

Spinal cord tethering following myelomeningocele repair

Clinical article

VIVEK A. MEHTA, B.S.,¹ CHETAN BETTEGOWDA, M.D., PH.D.,¹ SEBASTIAN A. AHMADI, M.D.,² PETRA BERENBERG, M.D.,² ULRICH-WILHELM THOMALE, M.D.,² ERNST-JOHANNES HABERL, M.D.,² GEORGE I. JALLO, M.D.,¹ AND EDWARD S. AHN, M.D.¹

¹Department of Neurosurgery, Division of Pediatric Neurosurgery, The Johns Hopkins Hospital, Baltimore, Maryland; and ²Department of Pediatric Neurosurgery, Charité—Universitätsmedizin Berlin, Germany

Object. Symptom response to spinal cord untethering, and the impact of duraplasty and scoliosis on retethering, are poorly understood in tethering after myelomeningocele (MMC) repair. In this retrospective study, the authors examined the outcomes of children who developed first-time spinal cord tethering following MMC repair. The response of symptoms to untethering and the role of duraplasty and scoliosis in retethering are explored.

Methods. The authors performed a review of 54 children with first-time symptomatic spinal cord tethering following MMC repair to determine the impact of untethering on symptoms, the impact of dural repair type on retethering, and the role of scoliosis on the prevalence and time to retethering.

Results. The average patient age was 10.3 ± 4.9 years, and 44% were males. The most common presenting symptoms of tethered cord syndrome were urinary (87%), motor (80%), gait (78%), and sensory (61%) dysfunction. The average postoperative time to symptom improvement was 2.02 months for sensory symptoms, 3.21 months for pain, 3.50 months for urinary symptoms, and 4.48 months for motor symptoms, with sensory improvement occurring significantly earlier than motor improvement (p = 0.02). At last follow-up (an average of 47 months), motor symptoms were improved in 26%, maintained in 62%, and worsened in 11%; for sensory symptoms, these rates were 26%, 71%, and 3%, respectively; for pain, 28%, 65%, and 7%, respectively; and for urinary symptoms, 17%, 76%, and 7%, respectively. There was no difference in symptom response with type of dural repair (primary closure vs duraplasty). Symptomatic retethering occurred in 17 (31%) of 54 patients, but duration of symptoms, age at surgery, and type of dural repair were not associated with retethering. Scoliosis was not associated with an increased prevalence of retethering, but was associated with significantly earlier retethering (32.5 vs 61.1 months; p = 0.042) in patients who underwent additional untethering operations.

Conclusions. Symptomatic retethering is a common event after MMC repair. In the authors' experience, sensory improvements occur sooner than motor improvements following initial untethering. Symptom response rates were not altered by type of dural closure. Scoliosis was associated with significantly earlier retethering and should be kept in mind when caring for individuals who have had previous MMC repair. (DOI: 10.3171/2010.8.PEDS09491)

KEY WORDS • tethered cord syndrome • scoliosis • myelomeningocele

FOLLOWING MMC repair, nearly all children will demonstrate radiographic evidence of spinal cord tethering,¹⁵ although only 10%–30% of them will experience symptoms that prompt surgical release.^{3,6,41,45} The role of surgical intervention in these children has been well established,³⁵ but the response patterns of preoperative symptoms to surgical release are not clearly defined, and are highly variable.^{7,9,13,15,26} This variability may be due to reporting responses to untethering from a variety of tether-

ing origins, variable timing of TCR following symptom onset, and nonstandardized definitions of symptom response.¹¹

Following MMC repair and subsequent index untethering, approximately 15% of children will experience symptomatic retethering that requires a second untethering procedure.^{4,13,24} Different techniques have been attempted to minimize retethering, including biological and synthetic dural grafts¹⁹ and a variety of suture techniques,^{38,44} but their efficacies have not been clearly defined, and factors associated with retethering remain unknown.

This article contains some figures that are displayed in color online but in black and white in the print edition.

Abbreviations used in this paper: LOS = length of stay; MMC = myelomeningocele; PVCSCO = posterior vertebral column subtraction osteotomy; TCR = tethered cord release; TCS = tethered cord syndrome.

Spinal cord tethering following myelomeningocele repair

Due to the nonspecific radiological findings of a tethered cord, the diagnosis of TCS is made clinically when symptoms suggestive of tethering occur in the setting of positive radiological findings.^{15,18,47} The term “retethering” is then applied based on symptom recurrence or progression, as every child will experience some degree of asymptomatic retethering.¹⁸ Therefore, a better understanding of the expected symptom response patterns following index untethering may help predict subsequent retethering. In this retrospective study, we examine the outcomes of 54 children who developed first-time spinal cord tethering following MMC repair. The response of symptoms to untethering and the role of duraplasty and scoliosis in retethering are explored.

Methods

Data Collection

Approval for this study was obtained from The Johns Hopkins Hospital Institutional Review Board. Fifty-four patients who experienced symptomatic tethering following an initial repair of MMC were identified from a total of 182 patients who underwent first-time untethering due to any TCS cause from 1997 to 2008. Tethered cord release was performed on symptomatic patients regardless of the presence or absence of scoliosis. Any asymptomatic patients who underwent untethering were excluded from this study. A retrospective review of medical records was performed that included all hospital records, pre- and postoperative clinic notes, and pre- and postoperative imaging studies. Patient demographics, symptoms, and neurological deficits that were present immediately prior to surgery, immediately following surgery, at regular follow-up visits, and at last available follow-up were recorded. Perioperative complications were recorded from operative notes and discharge summaries.

