1 Introduction to the Tethered Cord Syndrome

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The word *tether* means to restrain, an example of which is an animal held to the maximal range of motion by a rope. This definition leads to the connotation "the harder the pull on the rope, the tenser the rope." Applying the word *tethered* to the lumbosacral spinal cord, one visualizes an unnatural, unmitigated, abnormal constraint that aptly applies to the medical condition called tethered cord syndrome. However, there has been much uncertainty about this syndrome and its diagnosis and treatment because clinicians and scientists did not agree with the usage of this term and because perceptions were based on visual rather than on scientific evidence (F. Anderson, personal communication 1984). Interestingly, there is no word that has the same definition and connotation as English *tether* in any other language. The delay in recognizing this syndrome may be due in part to this linguistic difference.

There is now agreement that tethered cord syndrome (TCS) is a functional disorder caused by stretching of the spinal cord with its caudal end fastened by an inelastic structure.[1] The disorder is made worse as spinal cord tension increases with such influences as rapid growth in children, or forceful spinal flexion and extension.

■ Historical Background of Tethered Cord Syndrome

The concept of the tethered cord syndrome evolved slowly but with increasing interest among clinicians and pathologists. A suggestion that stretching of the spinal cord could induce a disorder came from a case with myelomeningocele in 1910.[2] Other articles followed on disorders such as a sacral lipoma,[3,4] and occult spinal dysraphism[5]; however, the expression *tethered cord* was avoided in such writings, which appeared to have linked the characteristic neurological deficits to "lipoma infiltration or congenital neuronal dysgenesis." Although Garceau and others attributed the neurological deficits to the traction effect that a tight filum[6,7] or myelomeningocele[8] exerted on the spinal cord, none of these articles was published in neurosurgical journals.

In 1940, Lichtenstein, an authoritative neuropathologist, was first to propose that tethering of the spinal cord may cause paraplegia and herniation of the brain stem and cerebellum through the foramen magnum.[9] However, his hypothesis was not accepted, particularly for the development of Chiari malformation.[10–12] Although surgeons noticed neurological improvements in their patients after what is currently called untethering of the spinal cord, two questions remained unanswered. First, if tethering-induced symptoms exist, what part of the nervous system is affected? And second, what is the pathophysiological basis for any reversible lesion?

In 1976 Hoffman et al adopted the term *tethered spinal cord* in a report on 31 patients presenting with incontinence and motor and sensory deficits in the lower limbs. These symptoms subsided after sectioning of a thickened filum terminale,[13] which indicated that the

neurological lesion was in the lumbosacral cord. In 1981, Yamada et al demonstrated impairments of oxidative metabolism in the lumbosacral cord before surgery and recovery from the impairments after surgery in patients who had the same clinical presentations as those described by Hoffman et al.[1] Simultaneously, electrophysiological impairments and recoveries were recorded before and after cord untethering, respectively.[1,14]

McLone [moderator] and a panel reviewed such pathophysiological and clinical information during a debate titled "Is the TCS Fact or Fiction?" One conclusion was that tethered spinal cord is a clinical entity based on scientific evidence.[15] Since then, the term *TCS* has increasingly appeared in the neurosurgical literature.[16–20]

■ Current Understanding of Tethered Cord Syndrome

Expanding the stretch-induced disorder from tethered spinal cord to tethered cord syndrome, Yamada et al included patients with neural spinal dysraphism located in the caudal end of the spinal cord, such as myelomeningoceles (MMCs) and lipomyelomeningoceles (LMMCs).[1] This definition engendered misinterpretations and questions despite the fact that these patients presented with the same symptomatology and oxidative metabolic impairment and postoperative metabolic and neurological improvement as those of tethered spinal cord of Hoffman et al.[13]

Two questions evolved: (1) How could the theory of TCS pathophysiology explain the downhill course in some MMC patients after repeated surgical repairs?[21] and (2) Why are surgical results of TCS patients so variable?[22]

