



Functional Outcomes in Children With Myelomeningocele Following Orthopedic Scoliosis Correction With or Without Prior Spinal Cord Untethering

Citation

En'Wezoh, Derick C. 2016. Functional Outcomes in Children With Myelomeningocele Following Orthopedic Scoliosis Correction With or Without Prior Spinal Cord Untethering. Doctoral dissertation, Harvard Medical School.

Permanent link

http://nrs.harvard.edu/urn-3:HUL.InstRepos:40620244

Terms of Use

This article was downloaded from Harvard University's DASH repository, and is made available under the terms and conditions applicable to Other Posted Material, as set forth at http://nrs.harvard.edu/urn-3:HUL.InstRepos:dash.current.terms-of-use#LAA

Share Your Story

The Harvard community has made this article openly available. Please share how this access benefits you. <u>Submit a story</u>.

Accessibility

Title: Functional Outcomes in Children with Myelomeningocele Following
Orthopedic Scoliosis Correction With or Without Prior Spinal Cord Untethering

Authors:

Derick C. En'Wezoh, B.S.¹, Kareem Halim, M.D.¹, Chris Nguyen, M.A.², Lawrence Karlin M.D.^{1,3,5}, Benjamin C. Warf, M.D.^{1,4,5}

¹Harvard Medical School, ²Stanford University, ³Boston Children's Hospital Department of Orthopedic Surgery, ⁴Boston Children's Hospital Department of Neurosurgery, ⁵Boston Children's Hospital Center for Spina Bifida and Spinal Cord Conditions

Last Revised: September 2015 Page 1 of 24

Abstract:

Purpose: Patients with surgically corrected myelomeningocele have a distally anchored spinal cord. These patients are predisposed to developing progressive scoliosis, often requiring surgery for curve correction and spinal fusion. Some authors have advocated prophylactic spinal cord untethering prior to corrective spine surgery to protect the tethered cord from injury. The purpose of this study was to identify any functional benefit from spinal cord untethering in otherwise asymptomatic myelomeningocele patients prior to corrective spinal surgery.

Methods: We retrospectively identified 45 patients who had corrective spinal surgery at Boston Children's Hospital between 1990 and 2011 who met the inclusion criteria. The patients were analyzed in two ways. 1) Pre- and post-fusion functional status of those who had an untethering operation at the time of or within the 6 months prior to spinal fusion (UT < 6m, 10 patients) was compared to that of those who were not untethered during this six-month period (NUT < 6m, 35 patients). 2) Pre- and post- fusion functional status of those who had an untethering operation at any time prior to spinal fusion (UT anytime, 25 patients) were compared to those who had never been untethered (NUT anytime, 20 patients). We collected demographic, radiographic, and pre/post-fusion functional status data for each patient from medical record review. Functional status was quantified using the Necker-Enfants Malades (NEM) and modified

Last Revised: 1/22/16 Page 2 of 24

Hoffer scoring systems. Spine radiographs were available for measurement of pre- and post-operative curve severity in 33 patients (20 UT, 13 NUT).

Results: For those with available radiographs, the average scoliotic curvature was 83 degrees pre-operatively and 22.3 degrees post-operatively for an average correction of 73% (73% for those who had been previously untethered and 71% for those who had never been untethered). In the first analysis, the mean change in NEM scores following fusion was +0.2 and +0.06 for the UT and NUT groups, respectively (p=0.71); and, mean change in Hoffer score was -0.2 and +0.06 for UT and NUT groups, respectively (p=0.13). In the second analysis, the mean change in NEM following fusion was +0.24 and +1.17 for the UT and NUT groups, respectively (p=0.29); and, mean change in Hoffer score was +0.65 and +0.00 for the UT and NUT groups, respectively (p=1.00).

Conclusions: This retrospective study was unable to identify any advantage of recent or remote spinal cord untethering in regard to overall functional outcome following corrective spinal surgery in MM patients.

