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Tethered Cord Syndrome in Low Motor Level Children with Myelomeningocele

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Abstract
The clinical presentation of tethered spinal cord and the results of tethered cord release were examined in a group of 30 low motor level (L3 and below) children with a history of myelomeningocele without concomitant CNS complications. Changes in orthopedic and/or neurologic status formed the basis of consideration for tethered cord release. Clinically, these patients presented with a new onset or recently progressing scoliosis, spasticity with or without contractures, decrease in motor function and low back pain at the site of closure. One or more of these findings was present in all cases and led to the suspicion of tethered spinal cord. The diagnosis of tethered cord was confirmed in all cases by MRI or CT myeolography. In order to isolate tethering as the etiology for the patients' clinical deterioration, patients with concomitant CNS complications, e.g. shunt dysfunction or hydromyelia were excluded from the study. Twenty-nine such patients, of an initial 59, who would have otherwise been considered, were excluded on the basis of this criteria of concomitant CNS complications. The results of release 1 year after the procedure were as follows: regarding scoliosis, in 75% of cases the curve either remained stable or decreased by more than 10°, with 25% experiencing curve progression of >10°. The most recent follow-up in this group revealed that 11.8% experienced a decrease in curvature of >10°; 47.1% remained stable, and 41.2% ultimately progressed 10°. In the group with spasticity, 43.8% improved; 56.3% remained stable, and none worsened. Most (78.6%) of the children who had experienced a decline in motor function improved postoperatively, and all those with back pain experienced complete resolution. In conclusion, tethered cord release in symptomatic low lumbar and sacral level children with myelomeningocele appears to be of benefit, especially with respect to stabilization of scoliosis in selected patients, back pain at the site of closure, and prior decline in motor function. Results in the cases with spasticity were more equivocal.

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Introduction

Symptomatic retethering of the spinal cord following primary myelomeningocele repair occurs in 11–27% of cases [1–4]. Though the conus medullaris is frequently found to be low lying in children with a myelomeningocele in general [5, 6], and may even appear adherent to the dorsal dura in the majority of cases [7], the significance of this finding depends on its association with the clinical signs and symptoms of the tethered cord syndrome [5, 7–9]. These signs and symptoms include back and leg pain, change in bladder tone, incontinence on intermittent catheterization, change in motor or sensory level in the lower extremities, spasticity of the lower extremities, and rapid progression of scoliosis [7, 10].

The pathophysiology of tethered cord has been examined. One study [11] revealed metabolic impairment which correlated with electrophysiologic spinal cord dysfunction in the feline spinal cord under traction. Another study revealed that traction on the cord increases the vulnerability to extrinsic compression [12].

Symptomatic retethering of the spinal cord in children with myelomeningocele is most frequently due to scarring, or adhesive arachnoiditis involving the neural placode or spinal cord with resultant adherence to the overlying dura or skin [3, 7, 13–15]. There is still controversy regarding the importance of the initial closure technique, with some neurosurgeons advocating pia-arachnoid repair while others have advocated leaving the neural placode open [16]. Another viewpoint is that the anatomic arrangement of the intraspinal compartment at infancy, which is described as a shallow dish, makes retethering very difficult to avoid [7].

This study retrospectively examines the clinical outcomes of a select group of 30 low motor level, pediatric myelomeningocele patients who underwent tethered cord release after developing signs of symptomatic tethered cord. The purpose of the study was to determine the effect of tethered cord release in this group of myelomeningocele patients in whom the only known etiology for their neurologic and/or orthopedic decline was a tethered spinal cord.

Materials and Methods

A retrospective review of operative records, charts, radiographs, and the PDMS data base, which has been used in the myelomeningocele clinic since 1982, was utilized to collect the data.