Answers to these questions may be derived from a better definition of the differences between TCS and the expressions *cord tethering* or *tethered cord* and from studies of TCS pathophysiology. Analyzing these questions, it became apparent that they are missing the fact that TCS is the manifestation of a stretch-induced lesion above the inelastic structure that exerts traction force to the spinal cord. In contrast, MMCs and LMMCs that are located dorsal to the spinal cord can cause neurological deficits by local compression or ischemia, or as a part of neuronal dysgenesis. It is clear that the lumbosacral neurological symptomatology in these patients is not caused by caudal traction effects and not considered TCS.[23]

At the request of Professor Sergio DiRocco to clarify the diagnosis of TCS (DiRocco S, personal communication, 2006), the editor felt it necessary to categorize such expressions as *cord tethering* and *tethered cord*, which are derived only from visuals. Based on the pathophysiological analysis on caudal spinal cord anomalies, Yamada and Won divided these into three categories based on experience with individual clinical cases.[24]

Category 1 represents patients with true TCS who exhibited neurological signs and symptoms due to anchoring structures restricting the spinal cord movement at its caudal end. They include an inelastic filum terminale, caudal lipoma or lipomyelomeningocele, or sacral myelomeningocele.[25]

Category 2 includes patients whose signs and symptoms resemble those of true TCS; however, signs and symptoms are associated with large MMCs, extensive dorsal or transitional LMMCs (see Chapter 11), and postoperative MMCs with extensive fibrous adhesions (category 2A). These structures cause local compression or ischemia to the spinal cord causing neurological deficits. In some cases the deficits are related to neuronal dysgenesis. These patients do not belong to true TCS. Only when a part of the signs and symptoms is indicative of a lesion above the anomalies is partial TCS an appropriate diagnosis (category 2B).

Category 3 patients typically present with thoracolumbar MMC and exhibit total paraplegia and urinary and bowel incontinence due to the lack of functional neurons in the lumbosacral region of the cord.

Surgical results differ depending on the patient's category. After surgical untethering,

category 1 patients can expect excellent outcome with pain relief and neurological improvement. Category 2B patients also have good outcome with similar symptomatic improvement. Category 2A patients have good pain relief with deficits stabilized but no neurological improvement. Category 3 patients have no predictable neurological improvement. No surgical treatment is indicated for category 3 patients. There is no hope for reversing incontinence in patients who have been performing intermittent catheterization for more than a few years.

There are special case reports other than the ordinary untethering surgery. One is a report of relief of severe back and leg pain after cord transection was performed on patients with severe adhesive arachnoiditis around the caudal spinal cord and cauda equina.[26] When the neurological signs indicated a neurological lesion above the transection, the diagnosis of TCS can be appropriate. Another report indicated reversal of TCS symptomatology by lumbar corpectomy in the patients with magnetic resonance imaging (MRI) evidence of severe arachnoiditis. This method relieved spinal cord tension by shortening the length of the vertebral column.[27] These atypical treatments will further stimulate the advancement of TCS studies.

Other chapters included in this book cover the pathophysiology of TCS (Chapter 3), embryological analysis of TCS (Chapter 2), neurological examination (Chapter 4), imaging studies of TCS (Chapters 5 and 6), pediatric tethered spinal cord (Chapter 10), adult TCS (Chapters 15 and 16), cervical tethered spinal cord (Chapter 9), TCS associated with myelomeningoceles and lipomyelomeningoceles (Chapter 11), folate studies in families of neural spinal dysraphism (Chapter 12), in utero repair of myelomeningocele (Chapter 13), TCS associated with dermoid (Chapter 14), urological aspects of TCS (Chapters 7 and 8), intraoperative stimulation studies on sacral cord and roots (Chapter 19), anomaly of the lumbosacral cord (Chapter 17), spinal cord length and filum thickness (Chapter 18), and electrophysiology with somatosensory evoked potentials (SSEPs) for TCS evaluation (Chapter 20), and for conservative and operative treatment (Chapter 21). Each chapter follows the principle of TCS with expertise and endeavors to improve the care of TCS patients to their full potential capacities by proper diagnosis and treatment.

