Tethered cord syndrome: a review of the literature from embryology to adult presentation

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Tethered cord syndrome (TCS) is a clinical condition of various origins that arises from tension on the spinal cord. Radiographic findings may include the conus medullaris in a lower than normal position, fatty infiltration of the filum terminale, lipomyelomeningocele, myelomeningocele, myelocystocele, meningocele, split cord malformations, dural sinus, anorectal malformations, and intraspinal tumors. The clinical constellation of signs and symptoms associated with TCS may include dermatologic, urological, gastrointestinal, neurological, and orthopedic findings. The current review focuses on TCS by age group of the more common causes of the condition, including myelomeningocele, lipomyelomeningocele, as well as the adult presentation of occult TCS. Pertinent review of the neuroembryology and normal anatomical position of the conus medullaris is included. (DOI: 10.3171/2010.3.FOCUS1079)

KEY WORDS • tethered cord syndrome • myelomeningocele • lipomyelomeningocele • occult tethered cord • natural history

TCS,13,35 Most of the patients with normal conus position but TCS are reported to have associated findings such as cutaneous stigmata, vertebral abnormalities, intradural lipoma, and neurological abnormalities on examination. In this patient population, symptoms of pain and bowel or bladder incontinence appeared to be responsive to detethering.

Embryology

Knowledge of neural embryology is essential to understanding TCS because it is commonly associated with a variety of disorders that are a result of abnormal development of the nervous system. A brief review of caudal nervous system embryology aids in understanding the anatomical basis of TCS (Figs. 1–3). The neural tube forms during the process of neurulation, which occurs during Days 18–28 of gestation. Initially, the ectoderm overlying the notochord proliferates to form the neural plate, which subsequently invaginates to form the neural tube, which then closes to form the neural plate, which subsequently involutes to form the neural folds and then closes to form the neural tube.5,24 The process of neural tube closure begins by Day 22–23 and extends cephalad and caudad with the anterior neuropore closing last by Day 25–27.26 Following neurulation, the distal neural tube undergoes canalization. Distal to the posterior neuropore, undifferentiated cells from the primitive streak form the caudal cell mass. The distal neural tube forms from fused vacuoles that developed from the caudal cell mass. This structure, in turn, develops into the conus medullaris, cauda equina, and filum terminale. During

Abbreviations used in this paper: MMC = myelomeningocele; TCS = tethered cord syndrome.
the end of the canalization period (Days 43–48), the ventriculus terminalis forms at the terminal end of the neural tube near the coccyx, marking the site of the future conus medullaris.26

As the gestational period continues the caudal spinal cord undergoes retrogressive differentiation, resulting in the filum terminale, cauda equina, and ascension of the conus in relation to the vertebral bodies. Neural tissue caudal to the ventriculus terminalis regresses to form the filum terminale. Simultaneously, the vertebral column grows at a disproportionate rate to the spinal cord, resulting in the ascension of the conus and elongation of the filum. The cauda equina forms as nerve roots grow longer to accommodate the differential growth. The regression process continues into the postnatal period with the conus reaching the adult level of L1–2 by approximately 3 months of age. The normal position of the conus postnatally has been elucidated in multiple anatomical reviews (Table 1). The cadaveric studies of Reimann and Anson37 and Pinto et al.34 demonstrated that the conus lies above the L-2 vertebral body in 95% of specimens. Similarly, in a study correlating the vertebral level of cord termination with gestational age, Barson3 demonstrated its ascent and final position to lie at or above the L1–2 disc space in the early postnatal period.

