Review and Classification of Occult Spinal Dysraphism and Tethered Cord Syndrome in Children

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Abstract

Tethered cord syndrome (TCS) or occult spinal dysraphism sequence is a collection of neurological conditions that potentially result from the abnormal fixation of the spinal cord secondary to a developmentally acquired or post-operative pathology. We present a schema for TCS consisting of etiology, embryology, pathophysiology, presentation, and classification in order to facilitate the comprehension and discussion of this complex topic. Our review focuses on closed rather than open spinal dysraphisms because those cases are generally more inconspicuous in presentation and, for that reason, likely to create diagnostic delays that can adversely affect patients.

Keywords: Tethered cord syndrome; Occult spinal dysraphism; Classification; Urodynamics; Pediatric neurosurgery

Abbreviations: TCS: Tethered Cord Syndrome; OSD: Occult Spinal Dysraphism; POD: Postovulatory Day; CCM: Caudal Cell Mass; UDS: Urodynamic Studies; SCM: Split Cord Malformation; MMC: Myelomeningocele; LMMC: Lipomyelomeningocele

Introduction

Tethered cord syndrome (TCS) in children is an entity described as an array of congenital anomalies, including cutaneous, urologic, neurologic, and orthopedic systems. It is thought to result from the abnormal fixation of the distal spinal cord secondary to a developmentally acquired or post-operative pathology. Unlike spina bifida aperta (i.e., myelomeningocele), which is readily diagnosed prenatally, spina bifida occulta often manifests more insidiously. As a result, these closed defects are usually discovered following symptom onset or incidentally during an unrelated work up of coincident comorbidities. Because the prognosis of this syndrome is highly dependent on symptom duration, it is important that all disciplines potentially involved (urology, orthopedics, dermatology, pediatricians, etc.) be aware of this condition as delays in diagnosis can have serious long-term effects. The objective of this paper is to review key aspects of tethered cord syndrome and offer up a classification schema to facilitate recognition and understanding amongst neurological surgeons as well as pediatricians and the surgical community at large.

Tethered Cord Syndrome: Then and Now

As early as the mid-19th century, there were descriptions of spinal cord tethering and related symptomatology. Johnson, in 1857, discusses a fatty sacral tumor connected with spinal membranes in a child. In 1891, in England, Jones performed the first successful intervention for tethered cord. In 1910, Fuchs observed incontinence as a pediatrician and the surgical community at large.

Radiographically, a low-lying cord in TCS refers to a conus sitting anatomically lower than the L2 vertebral body. TCS signs and symptoms result from damage the spinal cord endures while it is under traction (see Pathophysiology below). Several authors have attempted to determine what constitutes a “normal” conus level. Their findings vary from the T12 to the inferior aspect of L2, with the most common termination at or above L1/L2 [4-8]. For the remainder of this paper, normal conus will be considered one that terminates at or above the L2 vertebral body.

Embryology

TCS often associated with disorders that result from the abnormal development of the central nervous system. Because various TCS etiologies can be attributed to defects occurring at different stages of fetal spinal cord development, understanding the nervous system’s embryology is crucial to the comprehension and recognition of TCS.

The spinal cord forms as the result of two distinct processes: primary and secondary neurulation. Primary neurulation entails the proliferation and folding of neuroectoderm into a neural tube that ultimately comprises the spinal cord. This process begins on postovulatory day (POD) 18; the notochord induces the overlying ectoderm to proliferate as neuroectoderm, forming a groove that progressively elevates until it fuses and forms the neural tube. Cutaneous ectoderm (which eventually becomes skin) separates from the neuroectoderm and fuses on the midline during a critical process called “disjunction.” The mesoderm forms the posterior bony and soft tissue elements. Disruption of this stage is responsible for many spinal cord pathologies, including, myelomeningocele (nondisjunction), lipomyelomeningocele (premature disjunction), and dural sinus tract (incomplete disjunction). Closure of the neural tube begins around POD 22 at the site of future cervical levels; it precedes both rostrally and caudally. Closure of the rostral and caudal neuropore occurs by POD 26 and POD 28, respectively. The formation of the brain and the spinal cord mark the end of primary neurulation. Developmental failure of the primary neurulation process may result in an open neural tube defect.

