Occult tethered cord syndrome: the case for surgery

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Object. Controversy exists regarding proper indications for surgical lysis of the terminal filum in children with voiding dysfunction and tethered spinal cord. Recently, surgery has been offered to children who have a normally positioned conus medullaris and no terminal filum abnormality visible on 1.5-tesla magnetic resonance images (referred to as minimal or occult tethered cord syndrome [TCS]). The author evaluates existing clinical and scientific evidence relevant to this controversy.

Methods. Five retrospective, observational, noncontrolled studies of surgical terminal filum lysis for occult TCS in children were identified. Two further studies in which the authors reported surgical results in children with a normal-level conus medullaris were also identified.

Conclusions. These studies document encouraging clinical outcomes following surgery. Clinicopathological evidence suggests that occult TCS may result from radiographically occult structural abnormalities of the terminal filum. Although a preponderance of Class III clinical evidence supports the use of surgical filum lysis to treat occult TCS, no Class I or II evidence exists. Clinical practice varies; therefore, performance of a prospective randomized clinical trial of surgical terminal filum lysis for the treatment of occult TCS is advocated.

Key Words • terminal filum • tethered cord syndrome • pediatric neurosurgery

Tethered cord syndrome can include voiding dysfunction, lower-extremity weakness, sensory abnormalities, pain, and gait disturbance. It is associated both with major spinal dysraphism, such as lipomyelomeningocele, and with more subtle structural abnormalities of the terminal filum, including thickening, fat infiltration, and high content of dense fibrous tissue. Symptomatic spinal cord tethering is often associated with caudal positioning of the conus medullaris; however, TCS can also occur in the presence of structural filum abnormalities despite a normally positioned conus medullaris. Surgical filum lysis in this circumstance has been shown to improve urological and neurological function. Recently, results of a number of noncontrolled, observational studies have suggested that surgical filum lysis may also benefit children with medically refractory voiding dysfunction, normal positioning of the conus medullaris, and a structurally normal terminal filum (as defined on 1.5-tesla MR images)—a set of characteristics also known as minimal TCS or occult TCS.

Consideration of occult TCS as a possible new indication for surgical untethering raises three principal questions. 1) Is there a well-defined population of children (and presumably previously untreated adults) with neurogenic voiding dysfunction? 2) Is there a plausible pathophysiological mechanism by which occult TCS could account for voiding dysfunction in these patients? 3) Does surgery favorably alter the natural history of children with occult TCS? The current analysis outlines existing evidence relevant to these questions.

Literature Review

A MEDLINE English-language literature search was undertaken for the period between 1964 and the present that covered the following topics: tethered spinal cord syndrome, “filum terminale,” and voiding dysfunction. Subsequently, reference lists from relevant primary articles were reviewed to identify other pertinent reports. Studies were considered relevant if they featured reports of clinical outcomes after surgical filum lysis for TCS, as well as reports of voiding and lower-extremity dysfunction. Only reports featuring children (that is, patients ≤ 18 years of age) with a structurally normal lumbar spine as defined on MR images were included. In general, normal imaging results were defined as the position of the conus medullaris lying above the lower endplate of L-2, the absence of syringomyelia, normal caliber and signal of the terminal filum, and the absence of other structural defects with the exception of simple osseous spina bifida occulta.

Data from these studies were reviewed in the context of the existing literature on the prevalence and natural history of voiding dysfunction in older children and adults so that recommendations for further study and clinical practice in this area could be formulated.

Abbreviations used in this paper: MR = magnetic resonance; NLC = normal-level conus; TCS = tethered cord syndrome.
Surgical treatment for tethered cord syndrome

Clinical Evidence for Improvement Following Surgery

Using the aforementioned search strategy, five reports of clinical outcome after surgical filum lysis for occult TCS were identified. Two additional studies were identified in which the authors reported the outcomes after filum lysis in children with NLC as established using computerized tomography myelography or MR imaging (Table 1). In total, these authors reported outcomes in 161 patients whose ages ranged from 2 to 18 years; 61% were female.