Keywords: myelomeningocele, tethered spinal cord, scoliosis, spinal fusion, functional outcome

Last Revised: 1/22/16 Page 3 of 24

Background:

Myelomeningocele (MM) results from a failure of primary neurulation and requires surgical closure of the exposed neural elements to prevent infection. ^{2,16, 21} Effectively, all of these children secondarily develop a "tethered spinal cord" (a term originally coined by Hoffman, Hendrick, and Humphreys in reference to tight filum terminale) that can be observed on radiographic imaging. ^{3,7,9,10,13,21,25} Spinal cord tethering in this instance typically results from arachnoid scar formation and dorsal adherence of the initially exposed spinal cord placode to the dural repair site. ^{10,13} This can lead to chronic, repeated spinal cord injury likely resulting from decreased caudal spinal cord blood flow. ^{1,10,19,22,25} Approximately 10-30% of these patients will become symptomatic with loss of baseline motor function, gait deterioration, pain, new orthopedic deformities (such as progressive scoliosis or lower extremity deformity), spasticity, or altered urologic function. ^{3,7,10,11,14,21,23}

Patients with repaired MM frequently develop scoliosis, and tethered spinal cord may be a contributing cause. ^{1, 4,12, 13, 18, 24} An evidence based literature review performed by Dias found the incidence of scoliosis among children with repaired MM ranged from 40 to 85%. Herman and colleagues reported that 51% of children with symptomatic tethered spinal cord following closure of MM presented with scoliosis. Al-Holou and co-authors reported progressive scoliosis as the most common presentation (48%) in their series of

Last Revised: 1/22/16 Page 4 of 24

children with MM or lipomyelomeningocele undergoing reoperation for secondary tethered cord, including those in whom scoliosis was the only symptom. ¹ If the curve is not severe (less than around 50 degrees) untethering can arrest progression or lead to improvement, while more severe curves typically require operative correction, which increases the vertical height of the spinal column. ^{12, 13, 13, 14} Theoretical concern for developing new neurologic deficits under those circumstances, when the spinal cord is anchored distally, has led some to advocate prophylactic untethering of the spinal cord in these patients prior to curve correction. ^{1, 3} Those supporting prophylactic untethering view isolated progressive scoliosis as a symptom of tethering worthy of treatment and also suggest this may reduce the risk of neurological injury that could result from operative correction of the curve, while those opposed are concerned about increased operative risk and lack of data to support its benefit. ^{1,3,5,13,19}

Samdani and colleagues retrospectively reviewed seventeen patients with MM who underwent scoliosis corrective surgery, but had not recently had their spinal cord untethered. Since none of the study patients experienced any functional loss following spine surgery, these authors concluded that, for otherwise asymptomatic MM patients, prophylactic spinal cord untethering may be unnecessary. Al-Holoul and coauthors found no difference in functional outcome between their 34 patients untethered prophylactically prior to spinal fusion and those operated for clinical symptoms of tethering, but had no control patients undergoing fusion without untethering for comparison.

Last Revised: 1/22/16 Page 5 of 24

Historically, in the Boston Children's Hospital Center for Spina Bifida and Spinal Cord Conditions, there has been surgeon-dependent variation in practice regarding prophylactic untethering in MM patients prior to spinal fusion. Here, we present a retrospective analysis of changes in objective scaled functional scores before and after scoliosis curve correction in these patients, including both those with and without prior untethering. The goal of this study was to assess whether this variance in practice had any overall affect on functional outcome.

Methods:

Study Design

This retrospective study was approved by the Institutional Review Board (IRB) of Boston Children's Hospital. Initially, a query was performed in which operative notes in the Boston Children's Hospital medical record system between the years1990 and 2011 were scanned for the terms "myelodysplasia or spina bifida" and "scoliosis." This yielded 74 patients (**Figure 1**). Patients were then included in the study if they had: (1) a documented diagnosis of MM, (2) MRI imaging of the spinal cord, (3) no clinical signs or symptoms of tethered spinal cord other than scoliosis, (4) an operative report documenting the scoliosis correction and fusion procedure, and (5) at least 6 months of post-fusion follow-up documentation. Of the 74 patients sampled, 45 met the study's eligibility criteria (**Figure 1**). Patients were excluded from the study for not having a clearly documented diagnosis of MM (n=3), not having accessible MRI imaging (n=6),

Last Revised: 1/22/16 Page 6 of 24

not having had a scoliosis correction and fusion operation (n=8) and having insufficient post-fusion follow-up (n=12).

Analyses

Two separate analyses were performed on all 45 patients who met the study's inclusion criteria (**Figure 1**). In Analysis 1, the intervention was an untethering operation within the 6 months prior to scoliosis correction (including those who were untethered under the same anesthesia but prior to spinal manipulation). Within Analysis 1, 10 patients received the intervention (Untethered, UT <6mo Group), while 35 patients served as controls (Not Untethered, NUT <6mo Group). Patients who had undergone a previous spinal cord untethering more than 6 months prior to their scoliosis correction were included in the control group. For patients who had more than one corrective spinal operation, only the first operation was included in the analysis.

