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Functional fecal and urinary outcomes after sacrococcygeal mass resection in pediatric patients

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ABSTRACT

Background: Sacrococcygeal masses (SCM) are uncommon in children. The purpose of this study is to review the functional fecal and urinary outcomes following resection of SCM and to determine the impact of a multidisciplinary clinic (MDC) on these outcomes.
Methods: A retrospective review was performed of patients who underwent SCM resection between 1979 and 2019. Baylor Social Continence Scale (BCS), Vancouver Symptom Score (VSS) and Cleveland constipation score (CSS) surveys were used to assess fecal and urinary continence at time of most recent follow up. Age, tumor characteristics, histopathology, and type of anorectal malformations (ARM), if present, were also recorded.
Results: 75 patients were included. 51 (69%) patients were females and 23 (31%) had an associated ARM. The median age at resection was 8.5 months (IQR 0–26.8). 41 (56%) patients were followed in the MDC. 27 (82%) of patients seen in the MDC were clean for stool and 26 (87%) were dry for urine, while only 17 (59%) of patients not seen in the MDC were clean for stool and dry for urine ($p<0.05$). There was improvement in Baylor, Vancouver and Cleveland scores.
Conclusions: A multidisciplinary approach to the care of patients following SCM resection may improve bowel and bladder outcomes.

Abbreviations

SCM	Sacrococcygeal masses
SCT	Sacrococcygeal teratomas
MDC	multidisciplinary clinic
AFP	Alpha fetoprotein
CIC	Chronic intermittent catheterization
BCS	Baylor Continence Scale
CCS	Cleveland Constipation Scoring System
VSS	Vancouver Symptoms Score
AXR	abdominal x radiation
IQR	Interquartile range
OR	Odds ratio
SSI	surgical site infection
ARM	anorectal malformation
SNS	sacral nerve stimulator
BMP	bowel management program
LUTD	lower urinary tract dysfunction
QoL	quality of life
ACE	antegrade continence enemas

1. Introduction

Sacrococcygeal masses (SCM) are rare lesions with unknown etiology. The majority of these tumors present at birth and arise from all three germ layers: ectoderm, mesoderm, and endoderm [1]. The most common of these tumors are mature sacrococcygeal teratomas (SCT), but immature or malignant tumors may also occur in this region. SCT has a prevalence of 1 in 14,000–28,000 and may present alone or as part of Currarino triad. This rare disorder was first described in 1981, and consists of the triad of a presacral mass, bony sacral defects (typically partial agenesis), and an anorectal malformation (most commonly anal stenosis) [2]. It has a familial association and has been associated with a mutation of the HLXB9 gene on chromosome 7q36 [3]. Most patients are diagnosed with presacral masses in infancy (Altman I) although some may not be diagnosed until much later in childhood (Altman IV) being identified by incidental imaging, detected during screening imaging in the setting of Currarino syndrome, or presenting with symp-

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toms such as constipation, urinary retention, or fecal incontinence secondary to mass effect from the tumor on adjacent anatomical structures [4,5].

While the primary treatment modality for SCM is surgical resection, these tumors pose diagnostic and therapeutic challenges due to their specific anatomic location and potentially difficult operative approach [1,2,6]. Due to complex involvement and potential disruption of adjacent neurovascular structures in the pelvis, the resection and reconstruction can often be challenging and may affect long term bowel and bladder functional outcomes [6-8]. This may be secondary to iatrogenic disruption of or chronic attenuation of structures in close proximity to the tumor, including the pelvic floor musculature and the pudendal and hypoglossal nerve plexus [4,5,9,10]. As the majority of SCM are mature teratomas, the oncological outcomes following resection are generally excellent [1,11-17]. However, the long term functional bladder and bowel outcomes after these complex resections is less well defined [5,7,17]. The purpose of this study is to review the functional outcomes following resections of SCM and to determine the impact that a multidisciplinary clinic (MDC) may provide for affected patients. We hypothesize that patients who attend MDC following SCM resection have improved functional fecal and urinary outcomes.

2. Methods

2.1. Patient population

We performed a retrospective review of our institution's colorectal and pelvic reconstruction MDC and surgical oncology clinic database for all patients who underwent SCM resection between 1979 and 2019. Patients with incomplete functional outcomes records were excluded. This study was approved by the Institutional Review Board at our institution (IRB STUDY00000440).

