Occult tethered cord syndrome: a survey of practice patterns

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Object. Tethered cord syndrome (TCS) is associated with a number of congenital anomalies involving early development of the spinal cord. These include myelomeningocele, spinal cord lipoma, low-lying conus medullaris, and a fibrofatty terminal filum. Occult TCS occurs in patients when clinical features indicate a TCS but the typical anatomical abnormalities are lacking. It is controversial whether surgical release of the terminal filum leads to clinical improvement in a patient who does not have a previously identified anatomical abnormality. To assess the clinical standard used by practicing pediatric neurosurgeons, a practice survey was conducted at the 2004 Annual Meeting of the Joint Section for Pediatric Neurological Surgery of the American Association of Neurological Surgeons/Congress of Neurological Surgeons.

Methods. The survey examined clinical decision making for a same-case scenario with differing appearance on imaging studies. There was a clear consensus regarding diagnosis and treatment in the patient with symptoms, a low-lying conus medullaris, and a fatty terminal filum. The vast majority of respondents (85%) favored surgical untethering for this patient. A majority of respondents (67%) also favored treatment for the patient having symptoms and a fatty terminal filum. There was, however, significant disagreement regarding the diagnosis and treatment of disease in one patient with symptoms and an inconclusive magnetic resonance imaging study. Some respondents clearly favored surgery, whereas others believed that this patient did not meet the diagnostic criteria for TCS.

Conclusions. The results of this survey support the development of a randomized clinical trial to address the benefit of surgery for occult TCS.

KEY WORDS • tethered spinal cord • voiding dysfunction • spinal cord • terminal filum • pediatric neurosurgery

Tethered cord syndrome caused by a tight terminal filum has been known for more than 20 years and is recognized as a clinically important problem in children and adults.\textsuperscript{1,2,5} The pathogenesis of the clinical syndrome is believed to arise from traction on the lower end of the spinal cord by a thickened terminal filum. Traction on the conus medullaris then leads to decreased blood flow and decreased oxidative metabolism, which may eventually cause the clinical symptoms and signs of TCS.\textsuperscript{5} Its clinical features include neurological, musculoskeletal, and urological abnormalities that are often reversed or improved by sectioning of the terminal filum. The syndrome is typically associated with a low-lying conus medullaris (that is, its tip is located below the L-2 vertebral level). The thickening of the terminal filum and subsequent tethering of the spinal cord are thought to result from abnormal development of the filum during the retrogressive differentiation phase of secondary neurulation. Other features of spinal dysraphism, such as midline cutaneous lesions and osseous spina bifida, are commonly identified in these patients.

More recently, the concept of a TCS in which the conus is at the normal level has been proposed. In 1990, Khoury, et al.,\textsuperscript{,3} reported a case series of 31 children who had bladder instability that did not improve with conservative treatment, and in whom radiographic examinations revealed osseous spina bifida occulta and a normal-level conus. Because the clinical findings of neurogenic bladder dysfunction were similar to those in patients having TCS associated with the conus in a low position, a diagnosis of TCS was made. Postoperative improvement in urological symptoms occurred in 70% of the patients. Subsequently, a number of patients with a normally located conus were described in whom sectioning of the terminal filum resulted in improvement in the clinical findings of TCS.\textsuperscript{4,11,12} In almost all cases, the primary clinical abnormality has been urinary incontinence, which is thought to be of neurogenic origin. In some series, surgical
intervention has been considered only in cases in which there have been additional manifestations of occult spinal dysraphism, such as a cutaneous abnormality and/or the filum being found on MR imaging to be thicker than the normal 2 mm and to exhibit a signal consistent with it being fatty.\textsuperscript{3,11} In other series, however, the presence of a neurogenic bladder has been enough to justify sectioning of the filum.\textsuperscript{4,7,8,13}

Reports of clinical improvement of symptoms in children with so-called occult TCS following sectioning of the terminal filum are either anecdotal or derive from noncontrolled case series. No compelling scientific evidence exists that division of the filum in such children is beneficial. Adding to the difficulty of drawing conclusions from the reported case series is the fact that urinary incontinence in the school-age child, represented as a clinical diagnosis of either dysfunctional voiding or dysfunctional elimination, is a common problem, representing 20 to 40\% of office visits to pediatric urologists.\textsuperscript{9,10} The problem often resolves spontaneously, showing gradual improvement over time.\textsuperscript{6}

In light of this controversy, a general session was organized during the Annual Meeting of the Joint Section on Pediatric Neurological Surgery of the AANS/CNS in December 2004 to debate the existence of occult TCS and the evidence both for and against section of the terminal filum. This and the accompanying papers are based on presentations from the general session and the results of a questionnaire completed prior to and after the session.