Diagnosis

Clinical Presentation of TCS Based on Age

The clinical presentation of TCS is broad and varies with age at presentation as well as underlying cause. The physical examination plays a key role in determining the diagnosis of TCS. The dorsal spine should be examined for cutaneous manifestations of spina bifida as well as the presence of a scoliotic deformity. The presence of cutaneous stigmata associated with spina bifida may be the only evidence of a tethering lesion in the neonate and infant. Findings may include lumps, nevi, lipomas, hair tufts, hemangiomas, and dermal sinuses (Fig. 4).16,20,21 The lower extremities should be evaluated for orthopedic deformities. Motor and sensory testing should be assessed thoroughly as skip lesions are common. Gait assessment is extremely important and can be affected by orthopedic deformities (scoliosis or foot deformities) or spasticity.26 The deep tendon reflexes and muscle tone are variable. Sphincter disturbances may be difficult to discern in patients younger than 1 year of age, but lower-extremity deformities or anorectal malformations should raise suspicion for an associated cord tethering. Clinical presentation in toddlers and children is commonly associated with both motor and sensory dysfunction. Additionally, a
regression in motor function or bladder control or an arrest in achievement of developmental milestones may reveal important cues, including gait difficulties, sensory deficits, development or progression of scoliosis, or foot deformities. Sensory loss tends to occur in a nonsegmental distribution. Back and lower-extremity pain may be a presenting complaint in this age group. In the late childhood and teenage years, nondermatomal pain in the lumbosacral region, perineum, and legs is the predominant symptom. Progression of a scoliotic deformity often contributes significantly to complaints of pain. Sphincter dysfunction and incontinence may also be a predominant symptom. 

Weakness may be subtle and present only in a single muscle group. New onset diagnosis of TCS has been described following a history of sexual dysfunction. In the subset of adults who present without history of spina bifida, pain is the most common presentation, followed by weakness and urological dysfunction. In the patient without orthopedic deformities or urological dysfunction, trauma often leads to symptomatology. The trauma may be mild (pregnancy, childbirth, exercise) or involve a major direct trauma to the spine. It is hypothesized that the degree of tethering in this subset of patients is not significant enough to cause symptoms alone, but trauma increases the stress on the already tense spinal cord, altering microcirculation and cellular metabolism and eventually leading to neurological deterioration.

Urodynamic Studies

Sphincter dysfunction may play a part in the clinical presentation of TCS in all age groups. The most common finding is detrusor hyperreflexia, but decreased bladder compliance, dyssynergia, and decreased sensation can also occur. Important parameters in assessing a neurogenic bladder include total bladder capacity and pressure, leak point pressure, compliance, uninhibited contractions, electromyelogram activity, and sensation. Formal urodynamic testing plays an important role in establishing sphincter dysfunction during the primary diagnosis as well as serving as an indicator of deterioration during a course of watchful observation. Urinary sphincter worsening as detected by urodynamic studies often precedes

TABLE 1: Anatomical studies of the normal position of the conus*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Cadavers</th>
<th>Level of Conus Termination</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reimann &amp; Anson, 1944</td>
<td>129</td>
<td>at or above L-2 vertebra in 95.5%</td>
</tr>
<tr>
<td>Barson, 1970</td>
<td>252 (infants)</td>
<td>above L1–2 disc space</td>
</tr>
<tr>
<td>Pinto et al., 2002</td>
<td>41</td>
<td>at or above L-2 vertebra in 95%</td>
</tr>
</tbody>
</table>

* All cadaveric studies.
clinical manifestations of deterioration. In patients with abnormal formal urodynamic testing, whether clinically symptomatic or asymptomatic, improvement from 29% to 75% has been observed following detethering in a broad range of patients. Formal urodynamic tests have been used as a marker to document improvement or stability of function following a detethering procedure.

Radiographic Studies

Magnetic resonance imaging is the radiographic modality of choice for evaluating TCS (Figs. 4–7). Magnetic resonance imaging demonstrates the level of the conus, often visualizes the cause of tethering, and provides detail for potential surgical planning. Common lesions associated with TCS, including meningoceles, MMCs, split cord malformations, and dermal and lipomatous tumors, are all readily demonstrated on MR imaging. In the absence of spina bifida or a tumor, MR imaging is used primarily to identify the level of the conus and the nature of the filum terminale. Although there is a great deal of variability, a conus positioned at or above L-2 is considered to be at the normal level. Thickness of the filum terminale greater than 2 mm is considered abnormal in children, although this finding remains controversial. Additionally, supine and prone MR imaging can be performed to demonstrate evidence of motion of the spinal cord in the prone position. Lack of motion suggests TCS. Bone imaging with plain films or CT aids in the evaluation of scoliosis.