Data captured included gender, age at surgical resection, histopathology, chemotherapy received, duration of follow-up, time to tumor recurrence, and any occurrence of malignant degeneration. For those patients with an anorectal malformation (ARM), the anatomic subtype was recorded. Operative reports were reviewed to determine the procedure performed. Pathology reports were reviewed for the size and histology of the tumor. All preoperative alpha-fetoprotein (AFP) levels were drawn within 3 months pre operatively (some of these were in the newborn period). Since histopathologic diagnosis was not always known preoperatively, some of the AFP values are from patients who did not ultimately have an SCT. All post-AFP levels were drawn 3 months postoperatively or 3 months following completion of chemotherapy.

Patients were included in the study if the SCM resection occurred at our institution. Patients with pelvic tumors arising from the paraspinal region, such as meningoceles, were excluded. Patients were also excluded if they had incomplete medical records. All patients were followed in the surgical oncology clinic every 3 months after SCM resection for the first 2 years, then spaced to every 6 months until 5 years from resection. For the last 5 years, patients who have undergone resection of a SCM are routinely engaged in the MDC around toilet training age (18 months to 3 years of age) for evaluation and intervention as appropriate. The patients are then seen on annual basis in MDC or sooner if they need intervention. At each follow-up, functional fecal and urinary outcomes are assessed subjectively and objectively with surveys that include Vancouver, Baylor and Cleveland scores.

Bowel and bladder functional outcomes were captured from patient reports, chart information, and validated scoring systems. Urinary incontinence was defined as involuntary urine leakage in children older than 3 years of age. Chronic intermittent catherization (CIC) was defined as requirement for continued catherization but without leakage between catheterizations in patients with persistent urinary retention (>6 weeks post-operatively) [6]. Continence of stool was defined by the presence of voluntary bowel movements and ≤1 stooling accident per week as per Rome IV criteria [18].

2.2. Baylor, vancouver, cleveland scores

Standardized definitions and validated tools were used to assess fecal and urinary continence: the Baylor Continence Scale (BCS) (>4 years of age) [19], the Cleveland Constipation Scoring System (CCS) (>3 years of age) [20], the Vancouver Symptoms Score (VSS) (>3 years of age) [21]. BCS is a 23-question survey and is validated in children with ARM. BCS is completed by children or their parents using a psychometric response Likert scale. BCS scores ranges from 2 to 84, with lower scores reflecting better social continence. The VSS is a 14-item 5-point Likert scale questionnaire for which a score of ≥11 indicates dysfunctional elimination syndrome (DES) [21] and has been validated in the pediatric patients [22]. DES is defined as abnormal voiding habits associated with defecation symptoms. The CCS is a scoring system that was derived to obtain a universally objective definition of constipation. The score tabulated from a validated questionnaire ranges from 0 to 30, with 0 indicating normal and 30 indicating severe constipation [20] but has not yet been validated in pediatric patients.

2.3. Bowel management program (BMP)

The BMP is an outpatient program of 1-week duration for the purpose of intense management of constipation and/or fecal incontinence. A radiographic contrast enema to define relevant anatomy and an abdominal x-ray (AXR) are obtained prior to entering the program, which is then comprised of 3 outpatient clinic visits, 5 consecutive daily AXR's, and patient/parent reports of bowel function communicated by phone or email. Patients are placed on a medical regimen (stimulant laxatives for hypomotility or medications to slow motility for hypermotility) or a mechanical regimen (rectal enemas or antegrade flushes per cecostomy or Malone appendicostomy). The patient's bowel regimen is changed daily as needed based on the patient/parent functional report and assessment of the stool content on the AXR, with the goal of achieving social continence with an optimal bowel regimen. In addition, there is a psychosocial group for the patients and families during which they meet with the team of psychologists, social workers, and child life specialists. Following completion of this program, planned follow-ups occur at 3 and 12 months and annually thereafter.

2.4. Statistical analysis

Patient characteristics were reported as medians and interquartile ranges (IQR) for continuous variables and frequencies and percentages for categorical variables. Categorical variables were compared using the nonparametric chi-square test. Continuous variables were compared using the two-tailed Student's *t*-test where a paired *t*-test was performed to assess changes in the Baylor, Cleveland, and Vancouver scores. Bivariate logistic regression models were used to determine if tumor size, Altman classification, tumor recurrence, associated spinal anomalies, or associated ARM played a role in the patient's functional outcomes (clean for stool or dry for urine) – presented as odds ratios (OR) and corresponding 95% confidence intervals. Event-free survival (EFS) was calculated using the Kaplan-Meier method and measured as the time from initial operation until time of recurrence surgery, death, or last known follow-up if no event. A significance threshold of $p < 0.05$ was utilized for all tests. All statistical analyses for this study were per-

Table 1
Demographics and associated anorectal malformations.

