L5–S1 lipomyelomeningocele	N	Y
18	F	13.4	Lumbosacral myelomeningocele	N	Y
19	F	13.1	Lumbosacral lipomeningocele	N	Unknown
20	F	11.6	Lipomeningocele	N	Y
21	F	6.7	Lipomyelomeningocele	N	Y
22	M	12.2	S1 myelomeningocele	N	N
23	F	14.1	Lipomeningocele	N	Y
24	M	3.7	Lumbosacral lipomeningocele	N	Unknown
25	M	18.1	C7 ASIA spinal cord stroke	Y	N
26	F	14.6	L2–L5 lipoma	N	Y
27	F	10.1	Lumbosacral lipoma	N	Y
28	F	7.3	T10 terminal myelocystocele	N	Y
29	F	4.3	Lumbar meningocele	N	Y
30	F	14.2	L3 myelomeningocele	Y	N
31	F	7.4	T4 spinal cord injury	Y	N
32	F	6.4	C4–C5 anterior meningocele	N	Y
33	F	8.5	L1–L2 Myelomeningocele	Y	N
34	F	19.4	Partial sacral agenesis/diastematomyelia	N	Y
35	M	16.6	Lumbosacral lipomeningocele	Y	N
36	F	9.3	Lumbar myelomeningocele	N	Y
37	F	5.9	Sacral lipomyelomeningocele	Y	N
38	F	9.1	L4–L5 myelomeningocele	N	N

*Data are presented as means

starting the bowel management program at a delayed age (average = 8.9 years, ranging from 5 to 18 years).

Regarding the urinary tract, 128 children required clean intermittent catheterization to be dry of urine. Of those with voluntary bowel control, 60.5% (23/38) also had urinary control. Of those with urinary control, 88.5% (23/26) also had voluntary bowel control. Fifteen patients with myelomeningocele underwent in-utero repair, and 138 were repaired following birth. Eighty-one patients were wheelchair users, and of these 87.5% did not have voluntary bowel control. Contrast enemas were available for 54 patients. After review, findings were consistent with a non-dilated, redundant colon (Fig. 1), which differentiates this population from idiopathic constipation, anorectal malformation, and Hirschsprung disease that when suffering from constipation and hypomotility with or without bowel control, the distal colon gets very dilated. Antegrade continence enema (ACE) and antegrade channel for bladder catheterization (Mitrofanoff) with or without bladder reconstruction procedures were completed for 46 patients evaluated in the multidisciplinary clinic.

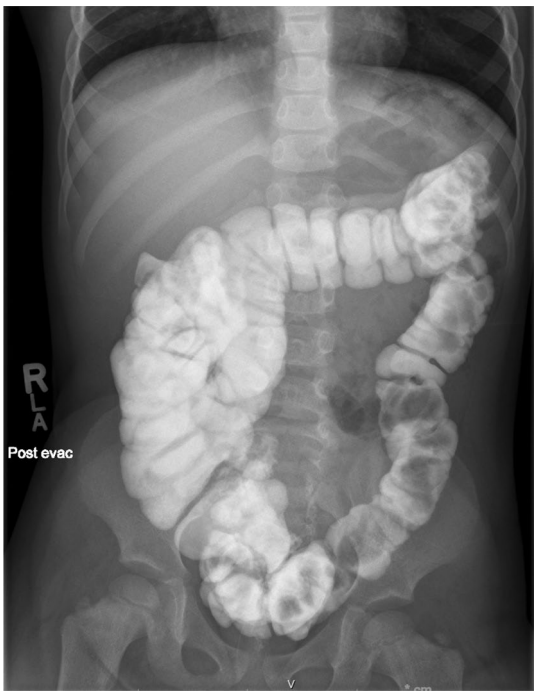

Fig. 1 Contrast enema of a patient with spina bifida showing characteristic image of a non-dilated, redundant colon, with signs of constipation (solid stool balls)

Discussion

Patients in the spina bifida and spinal cord injury multidisciplinary clinic at Children's Hospital Colorado have a variety of clinical diagnoses with myelomeningocele representing 81.0% of patients.