All patients with voiding dysfunction were treated medically before consideration of surgery. In general, medical therapy included timed voiding and other behavioral strategies, anticholinergic medications, and urological consultation. Various methods were used to select patients with NLC and occult TCS for surgery. In five studies, surgical candidates were chosen in part based on the presence of neurogenic changes on invasive urodynamics studies, including hypertonic bladder and sphincter-detrusor dyssynergia.18–21 In two other studies, neurological and/or cutaneous findings were used to select patients for surgery.18,19

The use of cine-MR imaging to evaluate conus medullaris excursion has also been advocated for this purpose.19

Averaged across all seven studies, 87% of patients improved after surgery (Table 1). When limited to studies of children with occult TCS as defined by nondiagnostic lumbar MR imaging, 92% improved with surgery. Similarly, 92% of patients with occult TCS improved regardless of whether they had18–21 or had not undergone19 urodynamical testing. In all studies, the preliminary method of outcome measurement was informal patient reporting to the neurosurgeon and/or urologist. The follow-up period ranged from 1 to 48 months and averaged approximately 1 year. Follow-up urodynamic test results were also reported in 59 patients from four studies.18–21 Sixty-three percent of these patients showed improvement in urodynamic variables after surgery (significantly fewer patients than those who showed clinical improvement). Three complications of surgery were reported in these 161 patients: one case of retethering leading to surgical reexploration and two cases of wound infection. No deaths or serious complications were reported.

Based on standard criteria for evidence review, all seven studies constituted Class III evidence for clinical outcome. The low level of evidence principally arose from the retrospective nature of these studies, the lack of formal, validated outcome measures, and the lack of randomized or cohort control groups.

Discussion

Multiple retrospective, observational noncontrolled studies suggest high rates of clinical benefit from surgical lysis of the terminal filum for occult TCS.18–21 Careful analysis of these results is required to determine the degree to which current clinical and scientific evidence supports the widespread adoption of surgery for occult TCS.

Voiding dysfunction is present in up to 17% of school-age children, although it occurs frequently in only 2%.15 Up to 16% of healthy nulliparous nursing students suffer regular and/or major episodes of urine loss.7 The finding that at least 15% of women and 3% of men in each decade of adult life suffer from voiding dysfunctions suggests that incontinence may persist into adulthood.16,17 Urinary incontinence is associated with a significant number of healthy adult men.3,6 Together, these findings reveal the existence of a large population of patients suffering from significant voiding dysfunction.

The concurrence of spina bifida occulta and voiding dysfunction may indicate the presence of TCS.18 Defined by these two criteria alone, the prevalence of TCS could range from 164,000 to 1,394,000 in 5- to 14-year-old children in the US (based on a rate of 2–17% incontinence and a 20% prevalence of spina bifida occulta19,20 as indicated on the US Census Bureau 2000 Report). These results should be interpreted with caution, however; for example, a prospective, long-term, observational study revealed no difference in the natural history of voiding dysfunction in children with or without spina bifida occulta.20

Surgical untethering of the spinal cord is postulated to prevent episodes of microcirculatory ischemia resulting from pathological spinal cord traction.21 The beneficial effects of spinal cord untethering in patients with occult TCS might then be explained in one of two ways. First, in patients with occult TCS the terminal filum may be structurally abnormal in a fashion not detectable on routine 1.5-tesla MR images.13 Alternatively, spinal cord parenchyma in patients with occult TCS may be abnormally sensitive to normal levels of traction resulting from the presence of an unremarkable terminal filum.

Data from recent reports demonstrate that the terminal filum is thicker and more fibrous in patients with occult TCS than in control patients. In patients with occult TCS, the terminal filum is also thicker than in patients without spinal cord tethering but not as thick as that found in patients with a radiographically apparent filum lipomata (that is, > 0.8 mm but < 2 mm in thickness).21 The filum is also more fibrous in patients with occult TCS than in control patients13 or in normal cadaveric specimens.12 These findings suggest that occult TCS involves a congenital abnormality of the terminal filum rather than the spinal cord itself. Thus, more sensitive 3-tesla MR imaging might be able to identify such patients noninvasively.