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Secondary neurulation refers to the formation of distal spinal elements caudal to S2 as well as the filum. This phase of development occurs between POD 28 and 48. Neuroectoderm caudal to the posterior neuropore, also known as the “caudal cell mass (CCM),” begins canalization. During this process, the vacuoles that form in the middle of the CCM coalesce with the vacuoles located in the neural tube’s central canal [9]. Subsequently, disproportionate growth rates between the spinal cord and the vertebral column cause the spinal cord to ascend and pull away from its sacral attachments. The cauda equina forms as nerve roots elongate to accommodate the differential growth. This process of retrogressive differentiation continues until the conus reach the adult level by three months of age. Errors that occur during canalization or regression are thought to contribute to the formation of low lying conus, terminal lipomas/myelocystoceles, and fatty filum pathology [9-11].

Pathophysiology

One of the first valuable pathological descriptions of tethered cord syndrome was published in 1982 by Pang and Wilberger [6]. The authors report that the degree of traction on the conus determines the age of symptomatic onset. If the traction is significant, it will cause symptoms to manifest in early childhood. If it is less severe, the patient will remain sub-clinically asymptomatic. Symptoms might manifest later in life due to additional stretching of the conus caused by growth spurts or precipitating events, such as strenuous exercise, pregnancy, childbirth, or trauma. Around 60% of adult TCS patients experience these types of events immediately before symptomatic onset. Because these events aggravate developmental errors that occurred much earlier in life, TCS is still considered a congenital anomaly even when it manifests during adulthood.

Our understanding of TCS pathophysiology has primarily been built upon the work conducted by Yamada et al. over the last 30 years [10]. The authors postulated that progressive low to moderate traction placed on the filum causes a reduction of cytochrome a, a3, which indicates an ischemic state. Using animal models, Yamada’s group showed that the degree of caudal traction on the spinal cord correlated with the severity of neurological deficit secondary to the impairment of oxidative metabolism [12]. They also demonstrated a proportional reduction in spinal cord blood flow in relation to the force of traction, which they designated as “traction induced hypoxia” [12]. Their model also showed how chronic tension can preload the cord in such a way that even minor additional traction can cause severe, permanent damage [13]. The authors demonstrated that many of these changes were completely reversible after persistent low to moderate traction was alleviated, but an application of severe traction still produced irreversible damage to the spinal cord. This damage was most likely caused by traction-induced mitochondrial dysfunction and cell death. Thus, there is an apparent threshold to persistent traction from which the spinal cord cannot recover. A study conducted by Steettel et al. showed that the tethering of the filum terminale caused a reduction in the blood flow to the spinal cord, leading to tissue hypoxia as a result of mitochondrial redox dysfunction [14]. Metabolic derangements were corrected following restoration of blood flow; however, if blood flow was interrupted for longer periods of time, recovery was only partial. Under these circumstances, it appears that excessive tension can cause irreversible changes and permanent dysfunction that will not be restored following tethered cord release.

Diagnosis

Clinical presentation

In the setting of Occult Spinal Dysraphism (OSD), a myriad of symptoms can suggest the presence of TCS in children. Thorough examinations are warranted for patients who demonstrate even the most subtle of findings associated with the condition. Common presenting signs and symptoms include cutaneous signatures associated with OSD (59%), neurogenic bladder (18%), lower extremity weakness, numbness, or spasticity (12%), leg or foot discrepancy (6%), foot deformity, spinal deformity, and non-dermatomal leg/back pain (6%) [15]. While children often present with a combination of findings, symptoms can also be isolated to one system. This diverse presentation is one of the reasons why it is so crucial for physicians to acquaint themselves with the clinical picture of TCS. The diagnosis of various syndromes should also encourage physicians to evaluate patients for OSD, as the two are often associated with each other.