Topics regarding TCS discussed in this book include the following:

- Correlation of pathophysiology to clinical manifestation of TCS
- Signs and symptoms of TCS to the spinal cord level of neurological lesions
- Distal motor and sensory lesions in lower limbs
- Patchy motor and sensory lesions in TCS patients
- Incontinence becoming irreversible earlier than motor and sensory dysfunction
- Hypoactive deep tendon reflex (DTR) seen in TCS patients
- Mechanism of later development of TCS in adult and late teenagers
- Difference in neurological improvement after what appeared to be complete untethering procedures
- Surgical indications and surgical techniques for TCS patients
- Essential part of surgical procedures for untethering
- Complications to be anticipated
- Rationale for surgical treatment and alternative treatment

The editor recommends that neurosurgeons identify the three categories for patients who present with so-called cord tethering and select the true TCS (category 1) and relative TCS (category 2B), non-TCS patients with symptoms similar to TCS. By following the categorization of cord tethering, neurosurgeons can correctly envisage the surgical outcome for the patients in each group. They will be able to convince referring physicians as well as patients and families of their ability to properly diagnose and treat adult TCS patients and to handle those who show similar symptomatology but belong to categories 2A and 3.

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18 Anomalies of the Spinal Cord Length and Filum Thickness

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The adults with tethered cord syndrome (TCS) are divided into two groups: group 1 adult TCS patients with spinal dysraphism and group 2 patients without dysraphism who develop signs and symptoms in adulthood.[1–3] A significant number of group 2 patients failed to show an elongated spinal cord and thickened filum terminale that Hoffman et al described in pediatric patients.[4] This chapter reports the statistics derived from the studies on the location of the caudal end of the spinal cord and the thickness of the filum terminale in group 2 adult TCS patients ($N = 104$). Only 35.6% of them were found to have both elongated cord and thickened filum. The data indirectly support that the essential factor for development of TCS is the inelastic filum.

Based on the neurological signs and symptoms localized in the lumbosacral cord and consistent imaging and operative findings, Hoffman et al adopted the term *tethered spinal cord*.[4] They attributed the motor and sensory deficits and incontinence to an increased tension in the elongated spinal cord anchored by a thickened filum terminale. As neurosurgeons' experience continued to increase, however, the concept of tethering-induced disorder broadened to the recognition of an inelastic filum as an essential mechanical factor for development of TCS.[5,6] Earlier in 1993, Warder and Oakes reported that 18% of TCS patients, including both children and adults, had the caudal end of the spinal cord at L1–2 intervertebral space or higher.[7] The authors present the retrospective analysis of the data on the caudal end of the spinal cord and filum thickness obtained from group 2 TCS in adult and late teenage patients as previously discussed.[8]

■ Materials and Method

One hundred and four patients within the group 2 adult and late teenage TCS, from 17 to 81 years of age, 39 males and 65 females, presented with typical signs and symptoms of TCS (Chapter 15). All the patients lacked spinal dysraphism on imaging studies as well as at operation, except for five patients with bony spina bifida occulta of the S3 through coccygeal vertebrae shown by plain x-ray films. During surgery, the level of the caudal end of the spinal cord was determined by the exit of the lowest coccygeal nerve root, and its location was expressed in relation to the lumbar and sacral vertebral bodies. The filum thickness was measured by the lateral diameter (usually slightly greater than the anteroposterior diameter).

■ Results

The locations of the caudal end of the conus and the diameter of the filum terminale are listed in **Table 18.1**. Only 35.6% of the group 2 TCS patients showed cord elongation and a thickened filum. The caudal end of the spinal cord was found at the L2–3 intervertebral space

Table 18.1 Locations of the Caudal End of the Conus and the Diameter of the Filum Terminale (from total 104 adult cases with TCS)

1. Normal range of caudal end and filum thickness	30 cases (28.8%)
2. Normal range of caudal end and abnormally thick filum	7 cases (6.7%)
3. Low-lying caudal end and normal range filum thickness	30 cases (28.8%)
4. Low-lying caudal end and abnormally thick filum	37 cases (35.6%)

Adding 1. and 2., 37 patients (35.6%) had the caudal end at L2–3 or above, and adding 3. and 4. the diameter of the filum was less than 2 mm in 60 patients (57.7%).

or above in 37 patients (35.6%), and below the L2–3 level in 67 patients (64.4%). The diameter of the filum was less than 2 mm (assumed to be in the normal range) in 60 patients (57.7%) and 2 mm or greater in 44 patients (42.3%).

