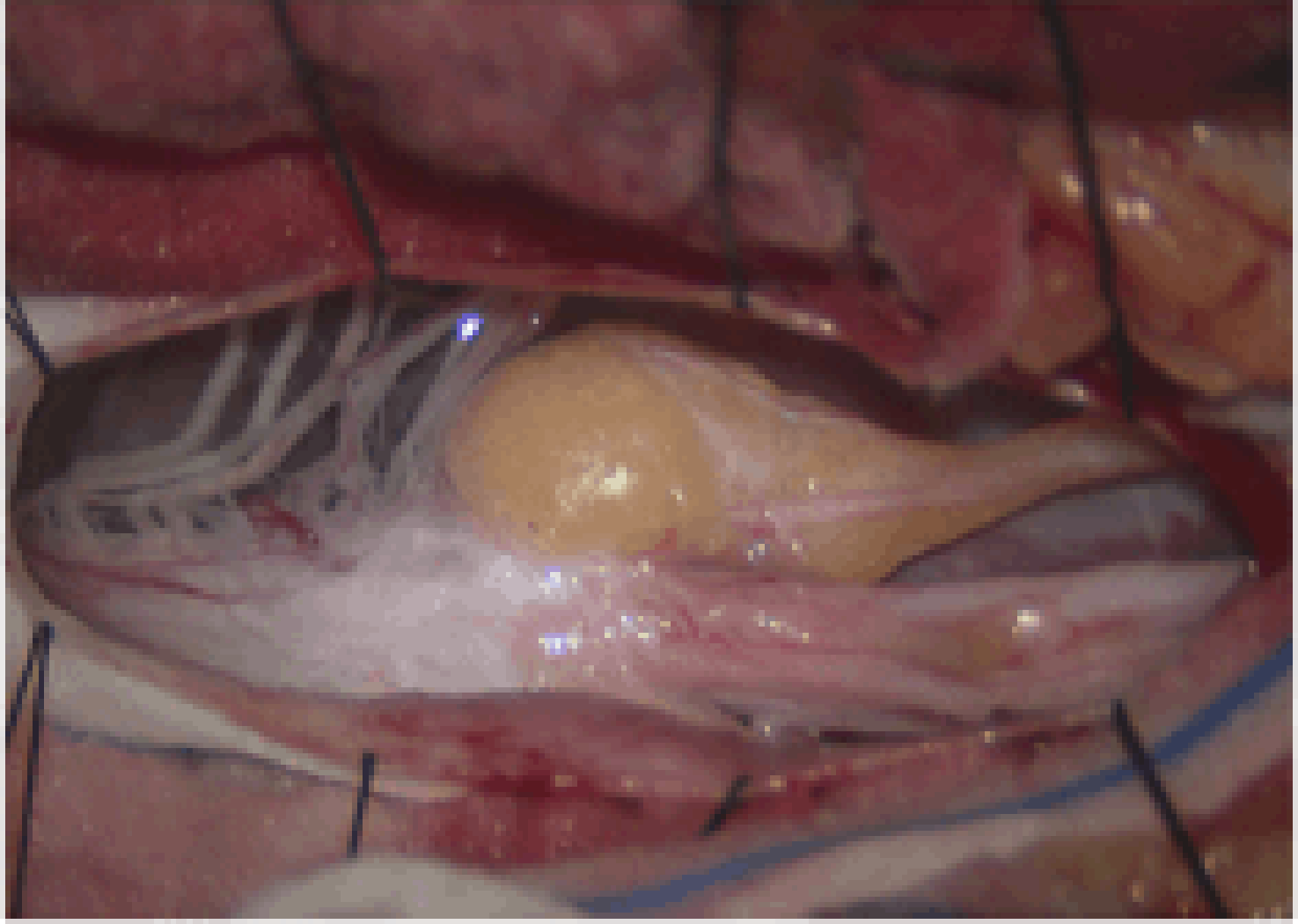

Introduction

Tethered cord

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For over 100 years physicians have recognized an association between spinal dysraphism and neurological deterioration. In the last 20–40 years, surgeons have begun to elucidate the pathophysiology involving the tethered cord. Despite the relative plasticity of the human nervous system, the spinal cord has a limited ability to accommodate acute and chronic tension. With a better understanding of the pathophysiology, surgeons are beginning to develop novel treatment strategies for the tethered cord.

This issue of *Neurosurgical Focus* presents several timely reviews of the history, embryology and etiology, and the pathophysiology and treatment of the tethered cord, 11 articles in total. Highlights include a review article with a historical perspective and analysis of neural embryology by Hertzler and colleagues. The Barrow Neurological Institute and University of Michigan neurosurgical groups provide an appraisal of the pathophysiology of pediatric and adult tethered cord syndrome.

