Response of Scoliosis in Children with Myelomeningocele to Surgical Release of Tethered Spinal Cord

Haluk Altiok,^{1,2} Anne Riordan, BA,¹ Adam Graf, MS,¹ Joe Krzak, PT,¹
and Sahar Hassani, MS¹

¹Shriners Hospitals for Children Chicago ; ²Loyola University, Department of Orthopedics
and Rehabilitation, Chicago, Illinois

Objective: To examine the effect of surgical tethered cord release (TCR) on scoliosis in children with myelomeningocele. **Methods:** A retrospective review of 65 pediatric patients with myelomeningocele and TCR. The final sample consisted of 20 patients with scoliosis who were managed conservatively after TCR. **Results:** Average age at TCR was 6.2 years with average follow-up of 3.8 years. Scoliosis of 1 (5%) patient improved, 7 (35%) were stable, and 12 (60%) worsened ($\geq 10^\circ$). Fifty percent of patients ultimately required definitive spinal surgery. TCR release delayed definitive spine surgery for an average of 3.2 years. Sixty-four percent of patients with curves less than or equal to 45° had progression of their curves compared to 50% with curves greater than 45° . For patients with curves less than or equal to 45° , curves progressed in 80% of those younger than 10 years as compared to 25% of those older than 10 years. For patients with curves less than or equal to 45° , 43% required definitive spine surgery as opposed to 83% with curves greater than 45° . Level of neurological involvement (ie, lumbar versus thoracic) and age at untethering emerged as factors influencing the effects of TCR for patients with curves less than or equal to 45° . Lumbar curves had more favorable results. **Conclusion:** Pediatric patients with myelomeningocele and scoliosis should be closely assessed and monitored. A selective approach for youth with lumbosacral level myelomeningocele and progressive curves less than or equal to 45° may result in scoliosis stabilization and avoidance of definitive surgery. **Key words:** myelomeningocele, scoliosis, tethered cord

Scoliosis is a frequent orthopedic manifestation in patients with myelomeningocele with an overall incidence of 50% to 80%.¹⁻⁷ It is characterized by early onset and rapid progression.^{1,4-9} The pathogenesis of scoliosis in myelomeningocele is considered to be multifactorial, involving neurological level, congenital vertebral anomalies, hydrocephalus and shunt malfunction, Chiari malformation, tethered cord, and hydromyelia.^{2,9-14} The level of last intact laminar arch/spinal dysraphism as well as the clinical level of paralysis are highly correlated with the occurrence of scoliosis,^{2,5,7,8} with almost all patients with thoracic level lesions (87% to 100%) having scoliosis.

Tethered cord syndrome is characterized by symptoms and signs that result from excessive tension on the spinal cord,¹⁵ including back and leg pain, change in bladder tone, change in motor or sensory levels, spasticity, deformities of lower extremities, new onset or progression of

scoliosis, and gait deterioration.¹⁵⁻²⁰ Symptomatic tethering of the spinal cord following primary myelomeningocele repair occurs in 2.8% to 27% of patients.^{9,16,18,19,21}

The association of tethered cord with scoliosis has long been assumed. In a study by McLone et al,¹⁷ 43 out of 91 patients with myelomeningocele who had surgical release of their symptomatic tethered cord had scoliosis as one of the signs of deterioration. Moreover, tethered cord alone (without any other central nervous system abnormalities) was the most common cause of scoliosis for 30 patients. Sixty-seven percent of these patients demonstrated stabilization or improvement of their scoliosis following release of their tethered cord at the 2- to 7-year follow-up period.

Tethered cord release (TCR) may decrease the progression of the spine deformity without additional procedures. However, many factors determine whether untethering will impact curve progression, including neurological level,

Corresponding author: Haluk Altiok, Shriners Hospitals for Children Chicago, 2211 N. Oak Park Avenue, Chicago, IL 60707; phone: 773-385-5457; e-mail: haltiok@shriners.org

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age at the time of untethering, curve magnitude, and possible retethering.^{16,17,19, 22-24} The purpose of our study was to study the effect of tethered cord release on scoliosis in children with myelomeningocele and identify the factors associated with favorable outcome.

Methods

This is an institutional review board–approved retrospective review of 65 youth with myelomeningocele who underwent tethered cord release between 1980 and 2010 in a single pediatric orthopaedic hospital. Patients who underwent MRI and/or CT myelography to rule out other possible causes of clinical deterioration (eg, shunt malfunction) were included in this sample. Data that were collected included patient demographics, neurological level, and clinical signs and symptoms leading to the diagnosis of the tethered cord syndrome. For patients with scoliosis and/or kyphosis and after TCR, information regarding Cobb angles was obtained at the time of tethered cord release and at final follow-up and/or time of definitive spine surgery. Age at time of the definitive spine surgery and surgical complications was also recorded. Scoliosis in this study is defined as a curve greater than 20°. The curve was classified as improved if it decreased by 10° or more and progressive if it increased by 10° or more.