Significant cutaneous lesions can be seen in up to 3% of the general population; in patients with OSD, the incidence approaches 80% and there is a greater chance that multiple lesions will be detected upon careful examination [11,16,17]. At times, these findings may be the only symptoms indicating underlying dysraphism. Cutaneous discrepancies include midline hairy patches, hemangiomas, dermal pits/sinuses, hypertrichosis, subcutaneous lipoma, “cigarette burns,” lumbarosacral appendage, and nevi. While the appearance of any of the aforementioned findings is sufficient to warrant investigation, recent work spanning 12 years of pediatric patients suggests that an isolated sacral dimple in an otherwise asymptomatic child has a significantly low association with tethered cord syndrome; the incidence of necessary surgical de-tethering in that population ranged from only 0.13 to 0.17% [18-20]. That being said, ultrasound is simpler to perform and easier to obtain than an MRI at a later point when the child may already have become symptomatic and will also require sedation.

Urologic dysfunction is most commonly the result of initial derangement in OSD-related tethering. Patients encounter problems that range from blatant incontinence to subtle changes observed during urodynamic studies. Presentation may include incontinence, urinary urgency, increased urinary frequency, and recurrent UTIs; in the pediatric population, these symptoms tend to be more subtle than other clinical findings [14]. Because bladder dysfunction is difficult to assess in infants, these problems may not even become apparent until children are much older. The most common bladder symptom among toddlers is delayed or unsuccessful toilet training; during testing, detrusor hyperreflexia is the most common finding. Because a disruption in urodynamics often proceeds clinical symptoms, this highlights the importance of urological work to aid in preventing delayed diagnosis and treatment. Besides detrusor hyperreflexia, other common symptoms include diminished bladder compliance, external detrusor-sphincter dyssynergia, decreased sensation, and hypocontractile detrusor function [21,22].

Neurological problems that manifest in TCS involve the disruption of the motor and sensory pathways of the lower extremities. Although they comprise elements of upper and lower motor neuron dysfunction, motor deficits are more prevalent than sensory problems [14]. However, presentation in toddlers and children is commonly associated with both motor and sensory dysfunction [1]. Neurologic disturbances that may be diagnosed include delayed gait development, hyper/hyporeflexia, muscular atrophy, and spasticity. Often times, the abnormalities are asymmetric. In a series conducted by Bui et al. 46% of children demonstrated changes in strength, tone, or reflexes [15]. Sensory deficits, if present, are in the feet or perineum or children may present with painless ulcerations of the feet/legs. Pain may also present as a neurological symptom; however, this is much less common in pediatric cases compared to adult cases (see below).
Orthopedic abnormalities are found in more than 90% of patients with TCS [14]. Among children, foot deformities are most common. These deformities most likely result from neuromuscular imbalance at a time when bones are growing and aligning; deformities are unlikely to arise later in life if mal-alignment does not occur during this period. Other abnormalities include limb length discrepancies, gluteal asymmetry, vertebral anomalies, and scoliosis. Progressive scoliosis or kyphosis can be seen in about 25% of children with TCS and may also contribute to complaints regarding pain.

Vertebral anomalies are commonly observed in children with TCS. These include bifiid vertebrae, laminar anomalies, hemivertebrae and sacral agenesis. Segmentation errors may be multiple and these bony abnormalities can be observed in approximately 95% of children with TCS [11].

It is well established that TCS is often associated with other congenital syndromes; as such, patients with these conditions should be screened for OSD/TCS. The incidence of OSD ranges from 15% to 60% in those with VACTERL association (vertebral anomalies, anal atresia, cardiac anomalies, tracheoesophageal fistula, renal and limb anomalies, most often radius) [23,24]. Two of the most common associations are caudal agenesis and anorectal atresia syndromes: OEIS (omphalocole, cloacal extrophy, imperforate anus, and spinal anomalies), VACTERL, VATER (vertebral anomalies, anal imperforation, TE fistula, renal radial anomalies) and Currarino/ASP triad (anorectal malformation or congenital anorectal stenosis, sacrococcygeal osseous defect, presacral mass). Miller-Dieker syndrome, a lissencephalic condition, has been reported to demonstrate symptomatic tethered cord pathology by way of a thickened filum and dermal sinus tracts [25,26]. An association between Chiari 1 malformation and tethered cord syndrome has also been described [27,28]. Because of this connection, the presence of the

