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Spinal column shortening for secondary tethered cord syndrome: radiographic, clinical, patient-reported, and urodynamic short-term outcomes

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OBJECTIVE Tethered cord syndrome (TCS) is a clinical and radiographic diagnosis of pathological stretching of the spinal cord leading to progressive loss of neurological function. The gold standard treatment for TCS is a tethered cord release. However, detethering involves significant risks of spinal cord injury and high rates of retethering. To mitigate these risks, the concept of spinal column shortening (SCS) to decrease spinal cord tension has become an alternative to detethering. In this study, the authors applied SCS to a pediatric and emerging adult population affected by secondary TCS.

METHODS A retrospective review of a prospective database at the authors' tertiary pediatric institution was performed. The Pediatric Quality of Life Inventory, patient- and parent-reported outcomes, and urodynamics were used to evaluate the outcomes of TCS treated with SCS.

RESULTS A total of 41 patients with secondary TCS were treated with SCS. The average age at the time of surgery was 15.9 years (range 5–55 years). Preoperative symptoms evaluated included pain (33 patients), weakness (30 patients), and bladder/bowel dysfunction (39 patients). The most common level of spinal column osteotomy was T12, with spinal fusion between T10 and L2. The mean follow-up time was 22.6 months (range 8–45 months). For patients with at least 12 months of follow-up, subjective clinical improvements were reported in 21/23 (91.3%) of those with preoperative pain ($p < 0.01$); in 16/24 (66.7%) of patients with weakness ($p < 0.01$), and in 15/29 (51.7%) of those with bladder/bowel dysfunction ($p < 0.01$). The median differences in initial and most recent Pediatric Quality of Life Inventory results were +5 for patient-reported scores ($n = 19$, $p = 0.04$) and +5 for parent-reported scores ($n = 19$, $p = 0.08$). Formal urodynamics performed at a median of 3.5 months after surgery documented stable to improved bladder function in 16/17 patients, with a median improvement in one classification category ($n = 17$, $p = 0.01$).

CONCLUSIONS SCS continues to represent a safe and efficacious alternative to traditional spinal cord untethering for TCS in children and emerging adults, as documented by objective formal urodynamics and patient- and parent-reported outcomes.

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KEYWORDS lipomyelomeningocele; myelomeningocele; spina bifida; spinal column shortening; tethered cord syndrome; spine

Tethered cord syndrome (TCS) is clinically defined by progressive loss of neurological function caused by pathological longitudinal stretch of the spinal cord. The most common neurological symptoms include leg pain and weakness; back and perineal pain; scoliosis; foot deformities; and sensory changes such as numbness or tingling, leg spasms, and bladder and bowel dysfunction.[1–6] Current knowledge suggests a pathophysiology of TCS involving decreased blood supply, altered spinal cord metabolism, and mechanical damage to the cord via abnormal cord attachments causing longitudinal stretch.[7] For spinal cord neurons, this cascade results in hypoxic stress and deterioration of oxidative metabolism leading to electrophysiological injury. Continued and accentuated neuro-

ABBREVIATIONS BMP = bone morphogenetic protein; EBL = estimated blood loss; MCID = minimum clinically important difference; PedsQL = Pediatric Quality of Life Inventory; SCS = spinal column shortening; TCR = tethered cord release; TCS = tethered cord syndrome; UTI = urinary tract infection.

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nal impairment can eventually induce structural damage to the neurons.

With TCS, a multitude of congenital and acquired etiologies can cause abnormal attachments of the caudal spinal cord to surrounding structures, thus anchoring the spinal cord within the lumbosacral region. These abnormal attachments can result from a spinal cord tumor, elongated cord, tight terminal filum, myelomeningocele, lipomyelomeningocele, or another spinal dysraphism.[7,8]

The current gold standard treatment for TCS is a tethered cord release (TCR) procedure that aims to remove abnormal attachments to the spinal cord, thus freeing the cord and alleviating tension.[2,6,9–11] Release procedures are a proven treatment for neurological pain, with improvement seen in an estimated 80% of patients.[3,12,13] To a lesser extent, release procedures are an effective treatment for subjective bladder function, with improvement seen in an estimated 50% of patients.[3,12,13] Although untethering procedures are effective, they come with significant risks, including a high rate of retethering and recurrence of neurological symptoms.[14] Secondary TCS in particular has been reported to have exceptionally high retethering rates. After initial release, the retethering rate of myelomeningoceles is approximately 3%–32% and can occur in up to 40% of patients after untethering of lipomyelomeningocele.[6,15] A primary cause for retethering may be natural scar tissue formation from previous surgical dissection into the dura mater. With each additional TCR, risks increase for infection, wound dehiscence, CSF leakage, arachnoiditis, and nerve root injury.[12] The increased difficulty of successfully untethering the spinal cord combined with the increased risk of neurological damage results in diminishing returns with each subsequent untethering procedure.[6,12,16]