Analysis 2 also included all 45 patients who met the study's inclusion criteria. In this analysis, the intervention was an untethering operation at anytime prior to (including under the same anesthesia) scoliosis corrective surgery. Within Analysis 2, 25 patients received the intervention (UT at any time), while 20 patients who had never undergone spinal cord untethering served as controls (NUT any time).

Last Revised: 1/22/16 Page 7 of 24

Surgical Technique

Corrective spinal surgery was accomplished using standard techniques with segmental instrumentation and fusion. Intraoperative spinal cord monitoring was routinely used. Spinal cord untethering, when done, was performed using microsurgical technique to free the distal cord from its immobilization by arachnoid scar. Intraoperative monitoring of nerve root responses, including the anal sphincter musculature, was routinely employed. The decision to prophylactically untether the spinal cord within 6 months of the spinal fusion was essentially random, being epoch and surgeon dependent.

Data Collection

We collected demographic, clinical, radiographic, and pre/post-fusion functional status data for each patient. Demographic information consisted of age at the time of fusion, sex, and race. Clinical and radiographic data included the deformity pattern, MRI appearance consistent with a tethered spinal cord, presence of a shunt, and whether there was imaging evidence of syrinx, cyst, fatty filum, and/or Chiari 2 malformation prior to scoliosis corrective surgery. Functional status data characterized the degree of motor, sensory, bladder, bowel and ambulatory impairment experienced by patients.

Radiographic Data

Upright PA or AP and lateral radiographs were evaluated. These included studies done within 1 month prior to the surgery and the first upright study

Last Revised: 1/22/16 Page 8 of 24

following the surgery. All post-operative radiographs were performed within 3 months of the procedure. The deformity pattern was recorded. The major scoliotic curvature was measured using the Cobb technique and the pelvic obliquity using the method of Osebold.¹⁵

NEM Score

For our analysis, we collected pre-fusion and post-fusion information pertaining to the motor, sensory, bladder, bowel, and ambulatory functional status of the patients. For patients who underwent simultaneous untethering or untethering within 6 months prior to the spinal fusion, the functional status data was that from just prior to the untethering operation. To quantitatively compare functional outcomes between the intervention (UT) and control (NUT) groups, we used two previously validated functional scales. We employed the Necker-Enfants Malades (NEM) scale to evaluate motor, sensory, bladder, and bowel function (Table 1). The NEM scale is a 5-point scale used to quantitatively assess the functional and social consequences of congenital spinal malformations.^{1,17}

Modified Hoffer Score

We also used the modified Hoffer ambulatory scale (**Table 2**) to evaluate ambulatory function. Initially developed by Hoffer et al., and later modified by Schoenmakers et al., the modified Hoffer scale is used to quantitatively assess ambulatory ability in patients who undergo spinal cord untethering.^{8,20} For both

Last Revised: 1/22/16 Page 9 of 24

the NEM and modified Hoffer scales, lower scores are associated with more severe functional deficits.

Pre-operative NEM and Hoffer scores were assigned to the intervention (UT) groups using information in their most recent medical record data preceding their spinal cord untethering operation for the UT <6month group or within 6 months prior to their spinal fusion for all others (control groups, and those untethered prior to 6 months before the spinal fusion). Post-operative NEM and Hoffer scores were assigned at approximately 6 months after the fusion operation for both the intervention (UT) and control (NUT) groups.

Data Analysis:

We calculated the difference in post-fusion and pre-fusion mean NEM and Hoffer scores and then conducted a t-test to assess for significance of any differences between intervention and control groups in regard to overall change in motor, sensory, bladder, bowel, and ambulatory functional status.

Results:

Patient Population

Analysis 1 included the 45 patients who met the inclusion criteria for our study. As displayed in **Table 3**, the intervention (UT <6mo) group included 10 patients (10% male) with an average age of 14.52±5.91 years. The control (NUT

Last Revised: 1/22/16 Page 10 of 24

<6mo) group included 35 patients (49% male) with an average age of 13.97±3.46 years. The intervention and control groups were similar in racial demographics.

Analysis 2 also included the 45 patients who met the inclusion criteria for our study. Those in the group untethered at any time prior to their fusion included 25 patients (32% male) with an average age of 14.13±4.37 years. The control group included 20 patients (50% male) with an average age of 14.05±3.74 years. The intervention and control groups were similar in racial demographics.