The entire group of children who had undergone tethered cord release, performed by a single neurosurgeon (D.M.), at Children's Memorial Hospital between 1981 and 1989, was initially selected from 77 patients. The focus was then restricted to the patients who had been functioning as L3 motor level or better (community ambulation). This decreased the group size to 59 patients. Twelve cases were then excluded because of moderate to severe hydromyelia, 3 because of shunt malfunction requiring shunt revision at the time of tethered cord release, 6 because of CNS complications other than hydromyelia (infection, cerebral palsy), 3 because posterior rhizotomy was performed at the time of release, 3 children were eliminated because of inaccurate records of their spinal cord imaging studies, and 2 for whom the clinical data concerning their course prior to the time of tethered cord release were incomplete.

The selected group of 30 patients consisted of 17 females and 13 males. The average age at the time of tethered cord release was 7 years. The range of ages was 1 year, 1 month to 14 years, 5 months.

Sixteen (53.3%) of the patients were at the low lumbar motor level (L3–L5) and 14 (46.7%) were at the sacral motor level.

Four of these patients underwent two separate tethered cord release operations. In 1 patient the tethered cord releases were performed within 1 year and only the results of the later release are included in the study. In the other 3 cases the releases were performed from 1 year and 2 months to 2 years and 4 months apart. In those instances, data from both releases are included in the study for the purpose of reporting their clinical presentations and 1-year follow-up results regarding scoliosis. Therefore, 33 cases were ultimately analyzed in this study for their clinical presentations. In those cases where scoliosis was present, the 1-year results include all cases with scoliosis, while the long-term results reflect individual patient data. Results in the cases with spasticity, decrease in motor function, and back pain at the site of myelomeningocele closure are not broken down into 1-year and long-term follow-up groups, but instead reflect the clinical response within 1 year of tethered cord release. The areas of clinical evaluation in this study included: (1) new or rapidly progressive scoliosis in patients with no primary vertebral anomalies; (2) deterioration of motor function; (3) spasticity (frequently seen in the hamstring muscles, ankle dorsiflexors/evertors), and (4) back pain at the site of closure. We did not look at changes in bladder function. All 33 cases presented with one or more of these signs/symptoms.

The operative technique for release has been described previously [17, 18]. A CO_2 laser was used for dissection in all patients. Information regarding the myelomeningocele initial closure technique was not available for all patients.

Results

Clinical Presentation

The 33 cases of tethered cord presented as follows: Twenty (60.6%) presented with new onset or recently progressing scoliosis in the absence of primary vertebral abnormality (congenital scoliosis). Seventeen of these were curves of 45° or less, and 3 were curves of >45°. Sixteen (48.5%) cases presented with lower extremity spasticity. Fourteen (42.4%) demonstrated a decrease in muscle/motor function detected by a manual muscle test, and 3 (9.1%) presented with low back pain at the site of closure of recent onset. Fifteen of the cases presented with more

than one of the above (table 1). All cases were demonstrations of secondary tethering or symptomatic retethering as no known predisposing risk for tethering was recognized except prior myelomeningocele repair in any patient.

Scoliosis Postoperative Results

Twenty presentations of tethered cord in 17 patients were for scoliosis. A postoperative decrease in the curvature of >10° was considered an improvement; an increase of >10° was considered continued progression, and everything in between was considered stable. First-year results revealed that 3 (15%) of the 20 cases have shown improvement, 12 (60%) were stabilized following release, and in 5 (25%) cases the curvature continued to progress. Therefore, in 75% of cases, the curvature either improved by >10° or was stable in the first year following tethered cord release (table 2). The 17 cases (14 patients) where the scoliotic curve was 45° or less preoperatively consisted of 2 (11.8%) who showed improvement in the first year, 12 (70.6%) where the curve remained stable, and 3 (17.6%) who worsened. 82.4% either improved or remained stable in the first year. Long-term (3- to 10-year) follow-up in this group of 14 patients revealed improvement in 1 (7.1%), stabilization in 8 (57.1%), and continued progression in 5 (35.7%). So, 64.2% ultimately improved or remained stable (table 3). No difference in response to cord untethering was noted between sacral and low lumbar level patients.

There were 3 patients in whom the curvature was >45° prior to the release. One of these improved and 2 continued to progress both at 1 year and also at the most recent follow-up (table 3).