With these established shortcomings of standard untethering, additional inquiry has been made into new treatment options for recurrent TCS to improve the limitations and technical challenges of repeated untethering procedures. Spinal column shortening (SCS) osteotomy has emerged as a potential alternative to direct untethering. SCS adequately alleviates tension on the spinal cord by decreasing the distance that the spinal cord is longitudinally stretched.[16–20] SCS bypasses the issue of retethering by eliminating the need to dissect the dura and directly manipulate neural elements, thus avoiding the formation of new intradural scar tissue. Although SCS has been shown to be safe and efficacious,[21] this study aims to evaluate clinical, radiographic, urodynamic, and patient- and parent-reported outcomes in a pediatric and emerging adult population treated with SCS.

Methods

Patient Population

We obtained and reviewed records of pediatric patients (< 18 years old) and adults (≥ 18 years old) with pediatric spinal disorders (i.e., patients with childhood spinal disorders who have entered adulthood and experience chronic medical issues due to their childhood disorder) who were treated with SCS for secondary TCS at our comprehensive tertiary pediatric center. Institutional review board approval was obtained for this retrospective chart review.

Surgical Indications

All patients showed signs and symptoms of progressive secondary TCS due to a history of myelomeningocele, lipomyelomeningocele, or prior TCR. Leg or foot weakness, back and leg pain, gait ability, and/or bowel and bladder dysfunction (defined as new leakage between catheterizations, febrile urinary tract infections [UTIs], or social incontinence) were evaluated as signs and symptoms. Furthermore, 34/41 (82.9%) patients underwent urology evaluation with formal urodynamics to document bladder function prior to surgery. The Pediatric Quality of Life Inventory (PedsQL) was administered to patients younger than 18 years of age and to their parents, when applicable, prior to surgery.

MRI was performed preoperatively on all patients in whom a low-lying conus, syringomyelia, or a terminal/transitional type of lipoma by Chapman classification was indicated.[22]

A classic untethering procedure was offered to each patient and his/her caregivers in addition to the SCS approach. Each treatment option was thoroughly discussed with patients, parents, and caretakers—including risks, benefits, possibility for additional surgery in case of failure of the procedure to resolve symptoms, and the lack of published data on the long-term outcomes of SCS, particularly in children. In equivocal cases, if the senior surgeon (A.J.) was asked for his opinion by patients, parents, or caregivers, he would favor SCS over traditional untethering based on admittedly weak evidence from the literature or anecdotal experience. Additionally, our indications conference discussed each case, and a panel of four other board-certified/board-eligible pediatric neurosurgeons deemed that the management plan was acceptable prior to proceeding with surgery, especially for patients in whom there had been no prior attempts at standard untethering.

Surgical Procedure

The SCS procedure used in this study has been previously described.[16,17] Vertebral column resection was the osteotomy approach used in all patients in this study. The most salient points of our surgical technique are included below.

A 6-mm diamond drill bit was used to drill the pedicle carefully, leaving a medial eggshell-thin rim of cortical bone to protect the dura. The size of the osteotomy gap was estimated using the width of the diamond drill bit, which was approximately 1.5 cm. This technique was used to continue drilling into the vertebral body toward midline; the anterior longitudinal ligament and the superior and inferior endplates were left intact. The nerve root one level below the level of the osteotomy was left skeletonized. After completing this process on one side, a temporary rod was placed on that side and the same process was completed on the contralateral side.

To allow the force of closure to be evenly dispersed through all the screws, segment rods were positioned across the 2 pedicle screws above and below the osteotomy defect. The screws securing the temporary rod were loosened to allow for use of a compressor to close the defect in a controlled manner. After complete closure, the

temporary rod was secured in place and the short rods were replaced by a unitized permanent rod.

Bone morphogenetic protein (BMP) was used "off label" to facilitate arthrodesis in this pediatric study. We acknowledge that the use of BMP in children is controversial. Whereas in-depth analysis of the risks and benefits of recombinant human BMP-2 use in children is outside the scope of this study, we have discussed this topic in previously published work.[23–26]

Outcomes

The primary outcome for this study was the need for reoperation due to retethering or surgical complications after SCS. Secondary outcomes included evaluation of spinal fusion as evident by solid arthrodesis on postoperative CT scans obtained at 12 months; subjective clinical symptoms (including pain, weakness, gait ability, bladder/bowel symptoms such as leakage between catheterization, febrile UTIs, and social incontinence); patient- and parent-reported outcomes (i.e., PedsQL); formal urodynamics; and the need for additional urological procedures. Considering recovery and rehabilitation time after surgery, only patients with at least 12 months of follow-up were included in the analysis of clinical symptoms and patient- and parent-reported outcomes.

Radiographic Evaluation

CT scans were used to assess fusion at the spinal level of interest 12 months postsurgery. To analyze sagittal alignment, a full-spine upright radiographic series was obtained at each clinical visit. MRI scans were collected only as needed for new symptoms or if there were concerns of pain or neurological decline after surgery.

