

A patient with myelomeningocele: is untethering necessary prior to scoliosis correction?

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Object. Tethering of the spinal cord is thought to increase the chance of neurological injury when scoliosis correction is undertaken. All patients with myelomeningocele (MM) are radiographically tethered, and untethering procedures carry significant morbidity risks including worsening neurological function and wound complications. No guidelines exist as regards untethering in patients with MM prior to scoliosis correction surgery. The authors' aim in this study was to evaluate their experience in patients with MM who were not untethered before scoliosis correction.

Methods. Seventeen patients with MM were retrospectively identified and 1) had no evidence of a clinically symptomatic tethered cord, 2) had undergone spinal fusion for scoliosis correction, and 3) had not been untethered for at least 1 year prior to surgery. The minimum follow-up after fusion was 2 years. Charts and radiographs were reviewed for neurological or shunt complications in the perioperative period.

Results. The average age of the patients was 12.4 years, and the following neurological levels were affected: T-12 and above, 7 patients; L-1/L-2, 6 patients; L-3, 2 patients; and L-4, 2 patients. All were radiographically tethered as confirmed on MR imaging. Fourteen of the patients (82%) had a ventriculoperitoneal shunt. The mean Cobb angle was corrected from 82° to 35°, for a 57% correction. All patients underwent neuromonitoring of their upper extremities, and some underwent lower extremity monitoring as well. Postoperatively, no patient experienced a new cranial nerve palsy, shunt malfunction, change in urological function, or upper extremity weakness/sensory loss. One patient had transient lower extremity weakness, which returned to baseline within 1 month of surgery.

Conclusions. The study results suggested that spinal cord untethering may be unnecessary in patients with MM who are undergoing scoliosis corrective surgery and do not present with clinical symptoms of a tethered cord, even though tethering is radiographically demonstrated. (DOI: 10.3171/2010.3.FOCUS1072)

KEY WORDS • myelomeningocele • neurological injury • tethered cord • posterior spine fusion • scoliosis

SCOLIOSIS often develops in patients with myelomeningocele.^{3,9,16} Risk factors for the development of scoliosis in these patients include motor level, ambulatory status, and last intact laminar arch, with scoliosis developing in almost 100% of patients with a thoracic motor level.^{9,16} In addition, virtually all patients with MM radiographically demonstrate a tethered spinal cord,^{6,13} although only 10–30% exhibit symptoms.^{1,2,15} This occurs due to a difficulty in closing the dura mater around the neural placode to keep it bathed in CSF and prevent adherence to the dura.

A tethered spinal cord can predispose patients to neurological injury when they undergo surgical correction of scoliosis.^{5,12,17} Theoretically, this occurs due to excessive traction on a tethered spinal cord. Symptoms can include those attributed to shunt malfunction, cranial nerve palsies, upper extremity weakness, and worsening lower extremity function. However, untethering the spinal cord in patients with MM also carries some risk of morbidity

including worsening neurological function and wound problems.^{2,6} No reports in the literature address whether untethering is necessary in an asymptomatic patient with MM who presents for scoliosis surgery. Practice patterns vary from institution to institution with some advocating untethering of all patients and others never untethering prior to scoliosis correction. In the present report, we describe our experience with patients with MM who did not undergo untethering prior to their scoliosis surgery.

Methods

After obtaining institutional review board approval we performed a retrospective review of all patients with MM who had undergone surgery for scoliosis correction at our institution. None of the patients had been untethered for 1 year prior to their surgical procedure. Patient charts and radiographs were reviewed for the major coronal Cobb angle, sagittal kyphosis and lordosis, sagittal and coronal balance, and presence of a shunt. Intraoperative and postoperative complications were noted with

Abbreviation used in this paper: MM = myelomeningocele.

TABLE 1: Summary of patient characteristics

total no. patients	17
M	7
F	10
mean age at surgery in yrs (range)	12.4 (10–17)
mean length of follow-up in yrs (range)	3.3 (2–8)
patients w/ shunt (%)	14 (82)

particular attention devoted to shunt issues and neurological function including the brainstem, a change in urological function, and upper and/or lower extremity function.

Results

Patient Demographics

Seventeen patients—10 females and 7 males—with an MM who underwent surgery for scoliosis were identified. None of these patients presented with symptoms suggesting symptomatic tethered cord, and none had been tethered in the year preceding their scoliosis surgery. The average age was 12.4 years (range 10–17 years). The affected motor levels were thoracic (7 patients), L-1 or L-2 (6 patients), L-3 (2 patients), and L-4 (2 patients; Table 1). The major curve was lumbar in 6 patients (apex L-1/L-2 disc to L-4), thoracolumbar in 5 patients (apex T-12 to L-1), and thoracic in 6 patients (apex T-2 to T-11/T-12 disc). Fourteen patients underwent a T-2 to pelvis fusion. The remaining 3 patients underwent fusion of the following levels: T10–L4 (2 patients) and T11–L4 (1 patient).