MRI Findings

Many of the patients demonstrated more than one radiographic abnormality (**Table 3**). In Analysis 1, the intervention (UT <6mo) group frequently included those with hydromyelia (40%), cysts (30%), fatty fila (30%), Chiari 2 malformations (90%), and ventricular shunts (80%). In the control (NUT <6mo) group, hydromyelia (57%), cysts (23%), fatty fila (29%), Chiari 2 malformations (83%), and shunts (91%) were similarly represented.

In Analysis 2, hydromyelia (52%), cysts (28%), fatty fila (28%), Chiari 2 malformations (88%), and shunts (84%) were present in the intervention (UT any time) group. In the control (NUT anytime) group, hydromyela (55%), cysts (20%), fatty fila (30%), Chiari 2 malformations (80%), and shunts (95%) were also similarly represented.

Last Revised: 1/22/16 Page 11 of 24

Baseline Disability

All patients involved in the study were diagnosed with severe progressive scoliosis (100%) for whom corrective surgery and fusion were indicated. As shown in **Table 4**, the number of patients with any degree of baseline motor, sensory, bladder, bowel or ambulatory functioning below normal (defined as a less than perfect NEM or modified Hoffer score) was documented.

In both Analysis 1 and 2, patients in the treatment and control groups demonstrated similar rates of baseline motor dysfunction, sensory deficit, bladder incontinence, bowel dysfunction, and ambulatory impairment. With the exception of bowel dysfunction, less than 15% of patients had normal functioning at baseline in any of the functional categories captured by the NEM scale and the modified Hoffer scale. All of the patients were asymptomatic for tethered cord at the time of the spinal fusion or at the time of untethering <6m prior to spinal fusion. Otherwise stated, none of the patients had recently experienced a worsening in their baseline degree of functioning that would necessitate performing an operation to release their spinal cord.

Operative Results:

All of the patients in this study had a fusion operation in an effort to correct their scoliosis. Many patients had fusions extending through more than one spinal region. For patients in the UT <6mo group of Analysis 1, 100% had a thoracic level fusion, 90% had a lumbar level fusion, and 60% had a sacral level fusion. For patients in the NUT <6mo group of Analysis 1, 100% had a thoracic

Last Revised: 1/22/16 Page 12 of 24

level fusion, 89% had a lumbar level fusion, and 37% had a sacral level fusion. Similar percentages were observed in Analysis 2.

A complete set of radiographs was available for retrospective analysis in 32 of the patients. Of these, there were 26 thoracolumbar curve patterns of which 13 were right and 13 left. The apices were: T8-1, T10-1, T11-1, T12-4, L1-8, L2-10, and L3-1. There were 2 double curve patterns, both right thoracic and left lumbar, 2 right thoracic and 1 right lumbar and 1 left lumbar. The average scoliotic curvature was 83 degrees pre-operatively and 22.3 degrees post-operatively for an average correction of 73%. The average pelvic obliquity was 22 degrees pre-operatively and 5 degrees post-operatively.

Functional Outcomes (Analysis 1)

Motor, sensory, bladder, bowel and ambulatory function were evaluated using the previously described NEM and modified Hoffer scores. As displayed in **Table 5**, patients in the intervention (UT <6mo) group of Analysis 1 experienced a mean post-fusion improvement in total NEM score (0.20±1.32 NEM Points), sensory function (0.10±0.99 NEM points) and bladder function (0.20±0.63 NEM points); whereas, they demonstrated mean post-fusion stability or worsening in motor function (-0.10±0.88 NEM points), bowel function (0.00±0.00 NEM points) and ambulatory function (-0.20±0.79 Hoffer points).

In the control (NUT <6mo) group of Analysis 1, patients demonstrated a mean post-fusion improvement in total NEM score (0.06±1.00 NEM points),

Last Revised: 1/22/16 Page 13 of 24

motor function (0.03 ± 0.17 NEM points), bladder function (0.06 ± 0.34 NEM points), and ambulatory function (0.06 ± 0.34 Hoffer points). These same patients experienced mean post-fusion stability or worsening in sensory function (-0.03 ± 0.92 NEM points) and bowel function (0.00 ± 0.34 NEM points).

Between the intervention (UT <6 mo) and control (NUT <6 mo) groups of Analysis 1, there was not a significant difference between the mean post-fusion changes in the NEM score, the modified Hoffer score, or any of the NEM parameters (P>0.05).