Clinical Evaluation

We retrospectively reviewed medical and operative records, radiographs, CT scans, and MRI sequences. Clinical assessments were made preoperatively, 2 weeks postsurgery, 3 months postsurgery, 6 months postsurgery, and annually. At each clinical visit we evaluated the following: presence or absence of subjective pain and sensory changes; gait ability; subjective bowel and bladder symptoms (including leakage between catheterizations, febrile UTIs, or social incontinence); and patient- and parent-reported outcomes. We used the PedsQL for our patient- and parent-reported outcomes. The PedsQL scale has been established as a generic quality-of-life assessment tool for pediatric populations.[27] Both parents and patients filled out PedsQL questionnaires for those 8–18 years of age. For patients < 8 years of age, only the parents filled out the PedsQL. The minimum clinically important difference (MCID) for PedsQL has been calculated to be 4.4 for patients and 4.5 for parents.[27] No patient-reported outcomes were used for patients > 18 years of age at the time of surgery, because PedsQL is only validated for children. Only patients with at least 12 months of follow-up were included in clinical and patient- and parent-reported outcomes analysis for this study. The need for reoperation due to retethering and surgical complications was documented.

Urodynamics

Urodynamics were documented preoperatively and were to be repeated at 3 months postoperatively to assess short-term surgical efficacy. Additional postoperative urodynamic testing was also performed whenever clinically indicated, such as by worsening symptoms of new leakage between catheterizations, febrile UTIs, and loss of social continence. Reasons that not all patients underwent urodynamic evaluation were multifactorial and included urgency of SCS and patient refusal. Preoperative and postoperative multichannel urodynamic testing was performed by a dedicated urodynamics team at our institution. These tests were reviewed by two independent pediatric urologists with experience in the care of patients with neuropathic bladder (K.M.S., R.M.). Urodynamics were classified as normal, safe, intermediate, or hostile bladder according to the National Spina Bifida Patient Registry Renal Protocol Group criteria.[28] Discrepancies in categorization, noted in 45.3% of cases, were resolved by re-reviewing and reaching consensus.

Statistical Analysis

Descriptive statistics including mean, standard deviation, and standard error were computed for all measurements. For pre- and postoperative presence or absence of clinical symptoms, p values were calculated using the 2-tailed McNemar test for matched data. The MCID was used to assess significance of the change in individual PedsQL from preoperative score to the score at the most recent follow-up. Both pre- and postoperative PedsQL and pre- and postoperative urodynamics were compared using the Wilcoxon signed-rank test for matched data. Statistical significance was set a priori at a p value less than 0.05 (Stata; StataCorp).

Results

We performed SCS in 41 patients with secondary TCS at our institution. As a point of interest, during the study period the senior author performed 13 (24%) standard untethering surgeries for secondary TCS, compared to the 41 (76%) SCS procedures. The mean operative time for the procedure was 3 hours and 52 minutes (range 2 hours and 18 minutes to 7 hours and 3 minutes). The mean estimated blood loss (EBL) was 662.8 ml (range 100–1700 ml). No patients have required reoperation due to retethering or complications (mean follow-up 22.6 months, range 8–45 months). Table 1 summarizes patient demographics, clinical data, and operative data. Table 2 summarizes all outcome results.

Complications

Three intraoperative complications occurred: 2 of acute blood loss and 1 of small unintended durotomy. The mean hospital stay duration was 5.1 days (range 2–25 days). No complications of new neurological deficit, CSF leakage, wound infection, or death were seen in our series. During surgery, 33 patients received an average transfusion of 1.6 units of packed red blood cells for blood loss leading to hemodynamic instability.

Patients, parents, and caretakers are counseled prior to

TABLE 1. Demographic, clinical, and operative data for 41 patients with secondary TCS