Spasticity Postoperative Results

It is known that objective measurement of spasticity is not yet available, therefore we utilized clinical evaluation by physicians and physical therapists for the assessment of spasticity in this study. Sixteen of the thirty-three cases had spasticity as part of their presentation. The possible outcomes were improved, no improvement, or worsened, and were based upon the physical examinations of the patients in the myelomeningocele clinic on serial examinations.

Seven (43.8%) of the patients were found to have some improvement following tethered cord release, 9 (56.3%) showed no clear improvement, and 0 (0.0%) worsened.

Table 1. Clinical presentation of tethered cord (33 cases total)

	Number of cases	% of total
New or progressive scoliosis	20	60.6
Spasticity	16	48.5
Decline in motor function	14	42.4
Back pain at closure site	3	9.1

Table 2. Postoperative results: Scoliosis

	Improved	Stabilized	Progressed
1st year (20 cases)	3 (15%)	12 (60%)	5 (25%)
Long-term (17 cases)	2 (11.8%)	8 (47.1%)	7 (41.2%)

Table 3. Scoliosis results by angle at presentation

	Improved	Stabilized	Progressed
>45°			
1st year (3 cases)	1 (33.3%)	0	2 (66.7%)
Long-term (3 cases)	1 (33.3%)	0	2 (66.7%)
≤45°			
1st year (17 cases)	2 (11.8%)	12 (70.6%)	3 (17.6%)
Long-term (14 cases)	1 (7.1%)	8 (57.1%)	5 (35.7%)

Motor Function Postoperative Results

Fourteen of the thirty-three cases had decreased muscle function or a change in motor level as part of their presentation preoperatively. Postoperative improvement was defined as improvement in motor level or increased strength at the same level. Eleven (78.6%) showed some improvement postoperatively, 3 (21.4%) showed no clear improvement, and 0 (0.0%) of the patients in this group deteriorated.

Back Pain Postoperative Results

All 3 patients who reported back pain prior to surgery experienced complete resolution of the pain.

Retethering

Four of thirty patients developed symptomatic retethering requiring additional surgical release. No difference was noted between these and other patients pre- and intraoperatively.

Discussion

Debate continues as to the significance of and the approach to symptomatic tethered spinal cord in children with a myelomeningocele [17].

Much of the debate arises from the fact that the conus is found to be low lying and/or adherent to the dorsal dura in most if not all children with myelomeningocele who underwent repair shortly after birth [5–7]. By contrast, only 11–27% of all myelomeningocele patients ultimately develop what is considered to be symptomatic retethering of the spinal cord with the clinical manifestations described earlier [1–3].

Additionally, some of the clinical manifestations of symptomatic tethered cord syndrome can often be explained by concurrent conditions in these patients. Spasticity of the lower extremities could be explained by hydromyelia [19, 20] for example. Scoliosis in these patients has even more possible explanations such as bony anomalies, including congenital hemivertebra leading to scoliosis. Other possible explanations include the anatomic or motor level of the lesion that may be asymmetric and lead to imbalance [21]. Even with these excluded, one must consider hydromyelia, or shunt malfunction with resultant hydrocephalus, as a possible explanation for the scoliosis [7, 17, 22–24]. Hydromyelia, and shunt malfunction with or without hydromyelia, occur concomitantly very commonly in these children and may make discerning whether the tethered cord is playing the greater clinical role difficult. In this study, all those patients who were found to have shunt malfunction at the time of release, or moderate to severe hydromyelia requiring treatment on MRI or delayed metrizamide CT were excluded from the study.

Similarly, the thoracic level patients and the high lumbar level patients were excluded since the incidence of developmental scoliosis occurs commonly and is dependent on the anatomic and neurologic level of the lesion. For example, nearly all thoracic level patients develop scoliosis by age 15 [25–27], 23–70% of low lumbar level patients [27–29] and <10% of sacral level patients [27, 28]. Therefore, it would be much harder to meaningfully state that scoliosis was due exclusively to tethered cord alone in those with higher level lesions if included in the study.