Functional Outcomes (Analysis 2)

As shown in **Table 6**, patients in the intervention (UT at any time) group of Analysis 2 demonstrated a mean post-fusion improvement in total NEM score (0.24±0.97 NEM points), sensory function (0.16±0.80 NEM points) and bladder function (0.08±0.40 NEM points). These patients demonstrated mean post-fusion stability or worsening in motor function (0.00±0.58 NEM points), bowel function (0.00±0.08 NEM points) and ambulatory function (0.00±0.65 Hoffer points).

In the NUT group of Analysis 2, patients demonstrated a mean post-fusion improvement in bladder function (0.10±0.45 NEM points) only. These same patients experienced mean post-fusion stability or worsening in total NEM score (-0.10±1.17 NEM points), motor function (0.00±0.00 NEM points), sensory

Last Revised: 1/22/16 Page 14 of 24

function (-0.20 \pm 1.06 NEM points), bowel function (0.00 \pm 0.00 NEM points), and ambulatory function (0.00 \pm 0.00 Hoffer points).

Between the UT and NUT groups in Analysis 2, there was not a significant difference between the mean post-fusion changes in the NEM score, the modified Hoffer score, or any of the NEM parameters (P>0.05).

Discussion:

In some MM patients, progressive scoliosis can be an indicator of clinically significant spinal cord tethering, and untethering of the cord is warranted when the goal is to arrest or reverse curve progression, thus avoiding or delaying corrective spinal surgery. However, for progressive curves greater than 50%, spinal fusion is generally not avoided with untethering. When there are no other indicators of spinal cord tethering in a patient with an advanced curve in need of correction, the question arises as to whether untethering prior to the resultant increase in spinal column vertical height is warranted. This is based upon the assumption that additional traction will be placed on the spinal cord, and possibly lead to new neurologic deficits. The results of this analysis do not demonstrate a statistically significant clinical benefit to untethering the spinal cord in otherwise asymptomatic MM patients prior to or during corrective spinal surgery. Patients with MM who underwent an untethering procedure at the time of or within 6 months prior to having a spinal fusion did not have significantly better or worse motor, sensory, bladder, bowel or ambulatory functional outcomes than patients

Last Revised: 1/22/16 Page 15 of 24

who did not have an untethering procedure performed in that time frame (**Table 5**). Further, patients with MM who underwent an untethering procedure anytime prior to having a scoliosis fusion operation did not demonstrate significantly better or worse functional outcomes than patients who never had an untethering procedure (**Table 6**).

These findings are consistent with the results of a retrospective study performed by Samdani and colleagues. ¹⁹ They found that no new negative neurological, urological, motor, or sensory outcomes occurred in a group of 17 patients with MM who had undergone scoliosis corrective surgery, but hadn't had a recent untethering operation. Hence, they concluded that untethering the spinal cord prior to scoliosis corrective surgery may be unnecessary.

Our larger study included both patients that had and had not undergone an untethering operation for a comparative analysis. Yet, it is still limited by its retrospective nature and small sample size. An argument could be advanced that the study did not have enough power to reveal meaningful differences in the post-fusion functional status between our intervention and control groups.

Another limitation of this study is the use of the NEM and modified Hoffer scores to quantitatively assess the functional status of patients. Despite being validated and previously used in the literature, these scores may not be sensitive enough to quantify small yet meaningful functional changes for patients post-

Last Revised: 1/22/16 Page 16 of 24

operatively. For example, in a patient with a post-fusion decrease in the frequency in which she must intermittently catheterize, her bladder NEM score would remain unchanged. Yet, this would represent an improvement in functional outcome not reflected by the NEM score. Also, in the Boston Children's Hospital Center for Spina Bifida and Spinal Cord Conditions, full urodynamic screening (including sphincter EMG) is now routinely obtained prior to untethering and at around 3 months following the procedure. But routine pre- and post-spinal fusion urodynamic studies have not been routinely performed in the past. Thus, subtle, subclinical changes in bladder or sphincter function might have gone undetected.

Despite these limitations, our results are clinically important in that they failed to demonstrate any difference in outcome between the two management strategies. Hence, our study provides no compelling evidence of any benefit to prophylactic spinal cord untethering in these patients prior to scoliosis surgery. Although this study might provide equipoise for a carefully designed prospective multi-institution study with a larger sample size and more sensitive functional outcome parameters, the practicality of doing a sufficiently powered study in light of evidence that functional downgrading is unusual with or without untethering would likely temper enthusiasm.