Retethering was defined as a new onset or worsening of symptoms suggestive of terminal spinal cord dysfunction (for example, urinary dysfunction, sensory deficit, motor dysfunction, and pain), occurring with or without positive radiological findings of a low-lying conus medullaris. Signs and symptoms were evaluated at regular postoperative clinic visits and were recorded based on the patient's subjective reports of symptoms and the assessment of signs by the physician. Perioperative complications occurring in the same patient were recorded as independent events. Symptom change was categorized as improved, maintained, or worsened as compared with the immediate preoperative status. The modified McCormick grading scale for spinal cord tumors was retrospectively applied to compare pre- and postoperative gross motor and sensory function (Table 1).²⁹

Data Analysis

The Kaplan-Meier²¹ method was used to compare re-tethering-free survival based on the type of dural closure and scoliosis status. Parametric data were expressed as means \pm SDs and compared using the Student t-test. The Mann-Whitney U-test was used to compare nonparametric data and reported as medians. The Fisher exact test was used to compare percentages.

TABLE 1: Modified McCormick grading scale for neurological function

Grade	Description
I	neurologically intact, ambulates normally, may have minimal dysesthesia
II	mild motor or sensory deficit, maintains functional independence
III	moderate deficit, limitation of function, independent w/ external aid
IV	severe motor or sensory deficit, limit of function w/ dependent patient
V	paraplegia or quadriplegia, even in the presence of flickering movement

Results

Patient Population

Fifty-four pediatric patients underwent TCR for the first time following MMC repair. The mean patient age at surgery was 10.3 ± 4.9 years (median 10.8 years) with 24 male patients (44%). A primary dural repair was performed in 22 patients (41%), and duraplasty was performed in 32 patients (59%). All patients presented with first-time symptomatic tethering following initial repair of an MMC. The level of untethering/prior MMC was upper lumbar in 4 patients, midlumbar in 5, lower lumbar in 20, lumbosacral in 20, and sacral in 5 patients. The average duration of symptoms before surgical intervention was 7.07 months, and the average modified McCormick grade at the time of surgery was 2.79, which improved to 2.66 after surgery. The most common presenting symptoms included urinary dysfunction (87%), motor dysfunction (80%), gait abnormalities (78%), sensory deficit (61%), and spasticity (52%) (Fig. 1). Scoliosis was noted in 50% of patients. At presentation, patients who received primary dural closure versus duraplasty were generally similar across 15 categories of presentation. A summary of all demographic and presenting symptoms is provided in Table 2.

Perioperative Outcomes

The average LOS was 9.80 ± 7.86 days (range 3–51 days). A surgical site infection occurred in 3 (5.6%) of 54 patients, wound dehiscence in 9 (16.7%), and CSF leak (transcutaneous or subcutaneous) requiring revision surgery in 6 (11.1%). Patients who developed a CSF leak requiring revision were also considered to have wound dehiscence, as each adverse outcome was independently recorded. The overall percentage of patients who experienced any complication was 18.5%. The use of a dural graft was not associated with any increased risk of perioperative complications, including CSF leak (Table 2).

Symptom Response

The mean follow-up period for all patients was 47.0 ± 37.7 months (median 49.5 months, range 0.3–163 months). The mean time to improvement in symptoms was as follows: sensory deficit improvement, 2.02 months; pain relief, 3.21 months; urinary dysfunction, 3.50 months; and motor deficit improvement, 4.48 months. Improvement in sensory

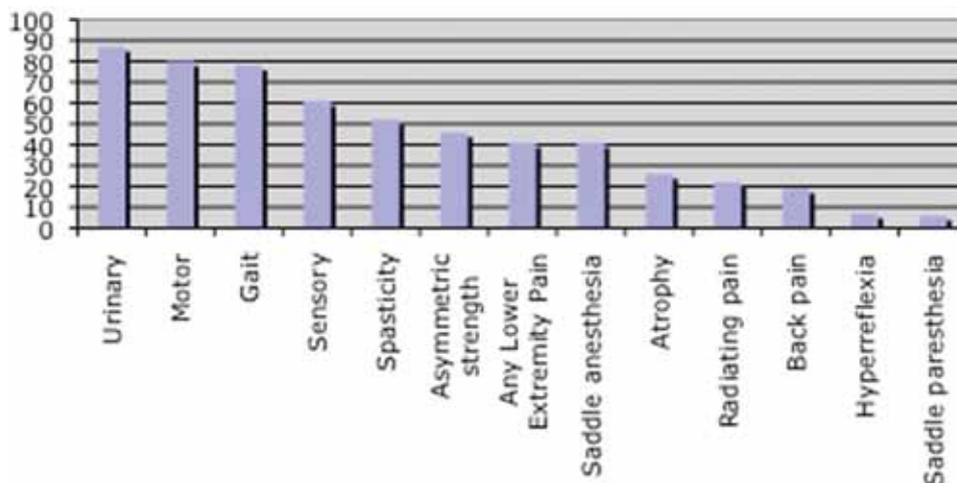


Fig. 1. Bar graph showing the relative incidence of symptom presentation.

deficits was found to occur significantly earlier than motor deficits (2.02 vs 4.48 months, respectively; $p = 0.02$) (Table 3). The time course to improvement demonstrated no other statistically significant pattern. Pain improvement was observed in 28% of patients, maintenance of preoperative state in 65%, and worsening in 7%. Preoperative sensory deficits were improved or maintained in 26% and 71%, respectively, with 3% reporting worsening. Improvement in motor function was reported in 26%, with maintenance or worsening in 62% and 11%, respectively. Stabilization of preoperative urinary symptoms was observed in 76%, with 17% experiencing improvement and 7% experiencing worsening. A complete report of symptom status at last follow-up is provided in Table 4. The duration of symptoms preoperatively and patient age at untethering were not associated with improvements in sensory deficit, weakness, motor deficit, or urinary complaints after surgery (Table 5). The level of tethering did not correlate with any functional outcomes postoperatively.