There is recent literature which supports the tethered cord as a causal factor in developmental scoliosis in myelomeningocele patients and which seems to indicate that tethered cord release is beneficial for many patients [10, 30–32] (fig. 1). McLone et al. [10] reviewed 30 myelomeningocele patients with developmental scoliosis and tethered cord, and found that of the 24 in whom the curvature was <45° at the time of release, all either stabilized or improved in the first year, and 63% ultimately stabilized or improved. They concluded that tethered cord contributes to developmental scoliosis, and that release of the tethered cord is of benefit in those cases where intervention occurred before the curve became >50° [10]. In this study we observed that in those cases where the scoliotic curve was <45° preoperatively, 82.4% improved or remained stable in the first year, and 64.2% ultimately stabilized or improved. These results are clearly very similar to those of earlier work at our institution. Of those that demonstrated continued progression, surgical stabilization and fusion were recommended.

Reigel [14] reviewed a large group of patients with tethered cord and subsequent release, nearly 90% of whom were myelomeningocele or meningocele patients. Of those with scoliosis, 71% stabilized and 29% continued to progress. Of those with new contracture, 52% improved, 47% remained the same, and 1% worsened. The results regarding weakness revealed that 76% improved, 19% remained the same, and 5% became worse subsequent to the tethered cord release. 87% of those with back pain improved, 10% remained the same, and 3% worsened. Again, the results support the release of the cord in symptomatic patients and are similar to the results of this study [14].

Numerous studies involving smaller patient groups indicate that there is some benefit to tethered cord release in the symptomatic patient [2, 3, 9, 21, 30, 33, 34].

The findings in this study are also in agreement with previous studies which cite MRI to be a reliable way to confirm the site of cord tethering in the symptomatic myelomeningocele child [2, 13, 36, 37]. All symptomatic patients who were found to have a cord or conus that abutted the dorsal dura on MRI demonstrated a tethered cord at surgery.

Symptomatic retethering after tethered cord release in 13% of our patients is concerning. However, this number is similar to pathologic adhesion of the spinal cord after primary closure [1–4] for myelomeningocele. In a similar study of adults, 25% of patients had evidence of retethering from 1 to 9 years after tethered cord release [31]. It would be reasonable to state that there is an inherent risk of adhesion/scarring following spinal surgery. Furthermore patients undergoing surgical untethering seem to be at risk of recurrence at a similar rate as after initial closure.

The relative duration of signs and symptoms related to outcomes of the procedure were not studied here. How-