Common Causes of TCS

Myelomeningocele

All patients with MMC are born with a tethered cord and at birth, or shortly thereafter, undergo a repair and closure (Fig. 6). Tethered cord syndrome occurs in as
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many as 2.8–32% of patients with MMC as a result of retethering as the vertebral column grows and lengthens (Table 2). Retethering occurs from scarring at the site of prior repair and anchors the cord, preventing it from ascending during growth. Symptoms usually consist of pain, weakness, foot deformity, scoliosis, and bowel and/or bladder dysfunction. Patient age at retethering is approximately between 5 and 9 years, usually during stages of rapid growth. Phuong et al. retrospectively reviewed 45 cases of repaired MMC with TCS that did not undergo a detethering procedure and found that 60% of the patients progressed with TCS and 89% needed other surgical procedures, including contracture release and bladder augmentation. Class 1 or 2 evidence does not yet exist for timing of detethering, but George and Fagan performed an evidence-based literature review and reported that tethered cord release should be considered when symptoms, imaging studies, urodynamics, and somatosensory evoked potentials are consistent with TCS. Patients should be treated within 5 years of onset of symptoms for best outcome and regular follow-up examinations are recommended to evaluate for retethering. Selber and Dias reported on 46 patients with MMC and noted that 12 patients required tethered cord release and all 12 experienced improvement of symptoms. Magnetic resonance imaging findings of the conus medullaris at S-1 or lower has been shown to predict late TCS in patients with MMC and poorer outcome after detethering. Most centers follow-up children with MMC regularly with multidisciplinary teams that evaluate urodynamic studies, muscle strength and neurological function, pain, and any orthopedic issues. Imaging alone is not a reliable

Fig. 5. Sagittal (A) and axial (B) T1-weighted MR images of 5-month-old patient. The conus medullaris terminates at the mid L-3 vertebral body, with marked thickening and fatty infiltration of the filum. The sagittal image demonstrates a syrinx that extends into the lower thoracic cord.

Fig. 6. Sagittal intrauterine MR imaging of a 22-week, 5-day-old fetus demonstrating a neural tube defect inferior to L-3. There is an absence of the posterior elements in the lumbar area. A large dorsal MMC measuring 3.7 × 1.4 cm with the neural placode in the sac is identified.
way to assess TCS as the majority of patients with MMC will have a low-lying conus on MR imaging but no related symptoms. Patients should therefore be followed for evidence of symptoms. Once growth is completed it is likely that fewer patients go on to develop TCS.

**Occult Spinal Dysraphism: Lipomyelomeningocele**

The true incidence of occult spinal dysraphism is unknown, but the incidence is increasing since the advent of MR imaging. Occult spinal dysraphism is often discovered by cutaneous manifestations such as hypertrichosis, capillary hemangioma, dermal sinus tract, subcutaneous lipoma, or an asymmetrical gluteal cleft. Other manifestations can include leg length discrepancy, foot asymmetry/deformity, scoliosis, neurogenic bladder, frequent urinary tract infections, upper and lower motor neuron signs, asymmetrical weakness, gait difficulty, spasticity, and back or leg pain. As availability and sophistication of imaging modalities has improved, occult spinal dysraphism is now often discovered incidentally during workup for other complaints. Lipomyelomeningocele is the most common form of occult spinal dysraphism and consists of a subcutaneous lipoma that is usually located in the lumbar or sacral region and attached to an intradural lipomatous mass (Fig. 7). Cutaneous manifestations are frequent, and while neurological deficits are not uncommon at the time of diagnosis, as many as 48% are normal at presentation (Table 3). The most common manifestation is a subcutaneous mass, followed by skin dimples, hemangioma, hair patches, skin tags, and depigmented regions. Progressive neurological deterioration is very common, and in 1 study 62.5% of patients who presented when < 6 months of age were asymptomatic compared with 29% of those presenting > 6 months. Koyanagi et al. reported that in their patient population, no child was asymptomatic after the age of 5. Retrospective reviews of a large number of patients demonstrated limited recovery if detethering is performed after symptom progression is documented. Kanev et al. in a large retrospective review, noted that urological and bowel dysfunction showed no improvement even with extended follow-up, but motor and sensory symptoms did demonstrate recovery as 10 of 11 patients undergoing operations for retethering returned to preoperative functioning. While asymptomatic presentation is not uncommon, the natural history of lipomyelomeningocele appears to more often demonstrate symptom progression with bladder dysfunction occurring first (often by the age of 2 years), followed by motor/sensory symptoms occurring later, often during the teenage years.