Total Patients	$n = 75$
Male	24 (32%)
Female	51 (69%)
Current median age (yrs, IQR)	9.5 (6–15.5)
Anorectal Malformation (ARM)	23 (31%)
ARM Type	
Anal stenosis	12 (52%)
Perineal fistula	5 (22%)
Rectourethral fistula	3 (13%)
Cloaca	2 (9%)

formed using Microsoft Excel and Stata 14.0 (StataCorp LP, College Station, Texas, USA).

Since analyses were univariate in nature, no adjustments were made for multiple comparisons, and due to the retrospective nature of the study no adjustments were made to control for the false discovery rate. Missing data were simply treated as missing – i.e. complete-case analyses were performed for variables in which missing data were present. Logistic regression models were bivariate, thus no model selection methods or use of interaction terms were necessary nor were concerns with collinearity. These unadjusted bivariate models were to explore general associations between a single independent variable and the outcome, not prediction, thus the use of ROC curves and model validation techniques were not appropriate. A footnote was added to the tables to clarify that the logistic regression models were unadjusted."

3. Results

3.1. Patient characteristics

Between March 1979 and October 2019, we identified 75 patients with a diagnosis of a SCM. Fifty-one (69%) patients were female. The median age at resection was 8.5 months (IQR 0–26.8 months). Twenty-three (31%) patients had an associated ARM, with the most common being anal stenosis ($n = 12$, 52%) followed by perineal fistula ($n = 5$, 22%), rectourethral fistula ($n = 3$ (13%), and cloaca ($n = 2$, 9%) (Table 1).

3.2. Tumor characteristics

The most common radiological method of tumor diagnosis was MRI, which also included fetal MRI for prenatally identified lesions ($n = 29$, 39%). The median tumor size for the cohort was 4 cm (IQR 2.6–7 cm). Fifty-six (76%) patients had grade 0 pathology with mature tissue only. One patient (1%) had grade 1 pathology with rare (<10%) foci of immature tissues. Ten (14%) patients had grade II pathology with moderate (10–50%) quantities of immature tissues. Three (4%) of patients had grade III pathology with large (>50%) quantities of immature tissue and 3 (4%) patients had malignant yolk sac elements. Four (5%) patients had neuroblastoma. For the patients with SCT, the most common Altman classification was type IV ($n = 20$, 27%), followed by type I ($n = 11$, 15%), type II ($n = 8$, 11%) and type III ($n = 3$, 4%) (Table 2). Sixty-one patients had an SCT but 19 patients did not have Altman class defined. The remaining histologies included neuroblastoma ($n = 4$), benign epithelial cyst ($n = 4$), tailgut duplication cyst ($n = 2$), rhabdomyosarcoma ($n = 1$), intramedullary lipoma ($n = 1$), ganglioneuroma ($n = 1$), and schwanomma ($n = 1$).

3.3. Oncological outcomes

Of 75 patients, 3 patients died. One died secondary to intraoperative exsanguination, and 2 died from progressive metastatic disease. The median preoperative AFP levels were 681 ng/ml (IQR 2.9–87,816 ng/ml) and the median postoperative AFP levels were 2.7 ng/ml (IQR 1.15–6.15 ng/ml) for all patients in this cohort. Eleven (15%) developed a recurrence with an average time to recurrence of 3.07 years. Seven (9%) received adjuvant chemotherapy and 16 ($n = 21$%) underwent reoperations (Table 2). 11 patients developed recurrence but 2 patients developed multiple recurrences and required multiple reoperations. The 5-year EFS for the entire cohort was 78.5% (95% CI: 67.6–91.1%).

Table 2
Tumor diagnosis and oncological characteristics.