**Clinical Material and Methods**

A questionnaire was created and distributed to attendees at the annual meeting of the AANS/CNS Section on Pediatric Neurological Surgery in December 2004. Attendees were asked to complete the questionnaire before and after a debate and discussion on the topic of occult TCS, specifically addressing whether or not to cut the terminal filum in children who presented with urinary incontinence but in whom the conus medullaris terminates at a normal level. In the questionnaire, a clinical case was presented as follows: A 6-year-old girl is referred by a pediatric urologist because the conus ending at L-2 with a normal filum (Fig. 1 upper left and upper center); Case 2, a normal conus ending at L-2 with a normal filum (Fig. 1 upper right); Case 3, a normal conus ending at L-1 with some fat in a slightly thickened filament (Fig. 1 lower left and lower center); and Case 4, a normal conus ending at L-1 with a normal filum and a T7–9 syrinx having a maximum diameter of 2 to 3 mm and no Chiari malformation (Fig. 1 lower right).

For each case scenario, participants were asked to indicate on a five-point Likert scale (1, agree; 3, unsure; 5, disagree) the extent to which they agreed or disagreed that 1) the patient had a TCS; and 2) the patient should undergo sectioning of the terminal filum.

Participants were asked to complete the questionnaire both prior to and after the debate and discussion on the topic. They were asked to indicate their professional role as designated by the following categories: resident or fellow; attending neurosurgeon certified by the ABPNS; attending neurosurgeon not certified by the ABPNS; registered nurse or physician assistant; or other. In addition, they were asked how many years they had been in practice. The information from the completed questionnaires was entered into an Excel spreadsheet and analyzed.

**Results**

**Overall Findings**

One hundred five participants completed some or all of the questions. Of the respondents who indicated their status, 51\% were ABPNS neurosurgeons and 47\% had 15 or more years of practice experience (Fig. 2). There was a strong correlation between ABPNS membership and experience level, with 66\% of ABPNS members having greater than 15 years’ experience, compared with 35\% of non-ABPNS members (\(p = 0.006\) for the correlation between increasing practice experience and ABPNS membership).

**Case 1**

Both before and after the discussion, virtually all respondents agreed with the diagnosis of TCS and the recommendation for surgical release (Fig. 3 upper left).

**Case 2**

In this case, involving a normal filum and normal position of the conus, there was both disagreement and uncertainty regarding the diagnosis of TCS and the recommendation for release by section of the filum. Before the discussion, 22 and 30\% of respondents had either some or full agreement, respectively, with the statement that the patient had TCS and should undergo an untethering procedure, whereas 41 and 43\% of respondents had some degree of disagreement with the diagnosis of tethering and recommendation for untethering, respectively. In addition, 36\% of respondents were uncertain as to whether or not a tethered cord was present and 26\% were uncertain as to whether or not operative release should be performed. After the discussion, 22 and 29\% reported some degree of agreement with the tethered cord diagnosis and treatment, respectively, whereas 36 and 35\% disagreed, and 42 and 35\% were uncertain. Overall, there was an even distribution of opinions across the spectrum for this question, strongly supporting the impression that equipoise exists (Fig. 3 upper right). Using a null hypothesis that the distribution of opinions around the midpoint is symmetric, there is a narrow statistical shift of the distribution to the left (toward disagreement) for the question as to the presence of a tethered cord, and no statistically evident asymmetry around the median value of 3. Our conclusion, arrived at using the Wilcoxon rank-sum test, is that there is uncertainty among clinicians over whether or not the patient should undergo an untethering procedure.

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Case 3

Given a normally located conus, the presence of fat in the terminal filum significantly altered the clinical impressions of the respondents, who expressed more agreement that this factor indicated cord tethering and that the problem should be addressed surgically (p = 0.0001, Pearson chi-square test, Fig. 3 lower left). Prior to the discussion, 67% of respondents had some degree of agreement with the diagnosis of TCS and 76% agreed with treatment involving section of the filum. The values for the postdiscussion results were similar.

Case 4

The respondents showed a broad spectrum of opinions on the diagnosis of a tethered spinal cord and treatment by section of the filum given the presence of a small thoracic syrinx (Fig. 3 lower right). Prior to the debate and discussion, 34 and 38% had some degree of agreement with the diagnosis of cord tethering and treatment by section of the terminal filum, respectively, whereas 40 and 43% disagreed and 23 and 16% were uncertain. After the discussion, the values were 41 and 43% for some agreement (diagnosis and treatment), 36 and 39% for some disagreement, 22 and 14% were uncertain, respectively. Compared with survey results in Case 2, the presence of a syrinx increased the likelihood of agreement with the diagnosis of TCS and with section of the filum as the treatment of choice, but the difference was not statistically significant (p < 0.05, Wilcoxon signed-rank test).

Impact of Experience on Opinions of Respondents

A strong positive relationship was found between ABPNS membership and number of years of practice experience (p = 0.006, Pearson chi-square test). On average, ABPNS members were more likely to support the diagnosis and treatment of a tethered spinal cord in the presented clinical scenarios than were non-ABPNS members (Fig. 4). Using the mean response for purposes of illustration,

![Magnetic resonance images. Case 1: A conus medullaris ending at L3–4 (upper left) and a thick, fatty terminal filum (upper center). Case 2: A conus ending normally at L-2 and a normal filum (upper right). Case 3: A normal conus ending at L-1 (lower left) and a fatty filum (lower center). Case 4: A normal conus ending at L-1 and a syrinx from T7–9 (maximum diameter 2 mm; lower right).]
ABPNS members were slightly more likely in each of the eight questions to support the diagnosis of TCS in the clinical scenario and more likely to agree with cutting the terminal filum. This difference was present both before and after the discussion. On a per-question basis, this was not a statistically significant difference, but taken over the entire questionnaire, it was significant ($z = -3.865$, $p < 0.000$, Mann–Whitney U-test).