Of 67 patients with the caudal end below the L2–3, 41 (39.4% of total) had the caudal end opposite to the L3 vertebra, leaving only 26 (25%) with the caudal end below the L3 vertebra. None of the cases with the caudal end at the L3 level was detected by imaging studies as having an elongated spinal cord (Chapter 3).

■ Discussion

Our data clearly demonstrate the variability of the cord length and filum thickness in adults and late teenage patients with TCS, which correspond to the previous report.[8] Warder and Oakes first described no elongation of the spinal cord in adult and pediatric TCS patients.[7] There are also an increasing number of pediatric TCS patients who failed to show these two features (Knierim DS and Won DJ, personal communication, 2007). Other authors diagnosed TCS in small children with a chief complaint of incontinence but without these two anatomical features.[9–11] These facts explain how difficult it is to diagnose TCS by relying on only two features, an elongated cord and a thickened filum. Despite these recent reliable findings, it is still widely believed that an elongated spinal cord and thick filum are the two fundamental features to establish the diagnosis of TCS. Further, this logic extends to such an assumption as the patients without two features are excluded from TCS or tethered spinal cord, even if they present with typical signs and symptoms of TCS (Chapter 15). The data reported by Younts et al are noticeable; the normal range of filum thickness was measured as 1.1 to 1.2 mm,[12] whereas the filum thickness in TCS patients ranged from 1.0 to 2.0 mm.[13–15]

We emphasize that the clinical findings[16] are the primary tools for the diagnosis of TCS (Chapter 15), and are assisted by the magnetic resonance imaging (MRI) findings, such as the two features mentioned earlier, fat signal in the filum and syringomyelia,[17] The most consistent MRI finding is the posterior displacement of the conus and filum. This feature is confirmed by intrathecal endoscopy before widely opening the dura and arachnoid membrane, or preoperative percutaneous endoscopy.[6] The further step to prove the lack of elasticity in the filum is the stretch test after exposing the filum. This is the logical approach that utilizes the experimental work on redox changes in cytochrome a,a_3[18] proportionate to cord elongation induced by traction,[18–20] detailed in Chapter 3.

Although fat tissue in the filum has been emphasized for the diagnosis of TCS,[17] fat itself is soft and elastic, and TCS may not develop until fibrous tissue is increased to sufficiently reduce filum elasticity. MRI studies with currently available resolution do not identify the caudal end of the spinal cord, particularly when it is located at the L3 vertebral level. This difficulty can be explained by the inability to locate the exit of the lowest coccygeal nerve root (100 μm in diameter),[2] which is too small to be identified as a landmark of the caudal end of the spinal

cord. Commonly, the caudal end at the L3 vertebra is not included in the group of low-lying cord before surgery.[1–3] It is advisable to consider the conus localization by MRI studies (see Chapter 5, Fig. 5.2). In some cases, fat tissue that extends from the filum into the conus makes the definition of the conus–filum junction impossible by imaging studies. Only at operation, the junction can be determined by the exit of the lowest coccygeal root with microscopic observation (see Chapter 15, Fig. 15.10A,B,C). The elongation of the spinal cord usually occurs in lumbosacral segments, and the conus diameter is often greater than the normally located conus (S2–4 segments). The relatively large bulk of the conus in these patients may resist traction forces as much as the lumbar cord segments do, and explain widely spread lumbosacral cord dysfunction in TCS.

■ Conclusion

The length of the spinal cord and the thickness of the filum terminale are of relative importance for the diagnosis of the TCS. Neurological and musculoskeletal abnormalities that indicate or strongly suggest a stretch-induced lumbosacral functional lesion make imaging studies useful. The symptomatic protocol presented in Chapter 15 will assist in the diagnosis for TCS in adults as well as in children.

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