Diagnostic imaging	
MRI	29 (39%)
X-Ray	7 (9%)
Ultrasound	6 (8%)
CT	3 (4%)
CT+MRI	2 (3%)
Altman type	
I	11 (15%)
II	8 (11%)
III	3 (4%)
IV	20 (27%)
Pathology	
Grade 0 pathology (only mature tissue)	56 (76%)
Grade I (rare foci of immature tissues)	1 (1%)
Grade II (moderate quantities of immature tissues)	10 (14%)
Grade III (large quantities of immature tissue).	3 (4%)
Neuroblastoma	4 (5%)
Mass type	
Solid	30 (40%)
Cystic	40 (53%)
Pre op AFP levels (median)	681 ng/mL (IQR 2.9–87,816)
Post op AFP levels (median)	2.7 ng/mL (IQR 1.15–6.15)

3.4. Functional bowel and bladder outcomes

The patients who followed up in the MDC were a median of 5 years from surgical resection with IQR 3–9 years, and the patients who did not follow up in the MDC were a median of 10 years from surgical resection with IQR 6–15 years. For the patients who followed up in the MDC, 2 (5%) had surgical site infection (SSI), 2 (5%) had fistulas (1 rectovaginal and 1 urethrovaginal), 2 (5%) had gait problems and 11 (27%) had neurogenic bladder. In comparison, for the patients who did not follow up in the MDC, 1 (3%) had SSI, 1(3%) had urethral stricture, 4 (13%) had gait problems, and 5 (16%) had neurogenic bladder.

Seventy-two patients were included in the functional bowel and bladder outcomes analysis. Forty-one (56%) were followed in the colorectal bowel management MDC. For all cases, standard posterior sagittal approach is used for resection of the tumor and reconstruction is done in collaboration with colorectal surgeons. Our practice for the last 5 years is to routinely engage patients who have undergone resection of a SCM into the MDC around toilet training age (18 months to 3 years of age) for evaluation and intervention as appropriate. The patients are then seen on annual basis in MDC or more frequently as required. All the patients over the age of 6 years of age who are fecally incontinent despite maximum medical treatment are referred for pelvic floor physical therapy.

The patients who were followed in the MDC were more complex than those who were not followed in the MDC: more likely to have an associated ARM [$n = 21$, 51% (95% CI: 35–67%) vs. $n = 2$, 6% (95% CI: 1–21%); $p<0.05$], more likely to have a tethered cord and/or sacral anomalies [$n = 27$, 66% (95% CI: 49–80%) vs. $n = 7$, 23% (95% CI: 10–41%); $p<0.05$], and more often required reoperation for recurrence [$n = 13$, 32% (95% CI: 18–48%) vs. $n = 2$, 6%