Retethering Outcomes

Symptomatic retethering occurred in 17 (31%) of 54 patients at a mean of 51 ± 33 months (range 10–133 months). Retethering was observed in 8 (36%) of 22 patients who underwent primary dural closure, and 9 (28%) of 32 who underwent closure with a dural graft, which was not statistically significant ($p = 0.537$). Mean time to retethering between primary dural closure (46 months) and dural graft (56 months) was also not statistically significant ($p = 0.580$). While it may be interpreted from Fig. 2 that duraplasty affords some early protection against retethering, that advantage was lost at final follow-up. When considering all patients, not just those who experienced retethering, there was no significant difference in retethering-free survival based on type of dural closure (Fig. 3). There was no significant difference in preoperative symptom duration in patients who experienced retethering (6.94 months) compared with those who did not undergo retethering (7.21 months; $p = 0.877$). The average patient age at the time of surgery was not significantly different in those who experienced retethering (9.26 years) compared with those who did not undergo retethering (10.66 years; $p = 0.345$)

(Table 5). Retethering was observed in 11 (41%) of 27 patients with scoliosis, and in 6 (22%) of 27 patients without scoliosis ($p = 0.149$). In patients with scoliosis, retethering requiring untethering occurred significantly earlier (32.5 months) as compared with patients without scoliosis (61.1 months; $p = 0.042$) (Fig. 4). However, when considering all patients, not just those who experienced retethering, there was no significant difference in retethering-free survival based on scoliosis status (Fig. 5). The level of tethering was not associated with retethering.

Discussion

Symptom Presentation

The decision to release a tethered cord is a clinical determination based on symptoms. Therefore, a better characterization of preoperative symptoms and their duration may lead to more timely intervention. A characteristic set of symptoms suggestive of tethering has been known to exist for many years, but a great variety of presenting symptoms exist, even in the subset of patients who experience index tethering following MMC repair. The 2 largest patient series concerning tethering after MMC repair illustrate the nonuniform nature of symptom presentation. In a review¹⁵ of 100 patients who developed a tethered cord following MMC repair, the most common presenting symptoms were motor dysfunction (55%), gait abnormality (54%), scoliosis (51%), pain (32%), orthopedic deformities (11%), and genitourinary dysfunction (6%). A later analysis of 114 children with TCS after MMC repair demonstrated that spasticity (47%), weakness (41%), scoliosis (40%), contractures (30%), urological dysfunction (26%), and pain (17%) were the most common presenting symptoms.⁷ The pattern of motor deficit, gait abnormality, weakness, and spasticity as dominant symptoms were also observed in this study, but in contrast to previous reports, urinary dysfunction was documented in more than 80% of patients. However, it is known that most children who present for nonurological signs/symptoms of tethered cord are found to have abnormal urodynamics when examined formally,³³ suggesting that the diversity of symptoms at presentation may instead represent variable preoperative evaluation and documen-

Spinal cord tethering following myelomeningocele repair

TABLE 2: Demographics, presenting symptoms, and operative/perioperative outcomes in 54 consecutive pediatric patients with tethered cord following initial MMC repair

Variable	Total Patients	Nonduraplasty	Duraplasty	p Value
demographics				
no. of patients	54	22	32	
mean age (yrs)	10.3	11.1	9.9	
males	24 (44%)	10 (45%)	14 (44%)	0.7
infants	3	2	1	0.16
preop presentation				
average modified McCormick grade	2.79	2.72	2.83	0.59
average symptom duration (mos)	7.07	5.9	8	0.17
bowel/bladder dysfunction	47 (87%)	18 (82%)	29 (91%)	0.38
lower-extremity motor dysfunction	43 (80%)	20 (91%)	23 (72%)	0.068
gait abnormalities	42 (78%)	19 (86%)	23 (72%)	0.19
lower-extremity spasticity	28 (52%)	8 (36%)	20 (63%)	0.62
any lower-extremity pain	22 (41%)	11 (50%)	11 (34%)	0.3
leg, perineal (saddle) anesthesia	22 (41%)	9 (41%)	13 (41%)	0.98
muscular atrophy	14 (26%)	6 (27%)	8 (25%)	0.86
leg pain (buttock, radiating pain)	12 (22%)	7 (32%)	5 (16%)	0.19
axial back pain	10 (19%)	6 (27%)	4 (13%)	0.18
lower-extremity hyperreflexia	4 (7%)	2 (9%)	2 (6%)	0.71
leg, perineal (saddle) paresthesia	3 (6%)	3 (14%)	0 (0%)	0.083
vertebral body defect	36 (67%)	14 (64%)	22 (69%)	0.88
scoliosis	27 (50%)	11 (50%)	16 (50%)	0.98
operative & perioperative outcomes				
surgical site infection	3	1	2	0.78
average LOS (days)	9.8	9.32	10.2	0.67
wound dehiscence	9	3	6	0.62
discharge to rehabilitation facility	6	4	2	0.22
average postop McCormick grade	2.66	2.5	2.77	0.29
revision op for CSF leak	6	2	4	0.69
deep venous thrombosis	1	0	1	0.33
average follow-up (mos)	47	44.72	48.56	

tation than a true difference in symptom frequency. This variability in preoperative urological evaluation methodology might explain the very high incidence of urological dysfunction in our study. The duration of symptoms prior to index TCR in our study (7.1 months) is consistent with that observed in other reports.²⁸ However, it is known that

the symptom duration prior to untethering increases with subsequent retetherings, which may suggest that a less aggressive approach is adopted once it becomes apparent that a patient is prone to tethering. Finally, as has been reported previously, pain was more commonly reported in older children, presumably because they are more able to articu-