Scoliosis Correction

The preoperative Cobb angle averaged 82° (range 56°–120°), and it measured 35° (range 20°–55°) at the last follow-up, for a 57% correction (Fig. 1). Maximal kyphosis averaged 50° (range 20°–110°) preoperatively, and improved to 33° (range 12°–66°) postoperatively (Table 2).

Neurological Complications

No patient experienced a shunt-related issue postoperatively. In addition, no new cranial nerve palsies, change in urological function, or upper extremity neurological deterioration occurred. One patient, whose MM motor level was at L-4, had moderate right quadriceps muscle weakness postoperatively, which improved to baseline function over 1 month. The etiology of the weakness was unclear as a postoperative CT was unremarkable. Although neurological complications were sparse, wound problems occurred in 4 (23%) of 17 patients, a rate similar to others reported in the literature.^{8,14}

Discussion

Our results suggest that untethering the spinal cord in patients with MM who do not present with symptoms of a tethered cord may be unnecessary before surgical correction of their scoliosis. In our series of 17 patients, significant correction of scoliosis was achieved, with only

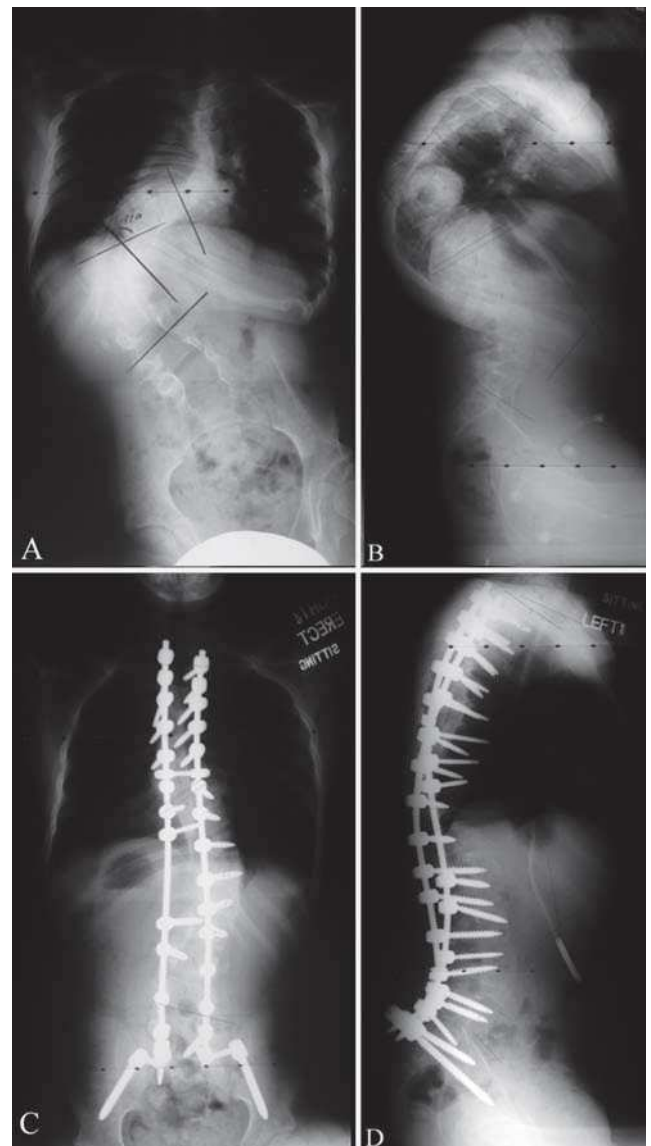


FIG. 1. Preoperative posteroanterior (A) and lateral (B) full spine radiographs obtained in a 15-year-old boy with an L-2 level myelomeningocele, showing a > 100° scoliosis curvature. The boy underwent a T-2 to pelvis posterior spine fusion without a prior untethering procedure. Postoperative posteroanterior (C) and lateral (D) radiographs showing good correction.

1 patient sustaining a temporary lower extremity deficit. No shunt problems, upper extremity weakness, or brainstem dysfunction was observed in the perioperative period. The main limitations of our study are its retrospective nature and relatively small sample size.

Virtually all patients with MM demonstrate radiographic tethering.^{6,13} Theoretically, the spinal column lengthening that occurs with scoliosis correction can result in neurological deterioration. Little has been published on whether patients should undergo untethering prior to correction of their scoliosis. Al-Holou et al.¹ commented on prophylactic untethering in patients with MM undergoing scoliosis surgery. Similarly, in a subsection of their paper, Bowman et al.² reported on 10 children with severe sco-

Patients with myelomeningocele and the need for untethering

TABLE 2: Deformity correction*

Deformity	Cobb Angle (°)	
	Preop	Most Recent Follow-Up
major coronal curve	82 (56–120)	35 (20–55)
kyphosis	50 (20–110)	33 (12–66)

* Values are presented as means with ranges in parentheses.

liosis who “underwent untethering immediately prior to spinal fusion to prevent neurological decline secondary to possible spinal column lengthening.” At our institution we do not untether patients with MM who are undergoing scoliosis correction surgery, as long as they do not present with symptoms suggestive of a tethered cord.