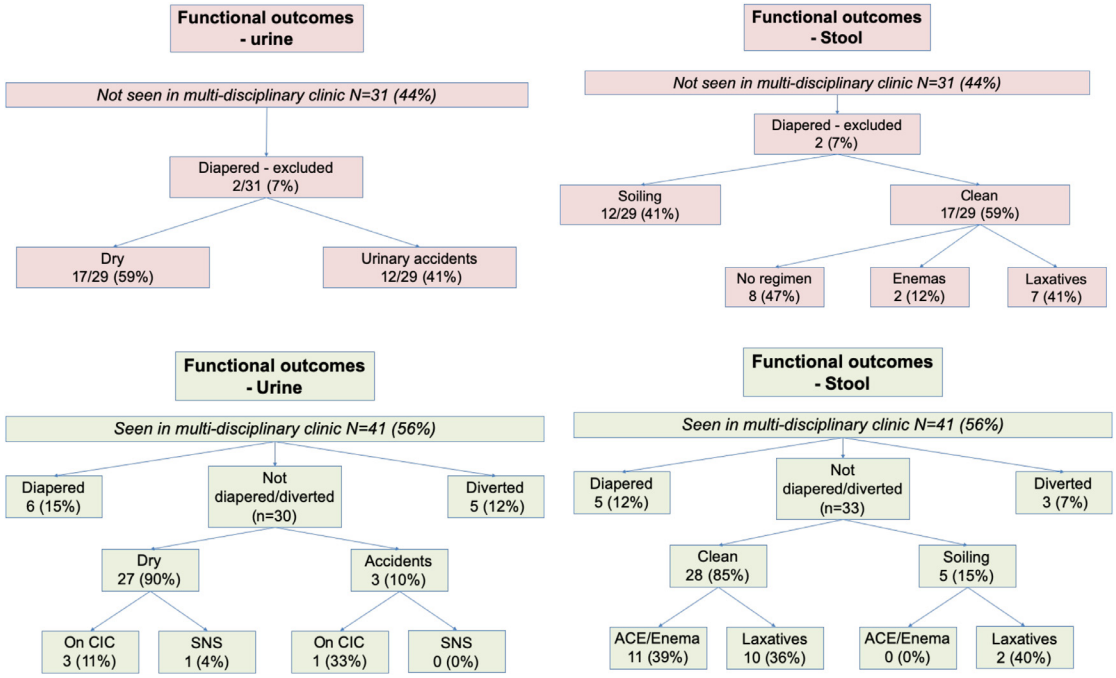

Fig. 1. Flowchart diagrams representing functional stool and urine outcomes in patients who attended multidisciplinary clinic vs those who did not. The patients who attended multidisciplinary clinic were more likely to be clean for stool and urine ($p<0.05$). *ACE=Antegrade continence enema. CIC=Chronic intermittent catheterization. SNS=Sacral nerve stimulator.

(95% CI: 1–21%); $p<0.05$]. Patients who followed up in MDC were less likely to be lost to follow up [$n = 2$, 5% (95% CI: 1–17%) vs. $n = 9$, 29% (95% CI: 14–48%); $p<0.05$].

For the urinary outcomes of patients who are followed in the MDC, 6/41 (15%) are diapered (not yet of age to toilet train) and 5/41 (12%) are diverted. Excluding those who are diapered or diverted ($n = 11$), 27/30 (90%) of those followed in the MDC are dry for urine, and 3/30 (10%) are having daily urinary accidents. Of the patients who are dry for urine, 3/27 (11%) require clean intermittent catheterizations, and 1/27 (4%) has a sacral nerve stimulator (SNS) in place. Of patients not seen in the MDC, 2/31 (7%) are still diapered. Excluding the 2 who are diapered, 17/29 (59%) are dry for urine, while 12/29 (41%) are having daily urinary accidents. Thus, the patients seen in MDC are more likely to be dry for urine compared to patients who were not followed up in MDC (90% vs. 59%, $p<0.05$).

For functional stooling outcomes of patients who were seen in the MDC, 5/41 (12%) are diapered and 3/41 (7%) are diverted. Excluding those who are diapered or diverted ($n = 8$), 5/33 (15%) met the Rome IV criteria for soiling (>1 stooling accident/week), and 28/33 (85%) are clean and continent. Of the patients who are clean, 11/28 (39%) are clean on either antegrade or rectal enemas, and 10/28 (36%) are clean on oral laxatives. Of the patients who are clean in the MDC group, 11/28 (39%) attended the formal bowel management program (Fig. 1). Of patients not seen in the MDC, 2/31 (7%) are diapered, 12/29 (41%) met the Rome IV criteria for soiling (>1 stooling accident/week), and 17/29 (59%) are clean and continent. Thus, the patients seen in MDC are more likely to be clean for stool compared to patients who were not followed up in MDC (85% vs. 59%, $p<0.05$). Of the patients who are clean, 8/17 (47%) are clean without any bowel regimen requirements, 2/17 (12%) are clean on rectal enemas, and 7/17 (41%) are clean on oral laxatives.

Tumor recurrence, presence of spinal anomalies, associated ARM, chemotherapy, tumor dimension and Altman classification did not play a significant role in the functional bowel and urinary outcomes in this small cohort (Table 3).

Of patients who were followed in the MDC and attended the bowel management program, a significant improvement in Baylor scores was noted (22.7 to 15.8, $p<0.05$), but no significant differences in Vancouver scores (12.85 to 10.44, $p = 0.18$) or Cleveland scores (11.06 to 9.36, $p = 0.07$) were detected.

4. Discussion

The primary purpose of this study was to evaluate the functional bowel and bladder outcomes in patients who underwent SCM resection at our institution. We hypothesized that patients who regularly attend a multidisciplinary clinic (MDC) with intense focus on bowel and bladder management following tumor resection will have improved urinary and rectal function compared to the patients who do not. We found that despite being more surgically and medically complex in nature, the patients who attended the MDC following resection had statistically significant improved bowel and bladder functional outcomes, reflected in Baylor score improvement. BCS has been validated and provides an objective measure of fecal continence in children with ARMs [19]. It is also a helpful tool to prospectively study interventions in children with incontinence after change in bowel regimens and/or BMP. Although we saw an improvement in VSS after the patients with SCM resection follow up in MDC, this did not reach statistical significance. Since there are currently no clinical guidelines regarding the long term follow up of the patients who undergo SCM resection, we recommend that all patients should be referred to a MDC following tumor resection for bowel and bladder evaluation and management, and optimization of outcomes.