TABLE 3: Average time to postoperative symptom improvement, and comparison of p values for average time to symptom improvement*

Symptom	Average Time to Improvement (mos)	Motor Deficit	Sensory Deficit	Weakness	Pain	Urinary Dysfunction
motor deficit	4.48	NA	0.02	0.10	0.40	0.52
sensory deficit	2.02	NA	NA	0.54	0.35	0.27
weakness	2.52	NA	NA	NA	0.62	0.49
pain	3.21	NA	NA	NA	NA	0.86
urinary dysfunction	3.50	NA	NA	NA	NA	NA

* NA = not applicable.

TABLE 4: Percentage of symptomatic improvement, maintenance, or worsening at last follow-up following untethering

Symptom	% Improved	% Maintained	% Worsened
nonduraplasty group			
motor deficit	29	62	10
sensory deficit	18	77	5
pain	36	55	9
urinary dysfunction	18	73	9
duraplasty group			
motor deficit	25	63	13
sensory deficit	30	68	3
pain	22	72	6
urinary dysfunction	16	78	6
all patients			
motor deficit	26	62	11
sensory deficit	26	71	3
pain	28	65	7
urinary dysfunction	17	76	7

late this symptom. It is difficult to account for this pattern when analyzing pain improvement following TCR.¹⁸

Perioperative Complications

In contrast to tethered cord symptom presentation, the perioperative complications are consistent across studies. In a review of a large national administrative database, an overall mortality rate of 0.0005% was reported with untethering, with an overall complication rate of 9.5%.²³ Hematoma and hemorrhage were the leading complications in this report, but CSF leak and surgical site infection are the ones most commonly reported elsewhere in the literature.^{16,37} Reports of CSF leak range from 1.6% to 17%^{7,13,15,23,28} and surgical site infections range from 1.1% to 8.4%,^{9,15,23} values that are consistent with observations in this report. An overall complication rate of less than 10% is consistently reported,^{13,23,28} but that does increase with sub-

TABLE 5: Impact of symptom duration and age at surgery on incidence of re-tethering and symptom improvement

Variable	Longest Symptom Duration (mos)	Age at Op (yrs)
re-tethering	6.94	9.26
no re-tethering	7.21	10.66
p value	0.87	0.34
improvement*		
sensory deficit	0.14	0.19
weakness	0.79	0.78
motor deficit	0.65	0.87
urinary dysfunction	0.64	0.77

* Symptom values represent p values of comparison between improvement and no improvement.

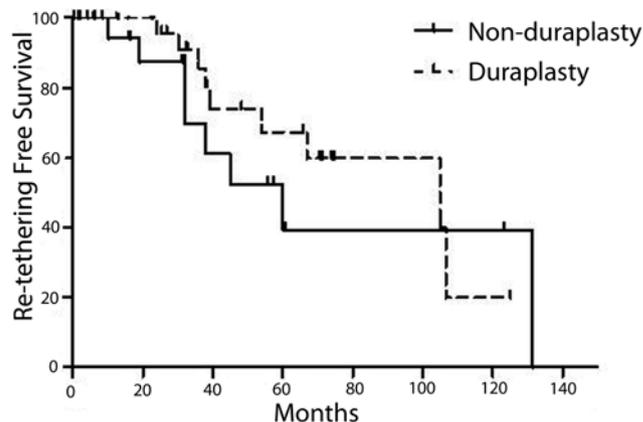


FIG. 2. Kaplan-Meier graph demonstrating the prevalence and time course to re-tethering in patients treated with duraplasty or primary dural closure (Non-duraplasty).

sequent untetherings.²⁸ The average LOS in this study (9.8 days) is also consistent with that reported elsewhere.^{23,28}

Symptom Response

While the role of surgical intervention might have previously been unclear, TCR in the correct clinical setting is now widely accepted. Persistent tethering results in symptomatic progression leading to an orthopedic or urological surgery in 90% of patients.³⁵ Additionally, the general goals of untethering and a pattern of response to untethering have been clearly shown in multiple studies. Surgical release of a tethered spinal cord is indicated in the proper clinical setting of progressive symptoms with positive radiographic evidence of tethering. The goals of untethering are to prevent further decline, prevent irreversible injury, and induce stabilization of preoperative symptoms.^{15,30,32}

Numerous studies have shown that following untethering, more than 80% of patients will experience stabilization of their preoperative symptoms, with significantly fewer patients experiencing mild improvement.^{13,15,26,28} The sole exception to this is the finding that pain consistently improves following cord release,²⁶ even after multiple untetherings.²⁸ Excellent improvements in pain following untethering have been documented in numerous studies, occurring in 80%–100% of patients.^{5,7,15,26,28}

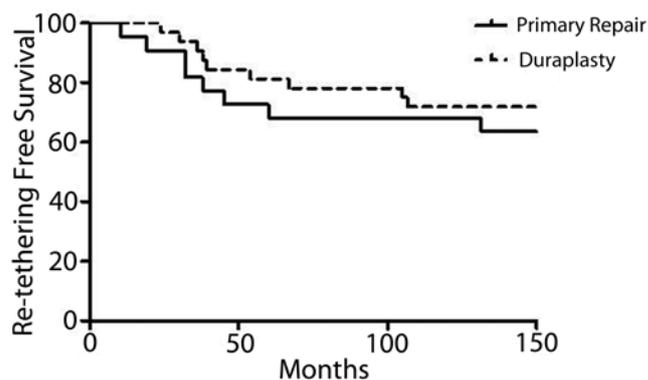


FIG. 3. Kaplan-Meier graph showing the incidence and time to re-tethering in all patients as a function of duraplasty or primary dural closure (Primary Repair).