Demographics			Clinical & Operative Data										
Case No.	Age at Op (yrs)	Sex	Form of Spinal Dysraphism	Presentation	No. of Prior Untetherings	Surgical Procedure	EBL (ml)	Op Time (H:M)	Hospital Stay (days)	Complications*	Concurrent Scoliosis (yes/no)	Other Spinal Instrumentation	FU (mos)
1†	44	F	MMC	Back/leg pain, leg weakness, gait inability, B/B dysfunction	5	T12 resection, T10–L2 fusion	1700	5:30	12	None	No	Prior L3–S1 spinal fusion	25
2†	20	F	LMMC	Back/leg pain, leg weakness, gait inability, B/B dysfunction	2	T12 resection, T10–L2 fusion	1000	4:40	5	None	Yes	None	39
3†	21	M	MMC	Back/leg pain, gait inability, leg weakness, & bladder/bowel dysfunction	1	T10 resection, T8–12 fusion	800	7:03	4	None	Yes	None	26
4†	19	M	MMC	Leg weakness, B/B dysfunction	2	L1 resection, T11–L3 fusion	700	4:39	4	None	Yes	None	33
5†‡	17	F	MMC	Back/leg pain	1	T12 resection, T10–L2 fusion	850	3:47	4	None	Yes	None	25
6†	17	F	MMC	Back/leg pain, leg weakness, B/B dysfunction	4	L1 resection, T11–L3 fusion	600	5:30	6	None	Yes	None	45
7†	20	M	LMMC	Back/leg pain, leg weakness, B/B dysfunction	1	T12 resection, T10–L2 fusion	500	3:40	8	None	No	None	19
8†	12	F	Transitional spinal lipoma	Back/leg pain, leg weakness, B/B dysfunction	1	T10 resection, T8–12 fusion	250	3:42	5	None	No	None	14
9†‡	12	M	LMMC	Back/leg pain, leg weakness, B/B dysfunction	4	T12 resection, T10–L2 fusion	500	3:50	5	None	No	None	38
10†‡§	18	F	MMC	Back/leg pain, gait inability, leg weakness, B/B dysfunction	2	T12 resection, T10–L2 fusion	750	3:15	6	Acute blood loss anemia	Yes	None	18
11†‡	16	M	MMC	Gait inability, leg weakness, B/B dysfunction	2	T12 resection, T10–L2 fusion	250	4:00	5	Acute blood loss anemia	Yes	None	33
12†‡§	13	M	MMC	B/B dysfunction	4	T12 resection, T10–L2 fusion	450	4:42	4	None	Yes	T2–10 spinal fusion performed at 28 mos FU	30
13†‡§	12	M	LMMC	Back/leg pain, B/B dysfunction	2	T12 resection, T10–L2 fusion	400	3:04	2	None	No	None	13
14†	39	F	LMMC	Back/leg pain, leg weakness, B/B dysfunction	1	T12 resection, T10–L2 fusion	950	3:59	2	None	No	None	22
15†§	21	F	MMC	Back/leg pain, leg weakness, B/B dysfunction	1	L1 resection, T10–L3 fusion	1500	6:56	7	None	Yes	None	38
16†‡§	11	M	Transitional spinal lipoma	Back/leg pain, leg weakness, B/B dysfunction	2	L1 resection, T11–L3 fusion	400	2:18	3	None	Yes	None	35
17†‡§	10	F	MMC	Back/leg pain, leg weakness, B/B dysfunction	2	L1 resection, T11–L3 fusion	500	5:32	3	Small unintended durotomy at sleeve of rt L1 nerve root	Yes	None	24
18†‡§	10	F	MMC	Back/leg pain, gait inability, leg weakness, B/B dysfunction	1	T12 resection, T10–L2 fusion	300	3:15	4	None	Yes	None	24

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TABLE 1. Demographic, clinical, and operative data for 41 patients with secondary TCS

Demographics			Clinical & Operative Data									
Case No.	Age at Op (yrs)	Sex	Form of Spinal Dysraphism	Presentation	No. of Prior Untetherings	Surgical Procedure	EBL (ml)	Op Time (H:M)	Hospital Stay (days)	Complications*	Concurrent Scoliosis (yes/no)	Other Spinal Instrumentation	FU (mos)
19†‡	13	M	LMMC	Back/leg pain, gait inability, leg weakness, B/B dysfunction	2	T12 resection, T10–L2 fusion	500	3:21	4	None	Yes	None	20
20†‡	7	M	MMC	Back/leg pain, gait inability, B/B dysfunction	1	T10 resection, T8–12 fusion	100	3:40	5	None	No	None	36
21†‡§	17	M	LMMC	Leg weakness, B/B dysfunction	1	T12 resection, T10–L2 fusion	800	4:15	3	None	No	None	21
22†‡	15	M	MMC	Leg weakness, B/B dysfunction	0	T11 resection, T9–L1 fusion	600	4:00	5	None	Yes	None	19
23†§	6	M	LMMC	Back/leg pain, leg weakness, B/B dysfunction	1	T11 resection, T9–L1 fusion	450	3:44	4	None	Yes	None	28
24†‡§	5	F	Transitional spinal lipoma	Back/leg pain, leg weakness, B/B dysfunction	3	L1 resection, T11–L3 fusion	250	3:00	7	None	Yes	None	41
25†‡§	12	F	LMMC	Gait inability, leg weakness, B/B dysfunction	2	T12 resection, T10–L2 fusion	450	3:00	2	None	Yes	None	13
26†‡§	11	M	MMC	Back/leg pain, B/B dysfunction	0	T12 resection, T10–L2 fusion	700	3:21	3	None	No	None	13
27†‡§	14	M	MMC	Back/leg pain, leg weakness, B/B dysfunction	2	L1 resection, T11–L3 fusion	500	5:00	4	None	Yes	None	44
28†‡§	17	F	MMC	Back/leg pain, gait inability, leg weakness, B/B dysfunction	6	T12 resection, T10–L2 fusion	475	4:30	5	None	Yes	None	37
29†‡§	12	F	MMC	Gait inability, leg weakness, B/B dysfunction	1	T10 resection, T8–12 fusion	250	4:15	8	None	No	None	30
30†‡§	9	M	MMC	B/B dysfunction	0	L1 resection, T11–L3 fusion	550	3:33	4	None	Yes	None	17
31	55	M	MMC	Back/leg pain, gait inability, leg weakness, B/B dysfunction	1	T12 resection, T10–L2 fusion	1200	4:30	6	None	Yes	None	11
32§	20	F	LMMC	Back/leg pain, gait inability, leg weakness, B/B dysfunction	1	T12 resection, T10–L2 fusion	950	3:30	4	None	No	None	11
33	19	F	LMMC	Back/leg pain, leg weakness, B/B dysfunction	2	T12 resection, T10–L2 fusion	900	3:52	7	None	No	None	10
34§	12	F	Functional tethered cord	Back/leg pain, gait inability, leg weakness, B/B dysfunction	1	T12 resection, T10–L2 fusion	750	2:32	4	None	No	None	10
35	22	M	LMMC	Back/leg pain, gait inability, leg weakness, B/B dysfunction	1	T11 resection, T9–L1 fusion	700	3:22	25	None	No	None	11
36	12	F	Transitional spinal lipoma	Back/leg pain, leg weakness	3	L1 resection, T11–L3 fusion	700	2:36	2	None	No	None	8
37§	8	F	MMC	Back/leg pain, B/B dysfunction	1	L1 resection, T11–L3 fusion	850	2:28	5	None	No	None	8
38§	7	F	MMC	Back/leg pain, B/B dysfunction	1	L1 resection, T11–L3 fusion	300	2:34	4	None	Yes	None	9