While the oncological outcomes of patients after SCM resection have been reported with excellent results [1,11-17], less is known about the long-term functional outcomes. Cozzi et al. retrospectively reviewed 18 patients who underwent SCT resection and interviewed and compared 13 of these surviving patients with focus on functional outcomes [23]. They reported improvement in functional outcomes as patients age and recommend conservative management with minimal interventions as well as life long follow-

Table 3
Bivariate logistic regression models showed that tumor recurrence, tumor size, Altman classification, chemotherapy, associated spinal anomalies and ARM did not play a significant role in the patient being clean for stool and dry for urine.

Characteristic	Clean for stool ($N = 44$)	Not Clean for stool ($N = 11$)	OR (95% CI)	p-value
Recurrence	6 (13.6)	3 (27.3)	0.42 (0.09–2.05)	0.28
Spinal Anomalies	12 (27.3)	6 (54.6)	0.31 (0.08–1.22)	0.09
ARM	13 (29.6)	3 (27.3)	1.12 (0.26–4.90)	0.88
Chemotherapy	4 (9.1)	1 (9.1)	1.00 (0.10–10.00)	0.99
Max Tumor Dimension[1]	4.0 (0.6–20.0)	3.5 (2.6–10.0)	1.08 (0.89–1.30)	0.43
Altman[1]	4 (1–4)	1 (1–4)	1.64 (0.84–3.21)	0.15
Data are presented as N (%) or Median (Range) [1]Missing data present				
Characteristic	Continent for urine ($N = 43$)	Not Continent for urine ($N = 7$)	OR (95% CI)	p-value
Recurrence	6 (14.0)	3 (42.9)	0.22 (0.04–1.22)	0.08
Spinal Anomalies	12 (27.9)	3 (42.9)	0.52 (0.10–2.66)	0.43
ARM	13 (30.2)	1 (14.3)	1.12 (0.26–4.90)	0.88
Chemotherapy	3 (7.0)	2 (28.6)	0.19 (0.03–1.41)	0.10
Max Tumor Dimension[1]	4.0 (0.6–20.0)	3.4 (2.6–6.5)	1.12 (0.86–1.45)	0.41
Altman[1]	4 (1–4)	1.5 (1–4)	1.67 (0.72–3.89)	0.23
Data are presented as N (%) or Median (Range) [1]Missing data present				

OR: Odds ratio, CI=Confidence Interval.

up. Our study is unique in that patients evaluated in the MDC are monitored longitudinally and will continue to be followed at least annually with patient visits and validated surveys, resulting in decreased recall bias and a more accurate assessment of function. We also anticipate reporting longer term data and potential changes in functional outcomes of these patients as they age. With this longitudinal data, we will be better able to counsel families regarding the anticipated challenges and expected functional outcomes following SCM resections. An additional benefit of MDC for these patients is the routine involvement of pediatric and adolescent gynecologists. Through annual examinations, these providers can counsel patients, and will allow us to not only understand functional bladder and rectal outcomes but long term sexual outcomes following SCM resection as well.

A recent study by Bai et al. demonstrated that improving the quality of life (QoL, social, physical and psychological functioning) in patients with ARMs is not only dependent upon successful surgical interventions but also on bowel training programs [24]. Validated questionnaires such as BCS and VSS provide an objective measure of such interventions, and MDCs provide supportive structure for bowel training programs. Other questionanaires including CCS, though not yet validated in pediatric patients, have been used in adults to study functional outcomes with excellent correlation with clinical outcomes [20]. At our institution, all three scores are obtained from patients who have undergone resection of a SCM at regular intervals, and the trends of these scores are followed over time. Interventions such as the change in a bowel regimen or participation in a formal BMP demonstrated a positive trend in improvement in all three scores.