| Common Presenting Signs and Symptoms in OSD/TCS          | Older Children/Adults | Pediatric         |
|---------------------------------------------------------|------------------------|-------------------|
| Cutaneous                                               |                        |                   |
| Hemangiomas                                             |                        |                   |
| Dermal pits/sinuses                                     |                        |                   |
| Hypertrichosis                                           |                        |                   |
| Subcutaneous lipoma                                      |                        |                   |
| “Cigarette burns”                                        |                        |                   |
| Lumbosacral appendage(s)                                 |                        |                   |
| Nevi                                                     |                        |                   |
| Neurologic                                               |                        |                   |
| Upper motor neuron signs: hyperreflexia, spasticity, etc. |                        |                   |
| Lower motor neuron signs: hyporeflexia, muscular atrophy, etc. |                |                   |
| Mixed upper and lower motor neuron signs                 |                        |                   |
| Feet/perineal sensory loss                               |                        |                   |
| Back/leg pain                                            |                        |                   |
| Gait difficult/Delayed ambulation                        |                        |                   |
| Urologic                                                 |                        |                   |
| Detrusor hyperreflexia                                   |                        |                   |
| Frequent urinary tract infections                        |                        |                   |
| Incontinence                                             |                        |                   |
| Delayed toilet training                                  |                        |                   |
| Orthopedic                                               |                        |                   |
| Foot deformities                                         |                        |                   |
| Limb length discrepancies                                |                        |                   |
| Gluteal asymmetry                                        |                        |                   |
| Vertebral anomalies                                      |                        |                   |
| Scoliosis                                                |                        |                   |
| Vertebral                                                |                        |                   |
| Bifid vertebrae                                          |                        |                   |
| Hemivertebrae                                            |                        |                   |
| Laminar defects                                          |                        |                   |
| Sacral aplasia                                           |                        |                   |
| Sacral agenesis                                          |                        |                   |

**Table 1:** Common presentation of tethered cord syndrome.

By studying the differences in TCS presentation that exist between various age groups, physicians will be better equipped to recognize any relevant symptoms and diagnose patients. In neonates and infants, tethering is often evidenced only by cutaneous manifestations of OSD. The presence of anorectal malformation is highly suspicious for a tethering lesion and warrants investigation. Toddlers and adolescents tend to present with motor and/or sensory deficits or bladder control regression. Teenage children and adults commonly present with severe pain. They usually describe it as a diffuse pain affecting the legs, groin, or perineum, but they might also characterize it as an electrical shock-like pain that travels along the spine.

Pain is an infrequent complaint among children. When reported, it is usually localized in the lower back region without radiation into the legs; sometimes it worsens with prolonged bed rest. The presentation of pain in children is often difficult to identify because it may be confused with general irritability or a temper tantrum. Long tract signs are rare in children with TCS but commonly encountered in adults. Barry et al. demonstrated that ischemic damage to large-diameter corticospinal fibers increases with the duration of tethering, which one reason why such findings are more common in the adult population [29]. With regard to orthopedic abnormalities, if an individual reaches adulthood without foot deformities or scoliosis, it is theorized that musculoskeletal development was successful at a younger age and the adult patient will not develop these conditions or experience the onset of related symptoms. Table 2 summarizes the contrasting presentation characteristics seen in pediatric and adult patients with TCS.

**Radiographic and urodynamic studies**

Diagnosis of TCS requires the correlation of clinical symptoms with relevant radiographic findings. Presently, to our knowledge, there has never been a case of TCS that was reported with normal imaging. Among the various radiographic procedures available, plain X-rays have the most limited application; they are primarily used to follow the progression of scoliosis. Ultrasound is ideal for infants because there is no need for radiation or sedation. It is also reported to have 96% sensitivity and 96% specificity [18]. That being said, ultrasound is limited by operator abilities and often difficult to interpret. Additionally, its use is restricted to infants 4-6 months old because spine ossification reduces the reliability of ultrasound findings [18,30]. It can, however, function as a screening tool. If ultrasound results are normal in the setting of sacral dimplies or isolated strawberry hemangioma, then the probability of TCS is relatively low and MRI studies can be postponed [31].