What might account for the apparently low risk of correcting a significant curve in the face of a tethered spinal cord in these patients? It might seem that increasing the vertical height of the spinal column by straightening the curve

Last Revised: 1/22/16 Page 17 of 24

would place increased tension on the caudally anchored spinal cord. However, unlike the relationship of a bowstring to its bow, the spinal cord has grown in length along with, and largely follows the curve of, the spinal canal, with the cervical and thoracic segments of the spinal cord being anchored to the lateral thecal sac by the dentate ligaments. The effect of the distally tethered placode is likely limited to the more caudal cord immediately proximal to the point of tethering. ²² The vertical height of the column may be increased without significantly increasing the actual length of the spinal canal or the tension on the distal spinal cord. The latter would be contingent on the degree of actual intersegmental distraction rather than on the extent of angular curve correction, which is largely accomplished by translation rather than distraction.

Conclusion:

This retrospective study failed to demonstrate any functional benefit to prophylactic spinal cord untethering in asymptomatic MM patients prior to corrective surgery for scoliosis. We found no compelling justification for prophylactic untethering prior to corrective spinal surgery when there is no other clinical indication for doing so.

Last Revised: 1/22/16 Page 18 of 24

References:

- Al-Holou WN, Muraszko KM, Garton HJ, Buchman SR, Maher CO. The outcome of tethered cord release in secondary and multiple repeat tethered cord syndrome. J Neurosurg Pediatr 4:28–36, 2009
- 2. Albright AL, Adelson PD, Pollack IF (eds). **Principles and Practice of Pediatric Neurosurgery**. 2nd ed., New York: Thieme, 2008
- 3. Bowman RM, Mohan A, Ito J, Seibly JM, McLone DG. Tethered cord release: a long-term study in 114 patients. **J Neurosurg Pediatr 3**:181–7, 2009
- Dias MS. Neurosurgical causes of scoliosis in patients with myelomeningocele: an evidence-based literature review. J Neurosurg 103:24–35, 2005
- 5. Hamzaoglu A, Ozturk C, Tezer M, Aydogan M, Sarier M, Talu U. Simultaneous surgical treatment in congenital scoliosis and/or kyphosis associated with intraspinal abnormalities. **Spine (Phila Pa 1976) 32**:2880–4, 2007
- 6. Herman J, McLone D, Storrs B, Dauser R: Analysis of 153 patients with myelomeningocele or spinal lipoma reoperated upon for a tethered cord. Presentation, management and outcome. **Pediatr Neurosurg 19**:243–9, 1993
- 7. Hertzler D a, DePowell JJ, Stevenson CB, Mangano FT. Tethered cord syndrome: a review of the literature from embryology to adult presentation.

 Neurosurg Focus 29:E1, 2010
- 8. Hoffer MM, Feiwell E, Perry R, Perry J, Bonnett C. Functional ambulation in patients with myelomeningocele. **J Bone Joint Surg Am 55**:137–148, 1973
- 9. Hoffman HJ, Hendrick EB, Humphreys RP. The tethered spinal cord: its protean manifestations, diagnosis and surgical correction. **Childs Brain 2**:145–155, 1976
- 10. Hudgins RJ, Gilreath CL. Tethered spinal cord following repair of myelomeningocele. **Neurosurg Focus 16**:E7, 2004
- Maher CO, Goumnerova L, Madsen JR, Proctor M, Scott RM. Outcome following multiple repeated spinal cord untethering operations. J Neurosurg 106:434–8, 2007
- 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, 1991
- 13. McLone DG, La Marca F. The tethered spinal cord: diagnosis, significance, and management. **Semin Pediatr Neurol 4**:192–208, 1997
- 14. Mehta V a, Bettegowda C, Ahmadi S a, Berenberg P, Thomale U-W, Haberl E-J, et al. Spinal cord tethering following myelomeningocele repair. **J Neurosurg Pediatr 6**:498–505, 2010
- 15. Osebold WR, Mayfield JK, Winter RB, Moe JH. Surgical treatment of paralytic scoliosis associated with myelomeningocele. **J Bone Joint Surg Am 64**:841–856, 1982