To determine the possible impact of the discussion on the opinions of the participants, the number of participants indicating a greater degree of agreement or disagreement with the diagnosis and treatment statements was calculated as a percentage of the total number of responses for which both before and after discussion information was available. Except for Case 1, between 20 and 30% of participants reported some change in opinion following the discussion. As shown in Table 1, the impact of the discussion on the participants was not uniform across all of the scenarios, and none of the differences in distribution between the pre- and post-discussion results were statistically significant on a per-question basis. There was little overall change in opinion regarding Case 1. For Case 2—a normal conus and filum—more individuals increased their agreement with the diagnosis and treatment than increased their disagreement. For Case 3—a normally positioned conus but an abnormal filum—there was an increase in disagreement with both diagnosis and treatment. For Case 4—a normal position of the conus and normal filum but a small syrinx—there was a somewhat greater movement toward agreement. A compar-

![Fig. 2. Bar graphs. Upper: The nature of the respondents. Lower: The number of years of experience of the respondents.](image)

![Fig. 3. Bar graphs showing responses before and after debate and discussion about Cases 1 (upper left), 2 (upper right), 3 (lower left), and 4 (lower right).](image)

![Fig. 4. Bar graph showing comparison of responses from those certified and not certified by the ABPNS.](image)
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**TABLE 1**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Increasing Agreement (%)</th>
<th>Increasing Disagreement (%)</th>
<th>Change (%)</th>
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<tr>
<td>1</td>
<td>1.0</td>
<td>3.1</td>
<td>4.1</td>
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<tr>
<td></td>
<td>treatment</td>
<td>3.1</td>
<td>4.1</td>
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<tr>
<td>2</td>
<td>17.9</td>
<td>10.5</td>
<td>28.4</td>
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<td>8.4</td>
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<tr>
<td>3</td>
<td>8.4</td>
<td>12.6</td>
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<td>16.8</td>
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<td>4</td>
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<tr>
<td></td>
<td>treatment</td>
<td>23.4</td>
<td>12.8</td>
</tr>
</tbody>
</table>

ision of the changed responses of ABPNS with non-ABPNS neurosurgeons showed no clearly definable trends or differences from Cases 2 to 4.

**Discussion**

For the questionnaire, Case 1 was selected as an example of a typical tethered cord with a low-lying conus and a thickened fatty filum demonstrated on MR imaging. We anticipated that in the setting of urinary incontinence, abnormal urodynamics, and these MR imaging findings, there would be agreement as to the diagnosis of TCS and that sectioning of the filum was indicated. The results of the responses to the questionnaire confirmed this viewpoint.

It was expected that there would be much less certainty regarding the diagnosis of TCS and/or the advisability of sectioning the filum in patients having the same clinical findings but with a conus in a normal position. The results of this survey indicate that for patients who have clinical symptoms of incontinence and urodynamics consistent with a tethered cord, as well as a normal filum and a normally positioned conus, there is uncertainty among pediatric neurosurgeons as to both the diagnosis of a tethered cord and the need for treatment involving sectioning the terminal filum. The presence of fat in the filum significantly increased the degree of agreement with the diagnosis of a tethered cord and a treatment plan involving sectioning the filum, whereas the presence of a syrinx in the absence of a fatty filum produced a nonstatistically significant increase in agreement with diagnosis and surgical treatment for a tethered spinal cord. Regarding the cases with a normally positioned conus, a number of respondents changed their opinions as a result of the debate and the discussion on the topic. The amount of variation in opinions was similar for experienced compared with less experienced neurosurgeons, and for neurosurgeons certified by the ABPNS compared with those not certified. In general, ABPNS-certified neurosurgeons were more likely to favor cutting the filum; this may reflect a higher comfort level with the surgical procedure, a more liberal view of how tethering of the cord might occur, or a combination of both.

These results indicate that sufficient clinical equipoise exists to justify ethically a randomized clinical trial for patients having a normally positioned conus and normal-ap- pearing terminal filum; inclusion of patients with syringo-

myelia in such a trial could also be justified if desired. Additional debate may be needed with regard to the inclusion of patients with fatty fila, as only 6% of respondents absolutely disagreed that sectioning the filum was advisable (although 44% were uncertain).

**Conclusions**

The results of a questionnaire-based survey of attendees at the Annual Meeting of the AANS/CNS Section on Pediatric Neurosurgery in December 2004 confirmed a high level of uncertainty over whether or not to cut the filum in a child with neurogenic urinary incontinence and a normally positioned conus. These results establish the presence of clinical equipoise, ethically justifying a randomized clinical trial to study this question and indicating that patient accrual would be acceptable.

**References**


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