Spinal cord tethering following myelomeningocele repair

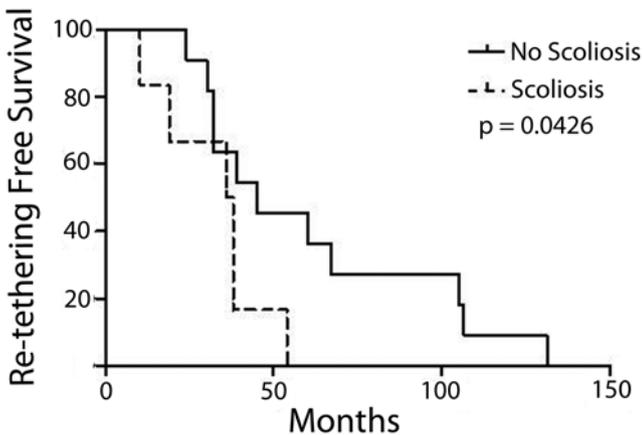


FIG. 4. Kaplan-Meier graph demonstrating the time to retethering in the presence or absence of scoliosis in patients who experienced retethering.

Improvements in pain following untethering often reach nearly 100% with prolonged follow-up.⁷ In this series, pain improved in one-quarter of patients, but stabilization was the most common response (65%) at last follow-up. Immediately postoperatively, pain improved in 47 (87%) of 54 patients, but the majority experienced regression to preoperative status by last follow-up. When retethering occurs, pain continues to respond well to surgery, but that response diminishes with each subsequent untethering. The same pattern of symptom stabilization/improvement is also observed in patients who undergo late untethering, suggesting that symptom duration or patient age at surgery does not play a significant role in symptom response.²⁸ Similarly, we observed that improvement in the other 4 major presenting symptoms occurred independent of either patient age at presentation or symptom duration.

While untethering consistently relieves pain, the impact on functional improvement is less reliable.²⁷ Improvements in urological signs and symptoms are the second most commonly reported to occur following untethering. Improvement rates as high as 60% have been reported,^{7,22} with an additional 36% achieving stabilization,⁷ prompting many to recommend urodynamic testing early and often.^{11,16,37,43} With repeat untethering, the response of urological symptoms still occurs in approximately 50%.²⁸ However, others have reported that the rates of improvement/stabilization of urological symptoms range from 16% to 67%, similar to the rates of other nonpain symp-

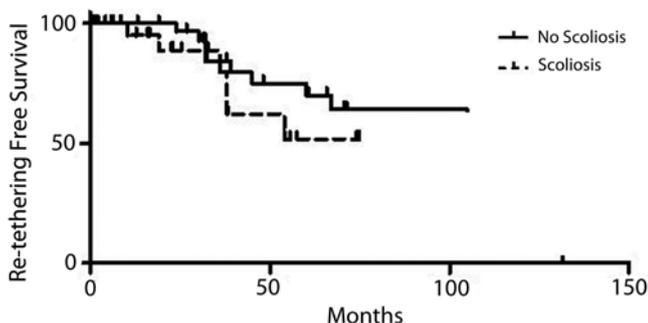


FIG. 5. Kaplan-Meier graph showing the time to retethering in the presence or absence of scoliosis in all patients.

toms.^{15,25,26,33} Again, the variability in reporting is likely due to variable definitions and measures of improvement, but may also represent a differential symptom response based on the cause of tethering.¹¹ The former explanation might explain our data that 75% of children experienced long-term stabilization and only 17% experienced improvement. Some have even argued that the reported urological improvement may not be reliable given the high rates of incontinence in young children and the limits of urodynamic testing.^{8,10}

Improvements in motor function appear to follow a similar pattern as urological dysfunction. Most studies report a response rate of at least 25%,^{7,15,28} with a range of 25%–80%.^{20,25,35} Interestingly, children who were not found to have preoperative motor deficits demonstrated motor improvements after surgery, leading some to believe the threshold for untethering should be lower.¹⁵ In this series, improvement in motor symptoms was observed in 26%, with 62% reporting maintenance and 11% worsening at last follow-up, consistent with the range reported in the literature. While most believe that pain, urinary dysfunction, and motor deficits respond best to untethering, few have reported some improvement in sensory and weakness following untethering.²⁸

The average time to symptom improvement is not well understood, but establishing a normal range for this variable may be important for evaluating when untethering has not been successful. In this series, we found that the average reported improvement occurred between 2 and 4.5 months. Interestingly, the improvement in sensory symptoms occurred significantly earlier than improvements in motor deficits, which may represent the increased time needed to regain strength in muscles that have atrophied secondary to neuronal damage from TCS. As has been shown in numerous prior studies, our data show that dramatic improvement is rare, and that stabilization is the most likely response of symptoms to untethering.^{12,46} While better results might be observed with other tethered cord causes, it is important to keep in mind that symptom presentation, response, and retethering behave uniquely in the patient after MMC repair.⁴²