Results

Of the 65 patients studied, 43 were diagnosed with myelomeningocele and 22 with lipomeningocele. Twelve patients had thoracic and upper lumbar level lesions, 42 had L3-4 level lesions, and 11 had sacral level lesions. The most common presenting symptoms of spinal cord tethering were scoliosis (50.7% of patients), weakness (38.4% of patients), gait change (36.9% of patients), foot deformities (36.9% of patients), and spasticity (32.3% of patients).

Thirty-three patients had progressive spine scoliosis and/or kyphosis as one of their presenting symptoms. Of these, 12 patients underwent TCR before their definitive corrective spine surgery as a staged procedure to prevent acute postoperative neurological deterioration with correction of the scoliosis, whereas 21 patients were managed

conservatively after the tethered cord was released. The average Cobb angle was $68.4^\circ \pm 21.8^\circ$ before the tethered cord was released and $44.5^\circ \pm 16.6^\circ$ after corrective spine surgery. Average age at the tethered cord release was 11.3 years, and average age at the time of definitive spine surgery was 11.6 years. Three patients had thoracic and 9 patients had lumbosacral level myelomeningocele. Two patients had complications: wound infection in one, and diminished bladder control after the untethered cord procedure in the other. In patients who underwent TCR to control the spine deformity, 12 (57%) were female. The majority of cases were myelomeningocele (86% vs 14% lipomeningocele) and scoliosis (95% vs 5% kyphosis).

Our final study sample consisted of the 20 pediatric patients with scoliosis (5 thoracolumbar, 14 L2-L4, and 1 L5 lesion level). The average age when untethering occurred was 6.2 years (range, 1.6-14.6 years). Fifteen patients had ventriculoperitoneal shunts and/or Chiari malformation: 11 had Chiari malformation and ventriculoperitoneal shunt, 3 had shunt but no Chiari, and 1 had Chiari alone. One patient had hemivertebrae. The average Cobb angle in this group was $41.6^\circ \pm 15.1^\circ$ before the tethered cord was released. The curves in 14 of the 20 patients were 45° or less at the time of TCR. Average follow-up was 3.8 years (range, 1-6.4 years). During the follow-up, 1 (5%) patient improved, 7 (35%) were stable, and 12 (60%) worsened. Ten out of 20 patients (50%) ultimately required definitive spinal surgery. TCR delayed definitive spine surgery for an average of 3.2 years from the time of untethering (range, 1-6.4 years) (**Figures 1** and **2**).

Although the number of patients in this sample was relatively small, more patients demonstrated progression of their curves if scoliosis was less than or equal to 45° (64%) compared to those with larger curves (50%). Younger age at the time of untethering appeared to be an important factor for this outcome. For the group of patients with curves 45° or less, curves progressed in 80% of those who were younger than 10 years of age (average age, 4.4 years) at the time of untethering as opposed to 25% of the patients who were older than 10 years (**Table 1**). For the group of patients with curves 45° or less, 6 out of 14 (43%) required definitive spine surgery as compared to 5 out of 6 patients (83%) with curves greater than 45°. Scoliosis progressed

Figure 1. (A) Eight-year-old male patient with 25° lumbar scoliosis and (B) 6 years after tethered cord release and lipoma resection.

Figure 2. Four-year-old male, with a thoraco-lumbar (42°/33°) scoliosis and spasticity. (A) Tethered cord release done at age 4 years with a (B) follow-up of 6 years before (C) the definitive surgery was done at age 10 years with a thoraco-lumbar (48°/65°) scoliosis.

Table 1. Curve progression related to the age and the severity of the curve at the time of tethered cord release

Age at release	Curve $\leq 45^\circ$ ($n = 14$)		Curve $>45^\circ$ ($n = 6$)	
	Stable or improved	Worsened	Stable or improved	Worsened
≤ 10 years	2 (20%)	8 (80%)	3 (50%)	3 (50%)
>10 years	3 (75%)	1 (25%)	0	0

in 3 patients who had curves 45° or less and who were thoracic level. In contrast, in the lumbar level group, 6 of 11 patients experienced curve progression and 5 stabilized, including 1 patient with hemivertebrae.