Symptoms suggestive of a tethered cord in these patients include pain, weakness, gait abnormality, lower extremity hip and foot deformity, urological changes, and rapidly progressive scoliosis.^{2,6} After untethering, improvements are commonly seen in pain and often in weakness, gait, and urological changes.^{1,2} Results of untethering in patients with a rapidly progressive scoliosis appear to be best when the curve is < 40°–45° and in patients whose motor level is below T-12.^{2,7,10,11} Pierz et al.¹⁰ reported on 21 patients with MM and rapidly progressive scoliosis who underwent untethering. None of the patients with curves > 40° or with an affected thoracic level demonstrated any benefit. Reigel et al.¹¹ examined 216 patients with MM who were untethered and found none of those with thoracic level MM stabilized.

Although none of our patients experienced shunt malfunction in the early postoperative period, this complication is potentially devastating in this population. In their series of 77 patients with MM undergoing scoliosis surgery, Geiger et al.⁴ described 4 patients who presented with acute postoperative shunt failure, with 1 death. The mechanism for acute shunt failure may include blood products entering the CSF and blocking the shunt, or shunt breakage from the change in alignment of the torso after scoliosis correction. Heavy narcotics for postoperative management should be used with caution to ensure adequate neurological assessment.

Conclusions

Virtually all patients with MM have a tethered spinal cord. Our results suggest that in a patient with MM who is asymptomatic, one may not need to untether the spinal cord prior to scoliosis surgery. These findings should be considered preliminary as the number of patients in our study was relatively small.

Disclosure

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: Samdani, Betz. Acquisition of data: Samdani, Sagoo, Cahill, Clements. Analysis and interpretation of data: Samdani, Fine, Sagoo, Shah. Drafting the article: all authors. Critically revising the article: Samdani, Fine,

Shah. Reviewed final version of the manuscript and approved it for submission: Samdani, Fine, Sagoo, Cahill, Clements, Betz. Study supervision: Betz.

References

1. Al-Holou WN, Muraszko KM, Garton HJ, Buchman SR, Mather CO: The outcome of tethered cord release in secondary and multiple repeat tethered cord syndrome. Clinical article. *J Neurosurg Pediatr* 4:28–36, 2009
2. 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
3. Dias MS: Neurosurgical causes of scoliosis in patients with myelomeningocele: an evidence-based literature review. *J Neurosurg* 103 (1 Suppl):24–35, 2005
4. Geiger F, Parsch D, Carstens C: Complications of scoliosis surgery in children with myelomeningocele. *Eur Spine J* 8: 22–26, 1999
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* 32:2880–2884, 2007
6. Hudgins RJ, Gilreath CL: Tethered spinal cord following repair of myelomeningocele. *Neurosurg Focus* 16(2):E7, 2004
7. McGirt MJ, Mehta V, Garces-Ambrossi G, Gottfried O, Solakoglu C, Gokaslan ZL, et al: Pediatric tethered cord syndrome: response of scoliosis to untethering procedures. Clinical article. *J Neurosurg Pediatr* 4:270–274, 2009
8. Mohamad F, Parent S, Pawelek J, Marks M, Bastrom T, Faro F, et al: Perioperative complications after surgical correction in neuromuscular scoliosis. *J Pediatr Orthop* 27:392–397, 2007
9. Muller EB, Nordwall A, Oden A: Progression of scoliosis in children with myelomeningocele. *Spine* 19:147–150, 1994
10. 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
11. Reigel DH, 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
12. Samdani AF, Asghar J, Pahys J, D’Andrea L, Betz RR: Concurrent spinal cord untethering and scoliosis correction: case report. *Spine* 32:E832–836, 2007
13. 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
14. Sponseller PD, LaPorte DM, Hungerford MW, Eck K, Bridwell KH, Lenke LG: Deep wound infections after neuromuscular scoliosis surgery: a multicenter study of risk factors and treatment outcomes. *Spine* 25:2461–2466, 2000
15. 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
16. Trivedi J, Thomson JD, Slakey JB, Banta JV, Jones PW: Clinical and radiographic predictors of scoliosis in patients with myelomeningocele. *J Bone Joint Surg Am* 84-A:1389–1394, 2002
17. Winter RB, Haven JJ, Moe JH, Lagaard SM: Diastematomyelia and congenital spine deformities. *J Bone Joint Surg Am* 56:27–39, 1974

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