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TABLE 1. Demographic, clinical, and operative data for 41 patients with secondary TCS

Demographics				Clinical & Operative Data									
Case No.	Age at Op (yrs)	Sex	Form of Spinal Dysraphism	Presentation	No. of Prior Untetherings	Surgical Procedure	EBL (ml)	Op Time (H:M)	Hospital Stay (days)	Complications*	Concurrent Scoliosis (yes/no)	Other Spinal Instrumentation	FU (mos)
39§	9	F	MMC	B/B dysfunction	1	T12 resection, T10–L2 fusion	1000	3:03	3	None	No	None	11
40§	10	F	Transitional spinal lipoma	Back/leg pain, B/B dysfunction	4	T12 resection, T10–L2 fusion	700	2:32	3	None	No	None	9
41	8	F	MMC	Back/leg pain, B/B dysfunction	1	L1 resection, T11–L3 fusion	1100	3:11	4	None	No	None	9

B/B = bowel/bladder; FU = follow-up; H:M = hours:minutes; LMMC = lipomyelomeningocele; MMC = myelomeningocele.
* Complications included new neurological deficit, CSF leak, infection, reoperation, or death.
† Included in clinical analysis (minimum of 12 months of follow-up).
‡ Included in PedsQL analysis.
§ Included in urodynamics analysis.

surgery that a blood transfusion will need to be undertaken either during or immediately after surgery. During the preincision timeout, the surgical and anesthesiology teams discuss the need for blood conservation and blood transfusion for potentially life-threatening blood loss. After the timeout, hourly EBL checks are performed between the surgical and anesthesiology teams to ensure that everyone in the operating room is on the same page.

Imaging Evaluation

A CT scan was performed at 12 months postsurgery. Solid arthrodesis was observed in all patients who reached at least 12 months of follow-up. The mean preoperative sagittal alignment was determined to be 6.3° of kyphosis (range 2°–16°). For patients with at least 12 months of follow-up, the mean sagittal alignment was 4.0° of kyphosis (range –7° to 13°) at latest follow-up. No signs of loss of alignment were evident at the most recent follow-up. An image obtained in a representative patient is seen in Fig. 1.

Postoperative MRI was performed for 4 patients. No new defects or pathology was observed in any patient; however, spinal cord detensioning is difficult to determine via MRI.

Clinical Evaluation

For the clinical symptoms and patient- and parent-reported outcomes aspects of this study, 30 patients with a minimum of 12 months of follow-up were included for analysis and 11 patients were excluded because they had yet to reach a minimum 12-month follow-up period at the time of analysis. These included 16 male and 14 female patients with an average age of 15.7 years (range 5–44 years) at the time of surgery. All of these patients were diagnosed with secondary TCS caused by myelomeningocele (18 patients), lipomyelomeningocele (9 patients), or transitional spinal lipoma (3 patients). At the time of study analysis, the mean postoperative follow-up period was 27.3 months (range 13–45 months) for this patient subset.