Discussion

Not every pediatric patient with myelomeningocele and scoliosis requires spine surgery. Potential indications for definitive spinal fusion include larger curves with poor sitting balance and need of upper extremities for balance, significant pelvic obliquity, associated pressure sores, and poor pulmonary function.^{6,13} It is important to identify factors that contribute to the scoliosis. Addressing these causative factors in a timely manner may help to control the scoliosis to the extent that there is no need for definitive surgical stabilization or it can be delayed, allowing for additional years of growth.

Tethered cord syndrome has long been considered as a causative factor in the development of scoliosis in myelodysplastic patients.^{16,17,19,22-24} McLone et al¹⁷ identified tethered cord as a causative factor for scoliosis in more than 70% of patients with lumbar level myelomeningocele in the absence of vertebral anomalies, shunt malfunction, or hydromyelia. Curves greater than 50° seemed to have less benefit following TCR. Moreover, in Sarwark et al's¹⁹ study of 30 pediatric patients with L3 or lower level myelomeningocele in whom the only known etiology for neurologic and/or orthopedic decline was a tethered cord, 64% of the curves that were less than 45° at the time of release either improved or remained stable over a 3- to 10-year follow-up.

Furthermore, in a study conducted by Bowman and colleagues,¹⁶ including 114 patients who

required TCR, 46 patients (40%) underwent untethering secondary to progressive scoliosis. Ten out of 46 patients had untethering performed prior to spinal fusion to prevent neurological decline. Long-term follow-up (average, 7.2 years) of the remaining 36 patients revealed progression of the scoliosis in 66% of 36 patients, with 36% of the 36 patients ultimately requiring spinal fusion. All 12 patients (33% of 36 patients) with curves smaller than 45° remained stable or had improved spinal curvatures after a TCR. Seven of 13 patients whose curves progressed and went on for a spinal fusion had thoracic level lesions. The authors concluded that untethering of the spinal cord does not appear to prevent the need for eventual spinal fusion in the majority of patients.

In the Pierz et al²³ study with 21 patients, untethering of the spinal cord failed to stabilize or improve scoliosis in patients with thoracic level function and curves greater than 40° . Regardless of the patients' preoperative functional status, untethering was associated with improvement of scoliosis in only 14% of cases while 29% remained stable. The authors concluded that the causes of the scoliosis alone must be analyzed critically before recommending untethering.

Reigel et al²⁴ had similar findings: Progression of scoliosis declined or stabilized in patients with lumbar and sacral level lesions, however, detethering did not stop progression of scoliosis in thoracic level group. They also concluded that TCR for recurring tethering is associated with further control of the progression of scoliosis. Dias²⁵ evaluated the level of evidence to support the causal relationship between scoliosis and spinal cord abnormalities of tethered cord, Chiari, and syringomyelia. The author concluded that there were insufficient data available to draw firm conclusions about the effects of tethering on the development or progression of scoliosis in the child with myelomeningocele, although there might be some potential benefit to untethering with high lumbar level myelomeningoceles or for those with curves less than 45° .

In our study, we found that age was a significant factor in regard to the effect of tethered cord release on scoliosis in children with myelomeningocele. Our study did not show any protective value of the curve magnitude at the time of untethering against

potential progression. For the group of patients with scoliosis less than or equal to 45°, curves progressed in 80% of the patients younger than 10 years at the time of untethering as opposed to 25% of patients who were older than 10 years. This may be explained by the impact of skeletal maturity on curve progression. However, regardless of age, only 43% of the patients with curves less than 45° required definitive spine surgery despite the progression of their curves as opposed to 83% of the patients with curves greater than 45°. Level of neurological involvement, lumbar versus thoracic, also seems to be a factor in regard to the effects of tethered cord release for those patients with curves equal or less than 45°. Curves of all 3 patients with thoracic level progressed whereas only 6 of 11 of those with lumbar level progressed.

Limitations of our study include the small sample size and retrospective nature. These preclude us from drawing firm conclusions about the impact of the untethering procedure on scoliosis in

myelomeningocele patients. Even though there are potential complications with untethering and symptomatic retethering may occur,^{16,19,23} the natural history of symptomatic tethered cord syndrome is consisted with progressive neurological decline in the absence of surgical untethering.^{15,18} Given the significant potential consequences of inaction, we believe that each patient with scoliosis should be individually assessed, taking into account the natural history of scoliosis in patients with myelomeningocele as well as the nature of progression of the curve, neurological level, age of the patient, and associated other signs and symptoms of tethered cord. Selective approach for young, lumbosacral level patients with progressive curves less than 45° may yield results with scoliosis stabilization and avoidance of definitive surgery. Even though scoliosis may progress after untethering, additional years of growth before potential definitive surgical stabilization could be of great benefit.

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