Last Revised: 1/22/16 Page 19 of 24

- 16. Piatt JH. Treatment of myelomeningocele: a review of outcomes and continuing neurosurgical considerations among adults. **J Neurosurg Pediatr 6**:515–25, 2010
- 17. Pierre-Kahn a, Zerah M, Renier D, Cinalli G, Sainte-Rose C, Lellouch-Tubiana a, et al. Congenital lumbosacral lipomas. **Childs Nerv Syst** 13:298–334, 1997
- 18. Reigel D, Tchernoukha K, Bazmi B, Kortyna R, Rotenstein D. Change in spinal curvature following release of tethered spinal cord associated with spina bifida. **Pediatr Neurosurg 20**:30–42, 1994.
- 19. Samdani AF, Fine AL, Sagoo SS, Shah SC, Cahill PJ, Clements DH, et al. A patient with myelomeningocele: is untethering necessary prior to scoliosis correction? **Neurosurg Focus 29**:E8, 2010
- 20. Schoenmakers M a GC, Gooskens RHJM, Gulmans V a M, Hanlo PW, Vandertop WP, Uiterwaal CSPM, et al.. Long-term outcome of neurosurgical untethering on neurosegmental motor and ambulation levels. **Dev Med Child Neurol 45**:551–555, 2003
- 21. Talamonti G, D'Aliberti G, Collice M. Myelomeningocele: long-term neurosurgical treatment and follow-up in 202 patients. **J Neurosurg 107**:368–86, 2007
- 22. Tani S, Yamada S, Knighton RS. Extensibility of the lumbar and sacral cord. Pathophysiology of the tethered spinal cord in cats. **J Neurosurg 66**:116–123, 1987
- 23. Tarcan T, Onol FF, Ilker Y, Simsek F, Simek F, Ozek M. Does surgical release of secondary spinal cord tethering improve the prognosis of neurogenic bladder in children with myelomeningocele? **J Urol 176**:1601–1606
- 24. Trivedi J, Thomson JD, Slakey JB, Banta J V, Jones PW. Clinical and radiographic predictors of scoliosis in patients with myelomeningocele. **J Bone Joint Surg Am 84-A**:1389–1394, 2002
- 25. Warf B. Pathophysiology of tethered spinal cord, in **Pediatric Neurosurgery: Surgery of the Developing Nervous System, 4th Edition.** Philadelphia: W. B. Saunders, 2001, pp 282–288
- 26. Winter RB, Haven JJ, Moe JH, Lagaard SM: Diastematomyelia Spine and Congenital Deformities *. **Surgery**:1974

Last Revised: 1/22/16 Page 20 of 24

Tables:

Score	Motor	Sensation	Bladder	Anus
1	Wheelchair	Skin ulceration or amputation	Incontinence day and night	Incontinence
2	Major orthesis or two crutches	Pain	Nocturnal incontinence	Painful constipation
3	Minor or distal orthesis	Painless sensory defect	Intermittent catheterization	Normal
4	Fatigue on walking	Normal	Dysuria or infections or stress incontinence	
5	Normal		Normal	

Table 1. NEM Scoring Criteria^{1,17}

Score	Symptom
1	Nonambulator
2	Exercise ambulator (only in therapeutic situations)
3	Household ambulator (using crutch or brace indoors, wheel-chair outdoors)
4	Community ambulator (ambulate outdoors w/ or w/o brace, uses wheelchair
	for longer distances)
5	Normal ambulator

Table 2. Modified Hoffer Score Criteria^{8,20}

Last Revised: 1/22/16 Page 21 of 24

	Analysis 1 Group					Analysis 2 Group			
	Untet	hered + Fusion		ethered + Fusion			hered) + Fusion		tethered) + Fusion
Number of patients, n (%)	10	TUSIOII	35	Trusion		25	j + rusion	20	j + Fusion
Mean age in years +/- SD	14.52	E 01	13.97	2.46		14.13	1 27	14.05	2 7/
•					:	•			
Male, n (%)	1	10%	17	49%		8	32%	10	50%
Race, n (%)		2221		2221					
White	9	90%		80%			84%	16	80%
Black or African American	0	0%	1	3%		0	0%	1	5%
Native Hawaiian or Pacific									
Islander	0	0%	1	3%		0	0%	1	5%
Asian	0	0%	1	3%		1	4%	0	0%
Hispanics	0	0%	0	0%	:	0	0%	0	0%
Other	1	10%	4	11%		3	12%	2	10%
Radiographic features, n (%)									
Syrinx	4	40%	20	57%		13	52%	11	55%
Ćyst	3	30%	8	23%		7	28%	4	20%
Fatty Filum	3	30%	10	29%		7	28%	6	30%
Chiari Malformation	9	90%	29	83%		22		16	80%
Shunt	8			91%			84%	19	95%
Mean number of shunt revisions	ŭ	0070	32	31/0			0 170		3370
+/- SD	0 00	1.60	2 26	3.07	:	2 08	3.07	1 20	2.63
Anatomical level of fusion, n (%)	0.90	1.00	2.20	3.07		2.00	3.07	1.00	2.03
Cervical	0	00/	1	3%		1	4%	0	00/
	0		1			-	.,.	_	0%
Thoracic	10	100%	35	100%	:	25	100%	20	100%
Lumbar	9	90%	31	89%		22	88%	18	90%
Sacra	6	60%	13	37%		10	40%	9	45%