Of the 30 patients included for analysis, significant subjective clinical improvement was seen in the preoperative symptoms of pain, weakness, and bladder/bowel dysfunction at the most recent follow-up (see Table 2). No patients with the absence of a given symptom preoperatively had developed that symptom postoperatively at the most recent follow-up.

Of patients with at least 12 months of follow-up, 19 had both pre- and postoperative PedsQL scores recorded. For these patients, the mean PedsQL score at the most recent follow-up was 64.8 (range 25–99) reported by patients and 64.3 (range 28–98) reported by parents. At the most recent follow-up, the median patient-reported score increase was +5 ($p = 0.04$). The median parent-reported scores increased by +5, but this was not statistically significant ($p = 0.08$). On an individual level, for patient-reported PedsQL, 11 patients (57.9%) reported an improvement greater than the MCID, 5 patients (26.3%) had stable scores with no significant change, and 3 patients (15.8%) worsened with a decrease in score larger than the MCID. For parent-reported PedsQL, 10 patients (52.6%) showed significant improvement, 6 patients (31.6%) had stable scores, and 3 patients (15.8%) worsened.

TABLE 2. Summary of outcomes and results in 41 patients with secondary TCS

Outcome	Inclusion Criteria	No. of Pts Included	Summary of Results
Reop due to retethering/ complication after SCS	All pts	41	0.0% pts required reop due to postop retethering or complications at mean 22.6 mos FU
Successful spinal fusion	≥12 mos of FU	30	100.0% had solid arthrodesis w/ proper alignment observed at 12 mos
Subjective clinical symptoms	≥12 mos of FU	30*	91.3% w/ preop pain resolved (n = 23, p < 0.01); 66.7% w/ preop weakness resolved (n = 24, p < 0.01); 51.7% w/ preop B/B symptoms resolved (n = 29, p < 0.01); 36.4% w/o preop gait ability gained gait ability—not statistically significant (n = 11, p = 0.13)
PedsQL	≥12 mos of FU w/ pre- & postop scores available	19	Patient-reported PedsQL—statistically significant median score improvement of +5 (n = 19, p = 0.04): 57.9% improved; 26.3% remained the same; 15.8% worsened Parent-reported PedsQL—statistically not significant median score improvement of +5 (n = 19, p = 0.08): 52.6% improved; 31.6% remained the same; 15.8% worsened
Urodynamics results	Pts w/ pre- & postop urodynamic testing	17	Median improvement in 1 classification category (n = 17, p = 0.01): 58.8% improved; 35.3% remained the same; 5.9% worsened
Subsequent urological surgery	Pts w/ preop urinary complaints	39	33.3%: botulinum toxin A detrusor injections in 8, bladder augmentation in 4, replacement of artificial urinary sphincter in 1

Pts = patients.
* Not all 30 patients presented with all the symptoms listed.

Formal Urodynamics and Urology Evaluation

All patients with urodynamic testing results were included for analysis regardless of follow-up length. A review was completed of 34 preoperative urodynamic evaluations performed at a median of 2 months before surgery. Preoperative urodynamic classifications included 55.9% hostile, 17.0% intermediate, 20.6% safe, and 6.9% normal bladder. For 19 patients, postoperative urodynamics were performed at a median of 3.5 months. Postoperative urodynamic classifications included 26.3% hostile, 31.5% intermediate, 36.8% safe, and 5.3% normal bladder.

A total of 17 patients had pre- and postoperative urodynamics available for comparison. The median change was an improvement of 1 classification category ($p = 0.01$). Urodynamics improved in 58.8%, remained the same in 35.3%, and worsened in 5.9%. Urodynamics and urology evaluation documented stable to improved bladder function in 16/17 patients in whom both pre- and postoperative urodynamics were performed. Among 39 patients with preoperative urinary complaints, 13 (33%) underwent urological surgery after SCS to optimize bladder function, including bladder botulinum toxin A detrusor injections (8 patients), bladder augmentation (4 patients), and replacement of artificial urinary sphincter (1 patient).

Discussion

As the current gold standard treatment for TCS, direct untethering has favorable outcomes but considerable drawbacks.[9,10,29] SCS represents an emerging surgical technique that has the ability to circumvent many prominent risks by relieving tension on the spinal cord without requiring entry into the dura.

Many authors have described and discussed SCS for TCS in case reports or small case series.[16,18,20,30] We previously reported our initial series of 7 children and emerging adults who underwent SCS for TCS.[21] We showed that pedicle subtraction osteotomy is an effective and safe technique for SCS, although it added a degree of unwanted lordosis at the thoracolumbar junction.

Since our initial experience, our surgical technique has evolved to incorporate vertebral column resection to ensure a more neutral closure of the osteotomy defect. Our technique for SCS is adopted from previously published techniques.[6,16,17,20,31] Similar to our previous study, the most common level of spinal column osteotomy was at T12 because minimal curvature of the spinal column in this region allows for easier fusion of the vertebra above and below the defect. Additionally, the thoracolumbar junction is selected for fusion in almost all cases because of the stiffness of this transitional zone where fusion would not be expected to add to the natural limited range of motion of the thoracolumbar junction.