Retethering

Following index untethering, retethering occurs in as many as 15% of patients after MMC repair,^{4,13,14,24,27} and in as many as 50% of patients when all TCS causes are considered.^{2,20,24,25} Various suture techniques,^{38,44} spinal cord/dural barrier materials,⁴⁷ and synthetic dural grafts¹⁹ have all been employed in an attempt to reduce retethering, with the latter being the most popular technique.¹ At some centers, patients are maintained in a flat position following surgery for at least 1–2 days. In severe cases, sectioning of the spinal cord at a functionless level has been proposed.⁵ However, none of these techniques has been reliably shown prospectively to prevent retethering.^{18,26,47}

In a recent report, Hsieh et al.¹⁷ reported on PVCSSO in 2 patients with repeated cord tethering. A technique previously used for spinal deformity, PVCSSO allows for a degree of untethering not achievable with standard intradural techniques due to the risk of neurological injury from adhesions. Posterior vertebral column subtraction

osteotomy is also a completely extradural technique, reducing the risk inherent with intradural surgery. However, with limited follow-up and few patients reported, further investigation is necessary before PVCSO is widely used.

As the efficacy of the above-mentioned techniques remain unproven, better identification of risk factors for retethering may help determine those at risk and lead to appropriate and earlier intervention in these patients. Some have proposed that such risk factors include younger age and a large placode at initial presentation of TCS.⁵ However, age has been shown in 2 separate reports to have no influence on pain, urinary, weakness, or sensory outcomes,²⁸ or any postoperative improvement.⁹ Similarly, we observed that age was not associated with improvement in sensory, motor, urinary, or weakness symptoms. Additionally, we observed that the average age of patients who experience retethering (9.26 years) was not different from the average age of those who did not experience retethering (10.66 years), suggesting that age may not be a risk factor for retethering.

A variety of dural grafting material is available, including adjacent fascia, bovine pericardium, Silastic, Gore-Tex, and MEDPOR, but it is not yet known if they slow or prevent retethering.^{1,47} Gore-Tex specifically has been suggested to be useful in closing dural defects, to be placed under a primary dural closure to provide a smooth layer to prevent adhesions, or to allow for the creation of an expanded dural compartment at the site of previous tethering.¹ Early work demonstrated that Gore-Tex limited the nature and extent of adhesion and resulted in a less inflammatory/foreign body response.³⁴ In a large retrospective review, duraplasty was not found to affect the rates of CSF leak, surgical site infection, or LOS in a variety of TCS causes.³⁹ Similarly, in TCS after MMC repair, we found that the use of a dural graft was not associated with an increased incidence of retethering, or increased incidence of any perioperative complications (CSF leak, surgical site infection, or wound dehiscence). Despite these promising data, duraplasty has mainly been found to confer some advantage against retethering in “complex” TCS origins. We did observe that patients with a dural graft experienced retethering at an average of 55 months, whereas the primary dural closure group experienced retethering at 45 months, but this did not reach statistical significance. It appears that dural grafts do not introduce an additional hazard, but their ability to prevent retethering remains debatable.

The analysis of scoliosis in spinal cord tethering has mainly focused on the response of preoperative curves to cord release. It is known that TCR can improve scoliosis,¹⁵ and that smaller preoperative curves are most likely to improve or stabilize following surgery.^{31,36,40} Furthermore, it has been shown that a tethered cord can be a cause of scoliosis in children with an MMC.³¹ However, the role of scoliosis in retethering has not been previously explored. In this series, children with preoperative scoliosis experienced retethering requiring untethering after index cord release significantly earlier than those without scoliosis. We propose 2 possible theories to account for this finding. First, the presence of a spinal curvature may increase the surface area over which the spinal nerve roots are in contact with the dura, which may promote an increasing

inflammatory reaction and scarring, causing earlier retethering. Alternatively, the presence of scoliosis might simply be an indicator of more severe dysraphism with accompanying lower-spinal cord dysfunction and presentation of symptoms that are interpreted as TCS symptoms leading to early untethering procedures. Further evaluation of these children is warranted to explore precisely the relationship between scoliosis and why these children undergo repeat untethering at an early time point.

The retrospective nature of this report should be considered in the interpretation of the data. The grading of outcomes as either improved, unchanged, or worsened is potentially an overall simplistic categorization and may be subject to bias. Additionally, patient selection pattern and surgeon preference introduces variation and potential bias that cannot be corrected retrospectively.

Conclusions

Patient age and duration of symptoms are not associated with retethering in children who undergo TCR following an initial repair of an MMC. The presence of scoliosis is associated with significantly earlier retethering. In patients who experience symptomatic improvement, sensory symptoms can be expected to improve before motor symptoms. The use of duraplasty is not associated with an increased incidence of perioperative complications or retethering as compared with primary dural repair. A CSF leak requiring revision occurred in approximately 11% of procedures and remains a significant complication. Considerable variation in the reporting of symptomatic response to TCR exists in the literature, likely a result of nonstandardized methods to assess symptoms and analyzing outcomes from multiple TCS origins. The establishment of a uniform method for symptom assessment in TCS may improve understanding of the response to TCR, which may help to better predict those patients most likely to respond to surgery.