Urodynamics studies were used to select patients for sur-

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**TABLE 1**

Clinical reports of surgical lysis of the terminal filum for NLC or occult TCS

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients†</th>
<th>Imaging Type &amp; Result</th>
<th>Urodynamics</th>
<th>% Improvement‡</th>
</tr>
</thead>
<tbody>
<tr>
<td>Khoury, et al., 1990</td>
<td>31</td>
<td>myelography, NLC in 27</td>
<td>yes</td>
<td>72</td>
</tr>
<tr>
<td>Warder &amp; Oakes, 1993</td>
<td>7</td>
<td>MRI, abnormal, NLC</td>
<td>no</td>
<td>71</td>
</tr>
<tr>
<td>Nazar, et al., 1995</td>
<td>32</td>
<td>MRI, normal</td>
<td>yes</td>
<td>97</td>
</tr>
<tr>
<td>Palmer, et al., 1999</td>
<td>8</td>
<td>MRI, normal</td>
<td>yes</td>
<td>100</td>
</tr>
<tr>
<td>Selçukı, et al., 2000</td>
<td>17</td>
<td>MRI, normal</td>
<td>yes</td>
<td>76</td>
</tr>
<tr>
<td>Wehby, et al., 2004</td>
<td>60</td>
<td>MRI, normal</td>
<td>no</td>
<td>92</td>
</tr>
<tr>
<td>present study</td>
<td>6</td>
<td>MRI, normal</td>
<td>yes</td>
<td>100</td>
</tr>
</tbody>
</table>

† The total number of patients was 161.
‡ Mean = 87%.
surgery in both of the studies that demonstrated structural fila-
num abnormalities in patients with occult TCS.12,13 Although
these studies may thus be useful in identifying appropriate
surgical candidates, no information about the prevalence of a
mild terminal filum abnormality exists for the general
population. Furthermore, no evidence is available regarding
the sensitivity and specificity of urodynamic studies in de-
tecting such patients.

Other methods, in addition to clinical evaluation and uro-
dynamic studies, may also be available to identify patients
with occult TCS.19 For example, cine-MR imaging of the
distal spinal cord has been shown to predict the response to
spinal cord untethering in patients with major spinal dys-
raphism.7 However, normative cine-MR imaging data are
not available for healthy children or those with minor dys-
raphism, including filum lipoma and occult TCS.

Evidence for Improvement Following Surgery

Existing evidence suggests that there is a large popula-
tion of children (and possibly young adults) with medically
refractory voiding dysfunction who may suffer from occult
TCS. Recent pathophysiologic evidence supports the
hypothesis that the terminal filum in such patients is struc-
turally abnormal, although not to a degree that is detected
on routine 1.5-tesla MR imaging. A preponderance of the
available Class III outcome evidence demonstrates high
rates (> 90%) of clinical benefit from surgical filum lysis
in patients with occult TCS (Table 1). Such benefits were
demonstrated in five studies in which patients were selected
for surgery based in part on invasive urodynamic testing5,8,
9,11,13 and in one study in which urodynamic studies were not
used.19

Significant controversy exists regarding proper indica-
tions for performing surgical filum lysis in children with oc-
cult TCS. There are wide variations in existing clinical prac-
tice.14 Based on a preponderance of favorable Class III
evidence but in the absence of Class I or II evidence,
performance of a prospective, controlled randomized trial is
recommended.

Conclusions

Voiding dysfunction is a frequent clinical problem in chil-
dren and young adults. Patients with medically refractory
voiding dysfunction and neurogenic abnormalities on inva-
sive urodynamic testing have abnormally thickened, fibrous
terminal fila. A preponderance of Class III evidence, but no
Class I or II evidence, supports the option of surgical filum
lysis in these patients. A prospective controlled trial should
be undertaken to evaluate surgical filum lysis in children
with occult TCS.

Acknowledgments

I thank Shirley McCartney, Ph.D., for assistance in manuscript
preparation and editing, as well as Paul Steinbok, M.D., and Nalin
Gupta, M.D., for the invitation to present these findings to the Sec-
tion for Pediatric Neurological Surgery of the American Associa-
tion of Neurological Surgeons and the Congress of Neurological
Surgeons in San Francisco, California, in December 2004.

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Manuscript received August 16, 2005.
Accepted in final form January 13, 2006.
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J. Neurosurg: Pediatrics / Volume 104 / May, 2006