As discussed in our previous series, we chose an osteotomy size of 1.5 cm for all our patients regardless of their height and age to maximize safety and efficacy.[21] However, this exact measurement may be an area of optimization in the future, especially when treating young children.

Most of the time, the apex for a patient's neuromuscular scoliosis in the setting of myelomeningocele is concordant with the levels of bony defect at the lumbosacral spine. By performing a relatively short-segment fusion at the thoracolumbar junction, we were able to avoid stopping our fusion at the apex of a patient's spinal deformity. Stopping a fusion at the apex of a scoliosis worsens that natural history and accelerates progression. In theory, if the apex of neuromuscular scoliosis involved the thoracolumbar junction, then we would plan for a more extensive fusion to address both SCS and scoliosis reduction.

In our experience, the risks presented by SCS appear to be no more than for traditional untethering for secondary TCS. Following at least 1 prior untethering operation, traditional TCR carries an estimated complication rate of 4%–17% for CSF leakage or new neurological deficits.[12,14,32] Our study showed no complications of CSF leakage, new

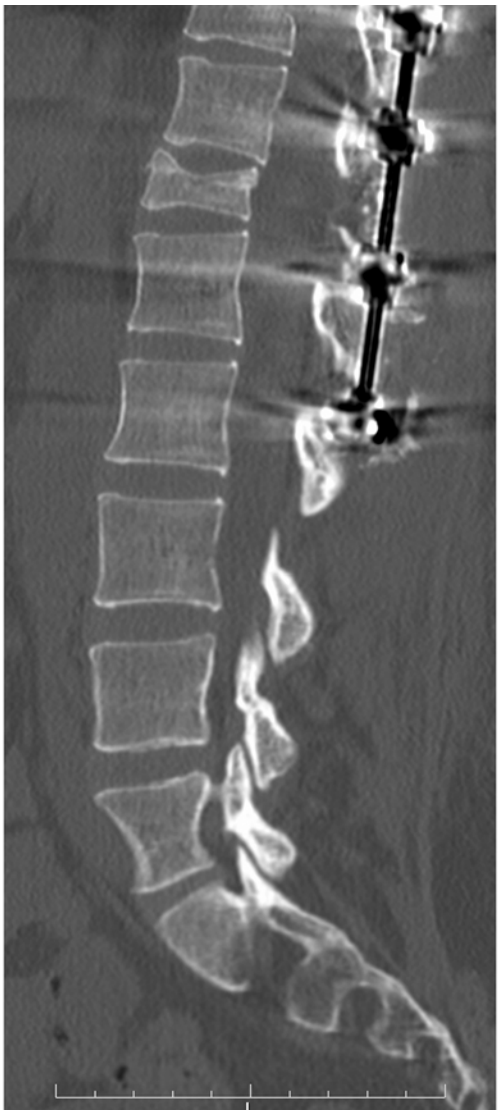

FIG. 1. Sagittal CT scan of the lumbar spine of an emerging adult patient documenting SCS at T12. T12 can be compared to the heights of its adjacent vertebral bodies to gauge the degree of shortening. In this case, it appears that approximately 50% shortening (approximately 1.5–2 cm) of the vertebral body was achieved. Note that the T11–12 and T12–L1 intervertebral discs and superior and inferior endplates of T12 were preserved.

neurological defects, infection, reoperation, or death in our 41 patients. Worsening of preoperative neurological symptoms was not considered a new neurological defect. Importantly, the most common complication of direct untethering—retethering—has not been observed in any patients at a median follow-up of 21 months. The short-term complication rates seen in this series improve upon the complication rates reported in other SCS procedures used to treat TCS.[16,18,30]

Most of our patients in this series with at least 12 months of follow-up (mean 27.3 months) demonstrated improvement in preoperative symptoms (see Table 2). These improvements were validated by the results obtained through standardized patient- and parent-reported outcomes and formal urodynamics. However, 1 in 3 patients with preoperative bladder abnormalities went on to have additional urological procedures.

Results from the PedsQL showed a significant increase in the median patient-reported PedsQL score, indicating a valuable improvement in quality of life. However, the median parent-reported PedsQL results did not result in a statistically significant improvement. From a clinical perspective, this difference may be due to parents being more critical of quality-of-life improvements in their child's health. This may partially explain the discrepancy between the significant improvements seen in the patient-reported outcomes versus the parent-reported outcomes.