**TABLE 2: Summary of studies of cord retethering after MMC repair**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>% Retethering After MMC Repair</th>
</tr>
</thead>
<tbody>
<tr>
<td>Phuong et al., 2002</td>
<td>1435</td>
<td>2.8</td>
</tr>
<tr>
<td>Bowman et al., 2001</td>
<td>118</td>
<td>32</td>
</tr>
<tr>
<td>Shurtleff et al., 1997</td>
<td>654</td>
<td>13</td>
</tr>
<tr>
<td>Tamaki et al., 1988</td>
<td>60</td>
<td>15</td>
</tr>
<tr>
<td>Bowman et al., 2009</td>
<td>502</td>
<td>23</td>
</tr>
<tr>
<td>Selber &amp; Dias, 1998</td>
<td>46</td>
<td>26</td>
</tr>
</tbody>
</table>
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TABLE 3: Long-term follow-up of patients with lipomyelomeningocele/occult spinal dysraphism tethered cord

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Patient Age Range</th>
<th>No. Asymptomatic</th>
<th>No. Symptomatic</th>
<th>Follow-Up (yrs)</th>
<th>No. Improved/Stabilized</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hoffman et al., 1985</td>
<td>97</td>
<td>6 days to 18 yrs</td>
<td>47</td>
<td>50</td>
<td>22</td>
<td>47/16</td>
</tr>
<tr>
<td>Kanev et al., 1990</td>
<td>80</td>
<td>1 wk to 17 yrs</td>
<td>38</td>
<td>42</td>
<td>35</td>
<td>38/41</td>
</tr>
<tr>
<td>Koyanagi et al., 1997</td>
<td>34</td>
<td>1–47 yrs</td>
<td>8</td>
<td>26</td>
<td>6</td>
<td>7/21</td>
</tr>
</tbody>
</table>

Adult Presentation of TCS

In adults, TCS most often presents with pain, motor or sensory deficits, and sphincter disturbances (Table 4). The cause of such symptoms is often diagnosed with a combination of physical examination findings and imaging demonstrating spina bifida, tumor, or an abnormality of the filum terminale. The adult presentation of TCS encompasses a variety of patients. Previous studies describe categories of adult TCS that are based primarily on the timing of presentation and the presence of dysraphism. Pang and Wilberger described 2 categories: 1) children that were healthy in childhood with symptoms developing later in life, and 2) those with stable deficits and previous diagnoses who were well until the onset of new or progressive deficits in adulthood. More recently, McLone described 3 categories: 1) those who were previously asymptomatic presenting with signs and symptoms of cutaneous stigmata; 2) the true occult group presenting with signs and symptoms and no cutaneous stigmata; and 3) those with a previous diagnosis of spinal dysraphism, with or without a detethering or repair procedure, with stable or progressive deficits. As stated previously, the adult presentation of TCS is similar to that in adolescent patients, with back or leg pain as a predominant feature. The pain is most often non-dermatomal, shocklike, or with a burning quality. Mostly the pain is insidious, but may present with acute onset or aggravation of existing pain. Sensorimotor deficits in the lower extremities occur at a similar rate and sphincter dysfunction is also frequently present. In many patients with adult onset symptoms, cutaneous stigmata are absent or had been previously missed. Both radiographic and surgical data may demonstrate a thickened filum, an intradural lipoma, and fibrous adhesions as the most common tethering lesions. Less common tethering lesions in the adult include split cord malformation and a dermal sinus.