Disclosure

Dr. Mehta received support from the AOA Medical Honor Society and the AANS for this study. 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 to the study and manuscript preparation include the following. Conception and design: all authors. Acquisition of data: Mehta, Ahmadi, Berenberg, Thomale, Haberl, Jallo, Ahn. Analysis and interpretation of data: Bettgowda, Mehta, Ahmadi, Berenberg, Haberl, Jallo, Ahn. Drafting the article: Bettgowda, Mehta, Ahmadi, Berenberg, Haberl, Jallo, Ahn. Critically revising the article: Bettgowda, Mehta, Jallo, Ahn. Reviewed final version of the manuscript and approved it for submission: all authors. Statistical analysis: Bettgowda, Jallo, Ahn. Administrative/technical/material support: Jallo, Ahn. Study supervision: Jallo, Ahn.

References

1. Aliredjo RP, de Vries J, Menovsky T, Grotenhuis JA, Mers J: The use of Gore-Tex membrane for adhesion prevention in tethered spinal cord surgery: technical case reports. **Neurosurgery** **44**:674–678, 1999
2. Archibeck MJ, Smith JT, Carroll KL, Davitt JS, Stevens PM: Surgical release of tethered spinal cord: survivorship analysis and orthopedic outcome. **J Pediatr Orthop** **17**:773–776, 1997
3. Balasubramaniam C, Laurent JP, McCluggage C, Oshman D, Cheek WR: Tethered-cord syndrome after repair of meningocele. **Childs Nerv Syst** **6**:208–211, 1990