Even though our hospital has a dedicated pediatric colorectal team and is a referral center for congenital colorectal conditions, including bowel management to treat fecal incontinence and constipation, the spina bifida and spinal cord injury population was not being properly referred, since 84.7% of patients were new to the colorectal team, and in 50.3% of the new patients, the bowel management had to be changed. These findings emphasize the importance of having each specialty represented in multidisciplinary clinics that treat patients with spina bifida and spinal cord injury. The hope of dedicated colorectal team participation for this patient population is that children will be referred to our bowel management program at an appropriate age avoiding, for example, the case of a patient who only learned of the possibility of being clean in the underwear for stool when he was 18-years old.

Overall, 76.7% of patients who were beyond toilet trained age suffered from fecal incontinence requiring enemas. Finding the right individualized enema recipe that keeps patients artificially clean for stool in the underwear takes time and dedication. At Children's Hospital Colorado, every month, we dedicate one week for bowel management sessions. We start by ordering a contrast enema to understand the anatomy and characteristics of the colon. Based on this we can estimate the volume that the patient will need to clean the rectum and left colon, since the stool in the right colon will usually take 24 h to reach the rectum. In the spina bifida population, interestingly, all patients have a non-dilated and redundant colon, in spite of the fact that this patient population suffers from a hypomotile colon, we do not have an explanation for this. These patients need both a large volume and concentrated enema to be clean in the underwear. We use normal saline solution as the base of the enema, between 100 and 1000 ml, and liquid glycerin is used as our main irritant, between 10 and 90 ml. If the patient needs an even more concentrated enema to clean the colon, then we add castile soap between 10 and 90 ml. Fleet is used only as the last resource since, in this population, patients may also have kidney problems. For patients with very slow motility, we may add bisacodyl, right before enema administration, if the enema process (administration and evacuation) is taking more than one hour. Based on reports from patients and/or parents about response to the enema, the number and time of stool accidents, as well as the amount of stool seen on the abdominal radiograph, the enema is changed

until one is found that keeps the patient clean of stool in the underwear and the abdominal radiograph is clean [10, 16, 17].

In addition to formal bowel management to be clean in the underwear for stool, at the same age when other children are out of diapers, properly treating constipation and diaper rash early in life may put these children on the proper track. The abdominal radiograph is not mentioned as a tool to monitor or diagnose the degree of constipation early in life in the spina bifida guidelines. We believe the abdominal radiograph is an objective way to evaluate constipation. We have seen young patients in which parents and other clinicians thought the child was doing well due to regular bowel movements, but the abdominal radiograph on routine clinical visit, demonstrated that the patient was fecally impacted (Fig. 2).

Unfortunately, due to the small number of patients with reported bowel control, and the variation in the lesion levels identified in this group, this study has not allowed us to identify prognostic factors that determine the chances for bowel control in this population. Our impression is that the best predictor for bowel control in this population is having urinary control, since 88.5% of patients with urinary control also had concomitant bowel control. However future studies are needed to confirm these preliminary findings.

While bowel management guidelines for this patient population exist in the literature, to our knowledge, none of the current guidelines recommend abdominal radiographs as a component of bowel management strategies. Another area of confusion is using the term "continence" for patients that are actually artificially clean in the underwear and mentioning "constipation" with or without bowel control as the same problem. Patients that suffer from constipation and bowel control can be properly treated with oral laxatives while patients that suffer from constipation and fecal incontinence should be treated with enemas.