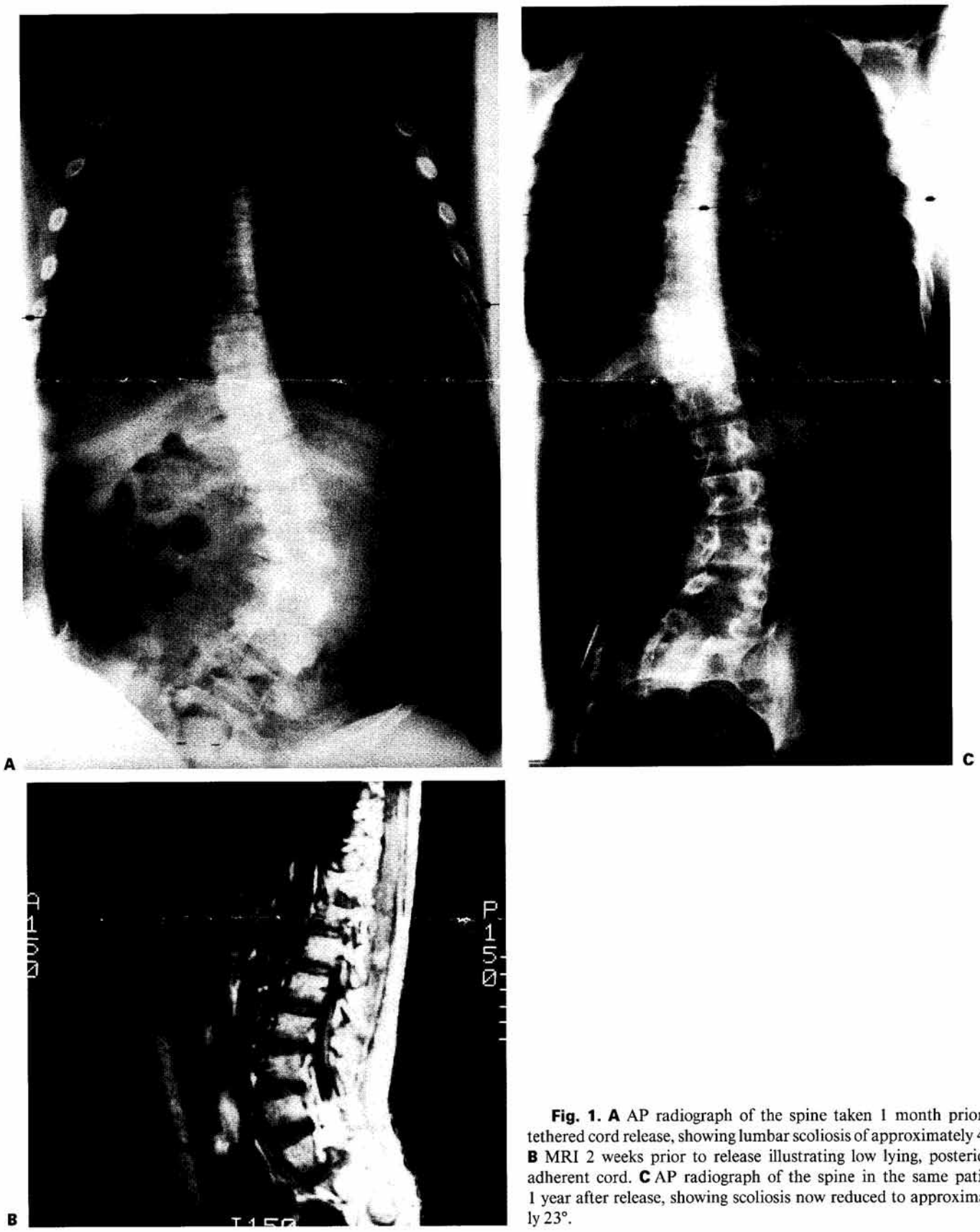

Fig. 1. A AP radiograph of the spine taken 1 month prior to tethered cord release, showing lumbar scoliosis of approximately 48°. **B** MRI 2 weeks prior to release illustrating low lying, posteriorly adherent cord. **C** AP radiograph of the spine in the same patient 1 year after release, showing scoliosis now reduced to approximately 23°.

ever, this remains an important question worthy of additional study. It is reasonable to infer, based on the suspected pathophysiology, that earlier intervention will lead to a better result such as reversal of scoliosis, resolution of spasticity and return of motor strength.

Conclusions

In conclusion, this study supports the release of a tethered spinal cord in low motor level children with myelomeningocele who are showing sings/symptoms of tethered spinal cord and who have MRI findings consistent with tethered spinal cord.

Clear benefit was shown with respect to scoliosis, with 75% of cases demonstrating stabilization or improvement in the first year. In those where the curve was 45° or less preoperatively, 82.4% showed stabilization or improvement in the first year. Tethered cord release is clearly beneficial to the child in a variety of ways, including that of potentially preventing sitting imbalance, skin instability and respiratory complications that can occur with severe levels of scoliosis [38]. Some of these curves will ultimately progress, but an additional year or more of growth before surgical stabilization by way of fusion and instrumentation is of great benefit, as the average age at the time of release was 7 years in this study, which is well below the optimum age for spinal fusion in these patients [19].

All patients who had complained of back pain prior to the release reported improvement or elimination of pain, and most (78.6%) of those had experienced a decline in motor function. None of the patients in this study experienced worsening of neurological symptoms following tethered cord release, confirming other studies [14, 39].

Therefore, the data support that release is indicated in those with symptomatic secondary retethering of the cord [8].

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