Table 3. Patient Information

	Analysis	1 Group	Analysis 2 Group			
	Untethered	Not Untethered	Untethered	Never Untethered		
	(<6mo) + Fusion	(<6mo) + Fusion	(anytime) + Fusion	(anytime) + Fusion		
Baseline Disability, n (%)						
Motor Dysfunction (NEM Motor < 5)	10 100%	32 91%	23 92%	19 95%		
Sensory Deficit (NEM Sensation < 4)	9 90%	30 86%	22 88%	17 85%		
Bladder Incontinence (NEM Bladder < 5)	10 100%	35 100%	25 100%	20 100%		
Bowel Dysfunction (NEM Anus < 3)	4 40%	20 57%	13 52%	11 55%		
Ambulatory Impairment (Hoffer <5)	9 90%	30 86%	20 80%	19 95%		

	Untethered (<6mo) + Fusion					Not U				
	Pre-Op	Post-Op	Mean Change	+/- SD		Pre-Op	Post-Op	Mean Change	+/- SD	P-Value
NEM Score	7.80	8.00	0.20	1.32		8.31	8.37	0.06	1.00	0.7122
Motor	1.40	1.30	-0.10	0.88		1.49	1.51	0.03	0.17	0.4066
Sensory	2.70	2.80	0.10	0.99		2.83	2.80	-0.03	0.92	0.7043
Bladder	1.40	1.60	0.20	0.63		1.91	1.97	0.06	0.34	0.3450
Bowe	2.30	2.30	0.00	0.00		2.09	2.09	0.00	0.34	1.0000
Hoffer Score	2.20	2.00	-0.20	0.79		1.91	1.97	0.06	0.34	0.1341

Table 5. Analysis 1 Mean Functional Outcome Score Change Post-Fusion

	Untethered (anytime) + Fusion					Never U				
	Pre-Op	Post-Op	Mean Change	+/- SD		Pre-Op	Post-Op	Mean Change	+/- SD	P-Value
NEM Score	8.32	8.56	0.24	0.97		8.05	7.95	-0.10	1.17	0.2912
Motor	1.60	1.60	0.00	0.58		1.30	1.30	0.00	0.00	1.0000
Sensory	2.76	2.92	0.16	0.80		2.85	2.65	-0.20	1.06	0.2000
Bladder	1.76	1.84	0.08	0.40		1.85	1.95	0.10	0.45	0.8751
Bowel	2.20	2.20	0.00	0.08		2.05	2.05	0.00	0.00	1.0000
Hoffer Score	2.28	2.28	0.00	0.65		1.60	1.60	0.00	0.00	1.0000

Table 6. Analysis 2 Mean Functional Outcome Score Change Post-Fusion

Last Revised: 1/22/16 Page 23 of 24

Figures:

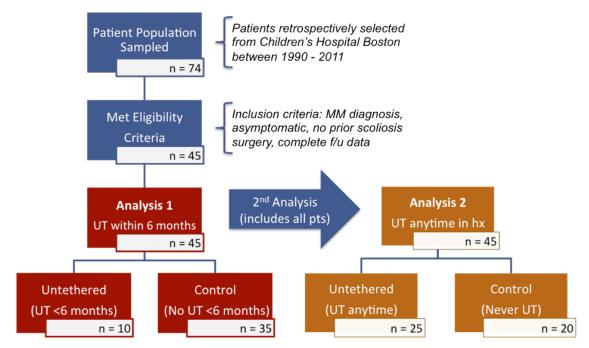


Figure 1. Study Design

Last Revised: 1/22/16 Page 24 of 24