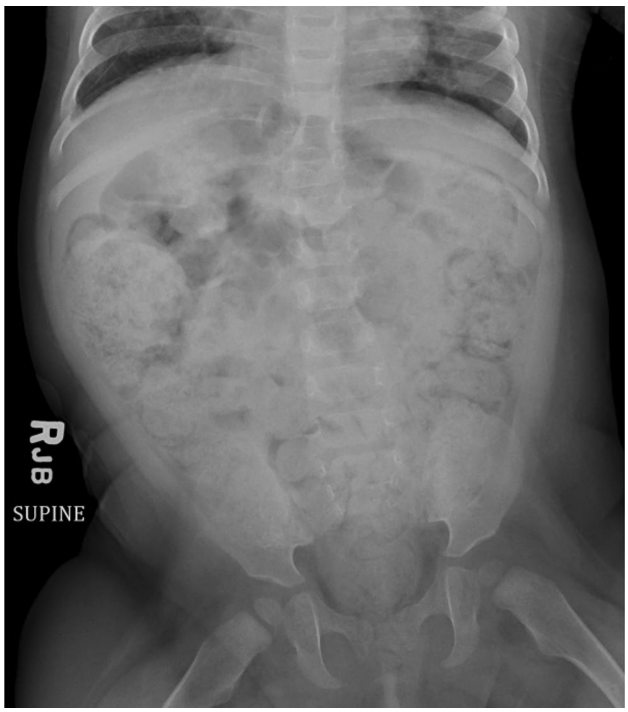

Fig. 2 Abdominal radiograph of one-year old, having daily bowel movements, showing severe accumulation of stool

Based on this experience, the importance of a dedicated colorectal team in the multidisciplinary clinic to care for patients with spina bifida and spinal cord injury cannot be overstated. Since most patients will suffer from neurogenic bladder and bowel, the collaboration between pediatric urology and colorectal surgery allows for combination of an anesthetic event when creating an antegrade enema procedure as well as an antegrade channel for bladder catheterization, but most importantly, optimizing their management prior to offering antegrade procedures is what determines success. Creating the antegrade enema procedure will not keep the patient clean in the underwear, however, finding the right enema recipe will make the difference. The antegrade enema procedure purely offers a more convenient way for administration of enemas.

Currently, patients with a diagnosis of spina bifida are seen in the spina bifida and spinal cord injury multidisciplinary clinic at 3, 6, 9, and 12 months of age following discharge from the Neonatal Intensive Care Unit (NICU). Patients are then evaluated in clinic every 6 months until they reach age 5, and then yearly afterwards. Based on this follow-up schedule and our experience of joining the multidisciplinary clinic, the pediatric colorectal team developed the following recommendations for bowel management in this cohort:

- An abdominal radiograph should be obtained at each clinic visit (to monitor the location and amount of fecal load)
- From age 3 months to 3 years constipation should be managed with MiraLAX or pedialax® (if the patient has constipation with a diaper rash), or large enemas if severe constipation is present and not responding to pedialax®, or if there was no improvement of diaper rash with pedialax®
- At age 3 formal bowel management with enemas should be started
- At approximately 5 years of age collaboration with urology should be coordinated to determine if the patient is a candidate for combined ACE + Mitrofanoff procedures with or without bladder augmentation/bladder neck reconstruction. The goal at this stage of the program is to be dry and clean in the underwear for urine and stool.

- Yearly follow up to determine if bowel management adjustments are needed

This paper has some limitations. This is a single-center, retrospective review and as such our findings may not be generalizable to all centers who care for patients with spina bifida and/or spinal cord injured patients. Future studies should be attempted evaluating the patient/parent's perspective since the colorectal team has joined the spina bifida and spinal cord injury multidisciplinary clinic, as well as evaluation of their quality of life before and after formal bowel management implementation.

Conclusions

Caring for children with spina bifida and spinal cord injury is complex, requiring a multidisciplinary and collaborative approach. The addition of the pediatric colorectal team to the Children's Hospital Colorado spina bifida and spinal cord injury multidisciplinary clinic allowed us to improve the quality of life of this patient population and contribute to a successful bowel management in such patients.

Declarations

Conflict of interest The authors have no conflicts of interest to report. No grant funding was used to support this research.

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