MRI is the imaging procedure of choice for the assessment of OSD/TCS. T1-weighted imaging provides clear anatomical detail of neural tissue and the filum. This enables visualization of vertebral levels, the conus position, and the presence of fat/thickening/syrinx.
Sagittal views allow for level localization while axial views display fat and the diameter of the filum. As mentioned before, a filum below the L2 vertebral body or thicker than 2 mm is considered abnormal in children. The absence of movement between supine and prone MR imaging would also indicate a diagnosis of TCS [1]. MR imaging can also reveal urologic sequelae, such as a distended bladder. T2-weighted imaging permits the identification of spinal cord tumors and fluid-containing structures. Complete imaging of the entire neuroaxis is important for the screening of “skip lesions” or other abnormalities that are often observed in association with TCS (i.e., MMC, SCM, dermal and lipomatous tumors, etc.).

Using only medical history and examination to determine the cause of bladder dysfunction in children can be problematic without the assistance of urodynamic studies (UDS). Because TCS symptom reversal is associated with the duration of dysfunction, patients will have a better chance for successful outcomes if urodynamic studies are implemented early to help establish a definitive diagnosis. UDS can also indicate clinical deterioration and provide a way for physicians to monitor patient improvement following a detethering procedure. For these reasons, physicians should obtain UDS prior to and following any surgical procedure. The most common UDS finding is detrusor hyperreflexia. The following metrics may also be identified: decreased bladder compliance, dysynergia, and decreased sensation. Important aspects of bladder function assessment include: bladder capacity, bladder pressure, leak point pressure, compliance, uninhibited contractions, EMG activity, and sensation [10].

**Classification of TCS Presentation**

Because there are several distinct tethered cord presentations, it is important to know how to differentiate and classify them based on natural history, comorbidities, severity, and progression. As recently at the early 20th century it was the tendency for untreated TCS to lead to progressive neurological decline; however the natural history of tethering lesions depends in part on the cause of cord tethering [2]. Van Leeuwen et al. suggested a tethered cord classification based on the origin of tethering with four main groups: 1) Post myelomeningocele (MMC) repair 2) Fatty/tight filum terminale 3) Lipomyelomeningocele (LMMC)/conus lipoma 4) Split cord malformation (SCM) [32]. This classification schema is outlined in Table 3 and Figure 1. Because the more subtle abnormalities (2-4) are skin covered, they often get grouped together in the literature. This conflation of categories creates confusion for readers and oversimplifies these processes. Our goal is to describe a variation of this classification system that focuses on tethering pathology to elucidate the subtle distinctions between these conditions. Appreciating these differences is imperative for physicians who wish to expedite workup, diagnosis and treatment.

The first group in Leeuwen’s model describes TCS following myelomeningocele repair. These patients present from birth with open NTD and consequential neurological deficits (motor/sensory/bowel/bladder) dependent on the level of the defect. Chiari 2 malformation with or without symptomatic hydrocephalus will also be present. Because of scarring created by the untethering of the spinal cord...
during surgical repair, this population is at risk for future tethering with resulting neurological decline. Recognition of prior repair is commonly obtained from patient history or physical exam; however, progression of urological dysfunction will often be the first indication of retethering [33]. If left untreated, the natural history of this process has demonstrated symptomatic progression in up to 60% of patients in the first five years [34]. In another study, approximately 25% of patients required an additional untethering procedure for symptoms related to retethering. Sixty-four percent of patients showed improvements in urological evaluations, 96% had improved or stable scoliosis, and 70% demonstrated increased lower extremity strength [35]. Of utmost significance is the understanding that once overt urologic dysfunction occurs in this population following retethering, it is less likely to be recovered, which further emphasizes the importance of early diagnosis.

Group two is comprised of patients with a fatty/tight filum terminale (filum lipoma). This form of OSD results from fat infiltrating the filum during the retrogressive differentiation stage of secondary neurulation. A fatty filum is defined as being >2 mm with or without a low-lying conus. Some authors describe a condition as “asymptomatic occult tight filum” despite nondiagnostic imaging. We have chosen not to include this form of OSD in our classification schema because of its controversial nature.

Patients with this type of OSD often demonstrate other caudal developmental abnormalities, such as VATER and Currarino/APS [33]. While the true incidence of Group 2 OSD among these patients is unknown, authors have published studies in which they report up to 46% for a given series; furthermore, a tight filum is implicated in up to a fourth of all lesions causing TCS [9]. Clinically speaking, this condition is less severe and often presents without classic cutaneous markers or neurologic/urologic symptoms. Patients are often older; their complaints more frequently involve pain as opposed to neurological dysfunction. That being said, urinary dysfunction with pain is observed as a common presentation in this group. Symptom onset is thought to be dependent on the degree and duration of traction placed on the spinal cord; severe traction sustained over longer periods of time results in more severe and potentially irreversible neurological symptoms. A precipitating event (exercise, trauma, childbirth, etc.) can often be identified among older patients diagnosed with this more insidious form of OSD.