Voiding dysfunction following SCM resection is likely multifactorial, secondary to direct mass effect, sequelae related to surgical resection, or related to associated anomalies [25]. Urodynamic studies have been shown to be abnormal in patients following resection of SCM in 67–79% of cases, but may not always correlate with clinical symptomatology [26,27]. Rehfuss et al. found that 51% of patients with SCM had subsequent lower urinary tract dysfunction (LUTD) and advocated for the early involvement of the multidisciplinary team, including urologists, in the management of such patients [6]. Similar to our findings, patients with LUTD in Rehfuss' study had comparable tumor size and histology to the patients who did not have LUTD, highlighting the challenge of properly identifying all affected patients. The involvement of a multidisciplinary team including urologists following SCM resection, and even screening patients for the presence of hydronephrosis or other subclinical evidence of LUTD provides great value in identifying and intervening in patients early with postoperative bladder dysfunction. Furthermore, we noted an improvement in VSS in patients with DES after SCM resection in patients who followed up in the MDC, although this did not reach statistical significance.

Most prior reports that have evaluated long-term functional outcomes for patients with SCM have been relatively small retrospective case series [23,25,26,28]. A small retrospective study reported excellent functional outcomes (13/14 had fecal and urinary continence) who underwent SCT resection, however none of these patients had recurrences, only one had a malignant tumor and only one had associated spinal anomalies [28]. A larger multicenter study from Swedish pediatric surgical centers reported bowel and urinary tract functional outcomes and health-related quality of life (QoL) for 85 pediatric patients who underwent SCT resection [7]. Their analysis demonstrated that urinary tract dysfunction correlated with immature histology and that bowel dysfunction correlated with tumor size, a trend we did not identify in our patient cohort [7]. A larger study of 148 patients from Netherlands reported soiling in 13.2% of patients post SCT resection [29], comparable to rates of patients who followed in our MDC (15%). However, they did not use standardized Rome IV criteria for defining incontinence. They also reported urinary incontinence of 30.7% compared to 10% of our patients followed in our MDC. Our MDC and BMP involve nurses, advanced nurse practitioners, colorectal surgeons, urologists, and gynecologists. Despite the increased complexity of patients evaluated in our MDC (more associated ARM, more likely to have recurrence, and more associated spinal anomalies), the majority of these patients were able to be rendered clean for stool and dry for urine. The 11 patients who continued to struggle with fecal incontinence underwent a formal BMP and are currently clean, 6 of them are on antegrade continence enemas (ACE). Of the patients requiring ACE, 4 have associated ARMs (perineal fistula, anal stenosis, rectourethral fistula and cloaca) and 4 have spinal anomalies. Once patients are clean and dry, follow up continues and BCS, CCS, and VSS are obtained on an annual basis. In addition, when patients are of appropriate age, they continue to receive care in a transition clinic with providers trained in complex colorectal and pelvic reconstruction in both pediatric and adult patients, with expertise in managing the diverse needs of patients who have undergone complex reconstructions in childhood.

There are several limitations to this study evaluating the long-term functional outcomes of patients with SCM. First, given the rarity of such tumors it is challenging to capture large numbers of patients to evaluate. Therefore our analyses run the risk of sampling error and bias. The patients are more likely to follow up in the MDC clinic if they have associated anorectal malformation. They are also more likely to follow up if they have aggres-

sive tumor pathology and require multiple interventions and/or chemotherapy. Second, our cohort is unique in that the patients are more complex with associated anorectal malformations, and a larger number of paitents in this cohort have Altman type IV SCTs, likely explaining the advanced median age at resection of 8.5 months compared to other series. This also highlights the type of patients being referred to a high-volume center such as ours, and therefore impacts the applicability of these results on a broader scale. Third, the retrospective nature of the study is limited to the information obtained the electronic medical record at the time of the review. Finally, the Cleveland constipation scores utilized in this study for assessing functional outcomes have not yet been validated in pediatric patients, and so conclusions regarding these scores are challenging to draw in this cohort.

Future directions include obtaining disease specific quality of life scores, including social, physical, functional, cognitive, psychosocial, and emotional, in all patients who have undergone SCM resection [30]. While we currently do not collect sexual functional outcomes surveys in our MDC, this is an important area to pursue data in patients as they age. Finally, larger prospective studies are certainly needed to determine the potential role of routine screening for bowel dysfunction with anorectal manometry and bladder dysfunction with ultrasound and/or urodynamics in patients following SCM resection.

5. Conclusion

Sacrococcygeal mass resection can have a significant impact on functional bowel and bladder outcomes. Care in a multidisciplinary clinic with expertise in colorectal and urinary performance may improve the functional outcomes in such patients undergoing complex resection and reconstruction.

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