Why do patients develop signs and symptoms in adulthood when the origin of these tethering lesions is often similar to those noted in the younger age groups? Pang and Wilberger identified the degree of cord traction rather than the level of tethering or origin of lesion as the predominant factor related to onset of symptoms. Patients with less severe traction remain asymptomatic in childhood and present with neurological dysfunction later in life. Yamada et al. identified several factors contributing to the onset of symptoms over time in patients with less severe traction, including: 1) increasing fibrosis of the filum leading to progressive loss of viscoelasticity, which results in progressively increased traction in the lumbosacral cord; 2) a growth spurt that could cause a rapid increase in spinal cord tension; 3) an increase in physical activity (sports, exercise); and 4) development of spinal stenosis that can restrict movement and may accentuate tension. Acute onset of symptoms of adult TCS may occur when trauma to the lumbar spine occurs in conjunction with an occult lesion already aggravated by the above-mentioned factors of progression. Natural head and neck flexion as minor trauma over time has been indentified as a contributor to the onset of symptoms in a tethered spinal cord. Similarly, changes in the lumbosacral ligamentous laxity with pregnancy can cause stretching of the conus, as can the act of childbirth in the lithotomy position. Additionally, changes in dimensions of the spinal canal, including a herniated disc or fracture, can induce symptoms. Other authors have noted straight leg exercises, sexual intercourse, and even prolonged sitting as precipitating factors. The understanding of adult TCS is evolving as the diagnosis becomes more prevalent than previously believed. Symptoms of adult TCS may mimic signs and symptoms associated with lumbar degenerative disease. Despite the routine use of MR imaging, there remains a delay in diagnosis. Patients may often be dismissed as having minor degenerative disease causing symptoms or given the diagnosis of failed back syndrome. As a result they may receive physical therapy, traditional conservative measures, or surgical management based on degenerative processes, and TCS may not be addressed as the primary or a related cause of the symptoms due to the subtle, nonspecific findings of TCS as well as deficits that do not correlate with specific myotomal or dermatomal patterns. Little data exists about the natural history of adult TCS.

TABLE 4: Summary of common symptoms in adults presenting with TCS

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Mean Age at Presentation</th>
<th>% w/ Pain</th>
<th>% w/ Motor Weakness</th>
<th>% w/ Bladder Dysfunction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rajpal et al., 2007</td>
<td>61</td>
<td>36</td>
<td>56</td>
<td>79</td>
<td>34</td>
</tr>
<tr>
<td>Lee et al., 2006</td>
<td>59</td>
<td>43</td>
<td>73</td>
<td>78</td>
<td>71</td>
</tr>
<tr>
<td>Huttmann et al., 2001</td>
<td>56</td>
<td>34</td>
<td>77</td>
<td>57</td>
<td>70</td>
</tr>
<tr>
<td>van Leeuwen et al., 2001</td>
<td>57</td>
<td>41</td>
<td>74</td>
<td>44</td>
<td>67</td>
</tr>
<tr>
<td>Pang &amp; Wilberger, 1982</td>
<td>23</td>
<td>39</td>
<td>78</td>
<td>65</td>
<td>56</td>
</tr>
</tbody>
</table>
Generally, the treatment of such patients is extrapolated from data regarding children with TCS. Understanding the pathophysiology of repeated traction and secondary vascular changes suggests that the process of the TCS, whether adult or pediatric, is a progressive one. Thus, the course of clinical presentation or worsening is related to the degree of traction and not necessarily its cause.

The timing of neurosurgical intervention in adult TCS is controversial. This debate parallels the uncertain course of the natural history. Some authors recommend surgical intervention with identification of cutaneous findings of dysraphism with or without neurological deterioration. In an asymptomatic patient with findings of occult spinal dysraphism, it will be difficult to predict if and when there will be a neurological deterioration. For this reason, some authors advocate intervention only if there is neurological deterioration, whereas others suggest intervention if the individual leads an active life and thus may be more susceptible to deterioration related to other factors, such as risk of trauma. In the presence of neurological deterioration, surgical intervention at the time of presentation is advocated.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Mangano, Hertzler, Stevenson. Acquisition of data: Hertzler, Stevenson. Analysis and interpretation of data: DePowell, Stevenson. Drafting the article: Hertzler, DePowell. Critically revising the article: Mangano, DePowell. Reviewed final version of the manuscript and approved it for submission: Mangano.

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