Patients with lipomyelomeningocele (lipoma of the conus medullaris) constitute the third group of OSD that can cause tethering of the spinal cord. This entity results from a premature disjunction that permits the migration of the mesodermal elements that form fatty tissue. The defect is closed (skin covered) as ectodermal fusion has occurred prior to formation. With an incidence estimated at 1:400 and a female to male ratio of 2:1, lipomyelomeningocele is considered the most common type of spinal lipoma [9,33]. Because it is characterized by cutaneous markers, diagnosis is typically made in infancy; up to 90% of patients present with a non-tender, subcutaneous fatty mass [29,33]. From a neurological/urological perspective, the natural history of these lesions is progressive neurological deterioration in the form of urologic dysfunction. In a prospective study, 50% of patients were and remained asymptomatic since birth; however, 25% of those asymptomatic at birth demonstrated progressive onset of neurologic symptoms within 5.5 years [36]. Another series demonstrated the progression of neurological symptoms in 60% of patients who were formerly asymptomatic [11]. Overall, 70% of lipomyelomeningocele patients require surgical intervention. The most common initial neurologic manifestation among lipomyelomeningocele patients is bladder dysfunction, which can be seen in up to 60% of this population [1]. Oftentimes, urologic abnormalities can only be diagnosed using urodynamic studies [33]. Because dysfunction has been shown to correlate with patient age, older children and adults are more likely to present with irreversible urological findings [37]. This condition’s inclination toward progressive decline and potentially irreversible neurological dysfunction make it crucial for physicians to establish an early diagnosis.

The final subgroup of TCS consists of patients with split cord malformation/diastematomyelia. This congenital anomaly accounts for 25% of OSD and results from problems that occur before primary neurulation, during gastrulation. During development, adhesions between ectoderm and endoderm lead to the formation of a mesenchymal tract that bisects the spinal cord [38]. Tethering occurs at the level of the bisecting bony spur/dorsal band as well as a fatty/thickened filum. Clinically, cutaneous stigmata, more specifically a tuft of lumbosacral hair, are also commonly associated with SCM. Orthopedic anomalies are also seen, with 85% of patients demonstrating bony abnormalities and 50% presenting with scoliosis. One unique aspect to SCM is the prevalence of tandem neurodevelopmental lesions, including fatty filum, lipomyelomeningocele, myelomeningocele, meningocele manqué, and chiari malformation. Fifty to eighty-five percent of patient with SCM will harbor a secondary abnormality. Like other etiologies of TCS, SCM patients are prone to developing urological abnormalities, which one study suggests will be seen in 75% of cases [39]. Because urologic dysfunction is not commonly diagnosed with clinical symptoms, we must once again highlight the important role formal urological evaluation plays in expediting a definitive diagnosis.

Conclusion

The prompt identification of TCS still represents a major clinical challenge. Some of the obstacles contributing to that challenge are symptom ambiguity, presentation discrepancy, and the absence of a standardized pathology accounting for the majority of OSD cases resulting in pediatric TCS. These factors are particularly problematic with respect to timely diagnosis and treatment of children. Because the duration of tethering is a key determinant in disability and recovery, it is paramount that physicians from various specialties establish a uniform understanding of the physiology, presentation, classification, and treatment of OSD. Additionally, given that presentation of OSD can be limited to subtle changes in urodynamic studies, it is imperative that a dedicated multidisciplinary team of specialists (including pediatric urologists, pediatric neurosurgeons, pediatric orthopedic surgeons, and physicists) evaluate patients with suspected TCS. We support a simple classification system that facilitates the identification of occult TCS pathologies in an effort to maximize the chance of intervention and minimize the appearance of severe, permanent neurological sequelae. While the true incidence of OSD is unknown, advanced imaging, a greater clinical awareness, and ongoing relevant publications assist in making this condition a more mainstream diagnosis among primary care physicians.

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