Spinal cord tethering following myelomeningocele repair

4. Begeer JH, Wiertsema GP, Breukers SM, Mooy JJ, ter Weeme CA: Tethered cord syndrome: clinical signs and results of operation in 42 patients with spina bifida aperta and occulta. **Z Kinderchir** **44** (Suppl 1):5–7, 1989
5. Blount JP, Tubbs RS, Wellons JC III, Acakpo-Satchivi L, Bauer D, Oakes WJ: Spinal cord transection for definitive untethering of repetitive tethered cord. **Neurosurg Focus** **23**(2):E11, 2007
6. Bowman RM, McLone DG, Grant JA, Tomita T, Ito JA: Spina bifida outcome: a 25-year prospective. **Pediatr Neurosurg** **34**:114–120, 2001
7. Bowman RM, Mohan A, Ito J, Seibly JM, McLone DG: Tethered cord release: a long-term study in 114 patients. Clinical article. **J Neurosurg Pediatr** **3**:181–187, 2009
8. Chin-Peuckert L, Komlos M, Rennick JE, Jednak R, Capolicchio JP, Salle JL: What is the variability between 2 consecutive cystometries in the same child? **J Urol** **170**:1614–1617, 2003
9. Daszkiewicz P, Barszcz S, Roszkowski M, Maryniak A: Tethered cord syndrome in children—impact of surgical treatment on functional neurological and urological outcome. **Neurol Neurochir Pol** **41**:427–435, 2007
10. Drake JM: Surgical management of the tethered spinal cord—walking the fine line. **Neurosurg Focus** **23**(2):E8, 2007
11. Fone PD, Vapnek JM, Litwiller SE, Couillard DR, McDonald CM, Boggan JE, et al: Urodynamic findings in the tethered spinal cord syndrome: does surgical release improve bladder function? **J Urol** **157**:604–609, 1997
12. Gilbert JN, Jones KL, Rorke LB, Chernoff GF, James HE: Central nervous system anomalies associated with meningomyelocele, hydrocephalus, and the Arnold-Chiari malformation: reappraisal of theories regarding the pathogenesis of posterior neural tube closure defects. **Neurosurgery** **18**:559–564, 1986
13. Haberl H, Tallen G, Michael T, Hoffmann KT, Benndorf G, Brock M: Surgical aspects and outcome of delayed tethered cord release. **Zentralbl Neurochir** **65**:161–167, 2004
14. Hendrick EB, Hoffman HJ, Humphreys RP: The tethered spinal cord. **Clin Neurosurg** **30**:457–463, 1983
15. Herman JM, McLone DG, Storrs BB, Dauser RC: Analysis of 153 patients with myelomeningocele or spinal lipoma reoperated upon for a tethered cord. Presentation, management and outcome. **Pediatr Neurosurg** **19**:243–249, 1993
16. Hoffman HJ, Taecholarn C, Hendrick EB, Humphreys RP: Management of lipomyelomeningoceles. Experience at the Hospital for Sick Children, Toronto. **J Neurosurg** **62**:1–8, 1985
17. Hsieh PC, Ondra SL, Grande AW, O'Shaughnessy BA, Bierbrauer K, Crone KR, et al: Posterior vertebral column subtraction osteotomy: a novel surgical approach for the treatment of multiple recurrences of tethered cord syndrome. Technical note. **J Neurosurg Spine** **10**:278–286, 2009
18. Hudgins RJ, Gilreath CL: Tethered spinal cord following repair of myelomeningocele. **Neurosurg Focus** **16**(2):E7, 2004
19. Inoue HK, Kobayashi S, Ohbayashi K, Kohga H, Nakamura M: Treatment and prevention of tethered and retethered spinal cord using a Gore-Tex surgical membrane. **J Neurosurg** **80**:689–693, 1994
20. Kang JK, Lee KS, Jeun SS, Lee IW, Kim MC: Role of surgery for maintaining urological function and prevention of retethering in the treatment of lipomyelomeningocele: experience recorded in 75 lipomyelomeningocele patients. **Childs Nerv Syst** **19**:23–29, 2003
21. Kaplan EL, Meier P: Nonparametric estimator from incomplete observations. **J Am Stat Assoc** **53**:457–481, 1958
22. Kaplan WE, McLone DG, Richards I: The urological manifestations of the tethered spinal cord. **J Urol** **140**:1285–1288, 1988
23. Lad SP, Patil CG, Ho C, Edwards MS, Boakye M: Tethered cord syndrome: nationwide inpatient complications and outcomes. **Neurosurg Focus** **23**(2):E3, 2007
24. Lagae L, Verpoorten C, Casaer P, Vereecken R, Fabry G, Plets C: Conservative versus neurosurgical treatment of tethered cord patients. **Z Kinderchir** **45** (Suppl 1):16–17, 1990
25. Lee TT, Arias JM, Andrus HL, Quencer RM, Falcone SF, Green BA: Progressive posttraumatic myelomalacic myelopathy: treatment with untethering and expansive duraplasty. **J Neurosurg** **86**:624–628, 1997
26. Lew SM, Kothbauer KF: Tethered cord syndrome: an updated review. **Pediatr Neurosurg** **43**:236–248, 2007
27. Linder M, Rosenstein J, Sklar FH: Functional improvement after spinal surgery for the dysraphic malformations. **Neurosurgery** **11**:622–624, 1982
28. Maher CO, Goumnerova L, Madsen JR, Proctor M, Scott RM: Outcome following multiple repeated spinal cord untethering operations. **J Neurosurg** **106** (6 Suppl):434–438, 2007
29. McCormick PC, Torres R, Post KD, Stein BM: Intramedullary ependymoma of the spinal cord. **J Neurosurg** **72**:523–532, 1990
30. McLone DG: Technique for closure of myelomeningocele. **Childs Brain** **6**:65–73, 1980
31. McLone DG, Herman JM, Gabrieli AP, Dias L: Tethered cord as a cause of scoliosis in children with a myelomeningocele. **Pediatr Neurosurg** **16**:8–13, 1990–1991
32. McLone DG, Naidich TP: Laser resection of fifty spinal lipomas. **Neurosurgery** **18**:611–615, 1986
33. Palmer LS, Richards I, Kaplan WE: Subclinical changes in bladder function in children presenting with nonurological symptoms of the tethered cord syndrome. **J Urol** **159**:231–234, 1998
34. Park YK, Tator CH: Prevention of arachnoiditis and postoperative tethering of the spinal cord with Gore-Tex surgical membrane: an experimental study with rats. **Neurosurgery** **42**:813–824, 1998
35. Phuong LK, Schoeberl KA, Raffel C: Natural history of tethered cord in patients with meningomyelocele. **Neurosurgery** **50**:989–995, 2002
36. Pierz K, Banta J, Thomson J, Gahm N, Hartford J: The effect of tethered cord release on scoliosis in myelomeningocele. **J Pediatr Orthop** **20**:362–365, 2000
37. Reigel D: Tethered spinal cord, in Humphreys R (ed): **Concepts in Pediatric Neurosurgery**. Basel: Karger, 1983, Vol 4, pp 142–164
38. Sakamoto H, Hakuba A, Fujitani K, Nishimura S: Surgical treatment of the retethered spinal cord after repair of lipomyelomeningocele. **J Neurosurg** **74**:709–714, 1991
39. Samuels R, McGirt MJ, Attenello FJ, Garcés Ambrossi GL, Singh N, Solakoglu C, et al: Incidence of symptomatic retethering after surgical management of pediatric tethered cord syndrome with or without duraplasty. **Childs Nerv Syst** **25**:1085–1089, 2009
40. Sarwark JF, Weber DT, Gabrieli AP, McLone DG, Dias L: Tethered cord syndrome in low motor level children with myelomeningocele. **Pediatr Neurosurg** **25**:295–301, 1996
41. Shurtleff DB, Duguay S, Duguay G, Moskowitz D, Weinberger E, Roberts T, et al: Epidemiology of tethered cord with meningomyelocele. **Eur J Pediatr Surg** **7** (Suppl 1):7–11, 1997
42. Talamonti G, D'Aliberti G, Collice M: Myelomeningocele: long-term neurosurgical treatment and follow-up in 202 patients. **J Neurosurg** **107** (5 Suppl):368–386, 2007
43. Tamaki N, Shirataki K, Kojima N, Shouse Y, Matsumoto S: Tethered cord syndrome of delayed onset following repair of myelomeningocele. **J Neurosurg** **69**:393–398, 1988
44. Tubbs RS, Oakes WJ: A simple method to deter retethering in patients with spinal dysraphism. **Childs Nerv Syst** **22**:715–716, 2006
45. Winston K, Hall J, Johnson D, Micheli L: Acute elevation of intracranial pressure following transection of non-functional spinal cord. **Clin Orthop Relat Res** (128):41–44, 1977
46. Yamada S, Won DJ, Yamada SM: Pathophysiology of tethered cord syndrome: correlation with symptomatology. **Neurosurg Focus** **16**(2):E6, 2004
47. Zide B, Constantini S, Epstein FJ: Prevention of recurrent tethered spinal cord. **Pediatr Neurosurg** **22**:111–114, 1995

Manuscript submitted March 24, 2010.

Accepted August 18, 2010.

Address correspondence to: Chetan Bettegowda, M.D., Ph.D., Department of Neurosurgery, The Johns Hopkins Hospital, 600 North Wolfe Street, Meyer 8-161, Baltimore, Maryland 21287. email: cbetteg1@jhmi.edu.