Of our patients with both pre- and postoperative formal urodynamic data, we saw objective improvements in 58.8% of patients, with 16/17 patients showing stable to improved urodynamics. Additionally, we saw symptomatic improvement in 51.7% of our patients who reported preoperative bladder/bowel dysfunction. These results compare favorably with previously reported symptomatic urological findings and formal urodynamic results for patients treated with a traditional untethering procedure. Although there is some variation in exact formal urodynamic testing procedures and grading systems, previous studies in which formal urodynamics were used to evaluate urological outcomes of secondary TCS indicate improvement in roughly 50% of patients.[33–35] Based on these findings, SCS shows promising results as an alternative to direct untethering for treating urological symptoms and urodynamic dysfunction.

Even with improvements seen in most patients, we had individuals who showed no improvements and a few patients with worsening symptoms after SCS. The cause for lack of improvement or worsening of preoperative symptoms is probably multifactorial. A possible explanation is that with prolonged excessive tension placed on the spinal cord causing alterations to blood flow and metabolism, irreversible damage may be done to the neurons that will not improve even upon relief of spinal cord tension. Clinical improvement might not occur in these cases. Because most patients in this series had at least 1 prior untethering procedure, they may have been at increased risk for irreversible damage prior to SCS. Another possibility for lack of clinical improvement may be lack of tension release due to inadequate SCS.

Last, of primary concern with SCS in this population is the spinal fusion of skeletally immature children. In this study, treatment of these children was deemed appropriate on an individual patient basis due to the severity of symptoms, serious risks associated with additional TCR, and concerns for permanent damage if treatment was delayed further. However, there remains potential for continued growth of the nonfused portions of the spinal column, thus leading to reoccurrence of pathological tension on the spinal cord. Additionally, spinal fusion in children is likely to result in growth retardation of 1 mm per year of remaining growth per spinal level fused, which was typically 5 spinal levels in our series.[36] Unopposed growth of the anterior and middle portions of the fused spinal segments while the posterior column is secured with fusion hardware has the potential to lead to a crankshaft deformity.[36] For these reasons, it could be best to delay SCS until skeletal matu-

rity in patients who are at an increased risk for these occurrences. Whereas these are all significant concerns, we have yet to find significant complications due to continued skeletal growth at our most recent follow-up in this series. Furthermore, these concerns regarding skeletal growth must be balanced with the increased risk of permanent neurological damage that can occur due to prolonged tension on the spinal cord if treatment is delayed.

Limitations

Although the urodynamic grading system used in this study has been widely advocated, limited published studies have used it longitudinally to assess TCS treated with either traditional untethering or SCS. Furthermore, although urodynamics were the urological outcome in the study, they were typically considered in the clinical context of urinary symptoms along with neurosurgical complaints and signs. Because postoperative urodynamic evaluations were not performed in all patients, only those with available tracings were amenable for review.

The median length of time of symptoms prior to surgery was not quantified for this study. Future studies are needed to investigate the length of symptoms prior to surgery and its association with clinical outcomes following TCR or SCS.

Despite growing evidence of safety and efficacy, many questions remain unanswered regarding SCS. Explanations are still needed for inconsistencies in outcomes, and the long-term durability of our outcomes still needs to be assessed. Exact indications for patient selection and treatment need to be established, including timing of surgery. Timing of surgery is critically important, especially when treating children who have yet to reach skeletal maturity. Moreover, recurrent TCS plays out over many years; therefore, long-term follow-up is necessary.

To address these concerns, further studies are needed. A prospective comparative study of SCS and traditional untethering for secondary TCS in the pediatric age group would provide invaluable insights. Additionally, the use of a patient- and parent-reported outcomes assessment specific to spina bifida would improve future assessments of outcomes of secondary tethered cord treatments.

Although our series was primarily focused on the pediatric age group, SCS was originally described for emerging adults.[16,18,20] Emerging adults with recurrent TCS were considered ideal candidates for SCS because they had reached skeletal maturity, and once the spinal column is shortened, the effects are likely to be permanent. Hence, we hold that SCS may apply to both children and the emerging adult population.

Conclusions

Direct untethering of the spinal cord is the current gold standard surgical treatment for secondary TCS due to myelomeningocele, spinal lipoma, and lipomyelomeningocele. However, severe shortcomings of this treatment exist for secondary TCS, including the frequent complication of symptomatic retethering of the spinal cord. Using formal urodynamics and patient- and parent-reported outcomes, our series of pediatric and emerging adult patients demonstrates that SCS is a safe and efficacious alternative treatment for secondary TCS, especially for patients with prior untethering procedures.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Jea, Szymanski. Acquisition of data: all authors. Analysis and interpretation of data: Jea, McVeigh, Anokwute, Belal, Zieles, Szymanski, Misseri. Drafting the article: McVeigh, Anokwute, Szymanski, Misseri. Critically revising the article: Jea, McVeigh, Anokwute, Belal, Raman, Szymanski, Misseri. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Jea. Statistical analysis: McVeigh, Zieles, Szymanski. Administrative/technical/material support: Raman, Zieles. Study supervision: Jea.

Supplemental Information

Previous Presentations

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