Pouratian and colleagues report on electrophysiologically guided untethering of the spinal cord after perinatal repair of a myelomeningocele. The authors review their surgical technique with the identification of an "autonomous placode" in 6 patients. The University of Washington group provides a unique case report on human motor evoked potential changes following spinal cord transection. The spinal cord was transected in 4 quadrants, and motor evoked potentials were lost unilaterally as each anterior quadrant was sectioned.

Mummaneni's group, from the University of California, San Francisco, is pioneering minimally invasive tethered cord release. They provide a comparison of the latter with the mini-open approach. Their critical assessment of the minimally invasive approach is refreshing. Shih and colleagues provide a review of the current management of recurrent adult tethered cord syndrome from detethering to vertebral column shortening. Hsieh and colleagues analyze the clinical outcomes obtained in patients reported on in the literature and who underwent a posterior vertebral column subtraction osteotomy for tethered cord syndrome. The discussion includes a comparison of the outcomes following detethering and vertebral column shortening.

A fascinating report from Samdani and colleagues calls into question the need to perform a spinal cord untethering procedure in cases involving myelomeningocele in which patients are undergoing scoliosis corrective surgery and who do not present with the clinical symptoms of a tethered cord, even though there is radiographic evidence of tethering. The issue also includes a review by Shin and Krishnaney on the rare tethred cord secondary to idiopathic ventral spinal cord herniation and a case report by Gupta, Heary, and Michaels showing reversal of long-standing neurological deficits after the late release of a tethered spinal cord.

We hope that you enjoy this issue of *Neurosurgical Focus* devoted to the tethered cord. *(DOI: 10.3171/2010.7.FOCUS.Intro)*

Neurosurg Focus 29 (1):E1, 2010

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, dermal 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**

Classically TCS has been defined as a spectrum of congenital anomalies resulting in an abnormally low position of the conus medullaris that may lead to neurological, musculoskeletal, urological, or gastrointestinal abnormalities.[1,13,15,28,45] One of the first modern published reports of TCS was written by Fuchs in 1910,[7] who noted incontinence upon flexion in patients with MMC and inferred that this symptom was a result of tension on the spinal cord. Lichtenstein furthered this theory in 1940[27] by linking spinal cord dysfunction and tethering lesions. Garceau in 1953[8] coined the term "filum terminale syndrome" after observing 3 patients with progressive spinal deformity and neurological symtoms. He surmised that it was due to a thickened filum terminale, which he sectioned. The term "tethered spinal cord" originated from the article in 1976 by Hoffman et al.,[15] wherein the authors described 31 patients with elongated cords whose symptoms improved following the sectioning of the filum terminale. Most commonly TCS is related to spinal dysraphism. The signs and symptoms correlate with the radiological definition in which the conus medullaris is anatomically lower than the L-2 vertebra or below the L1–2 disc space.[3,31] More recently, there have been descriptions of TCS in which patients are described to have the conus medullaris in a normal position on imaging but presenting with signs and symptoms consistent with 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.[44]

Abbreviations used in this paper: MMC = myelomeningocele; TCS = tethered cord syndrome.

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 involutes to form the neural folds and then closes to form the neural tube.[6,24] The process of neural tube closure begins by Day 22–23 and extends cephalad and caudad with the posterior 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

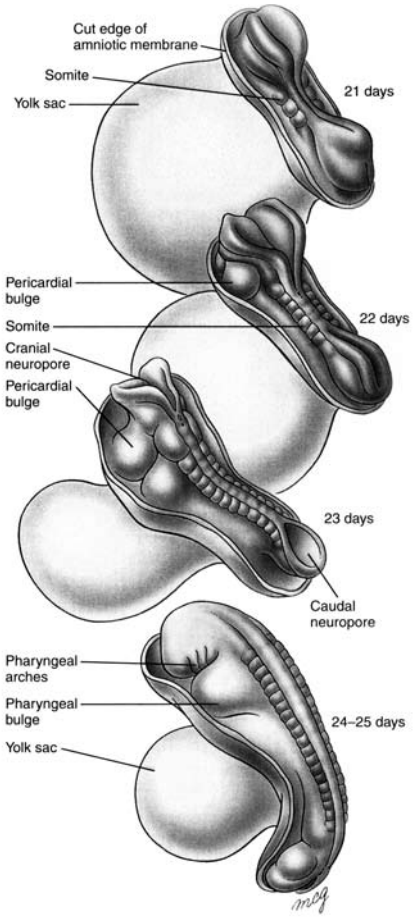

Fig. 1. Artist's illustration of primary neurulation. The lateral edges of the neural folds meet in the midline and fuse. Reproduced with permission from Larsen WJ: **Human Embryology**, 3rd ed., Elsevier Science, 2001.

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 Anson[37] 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, Barson[3] 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

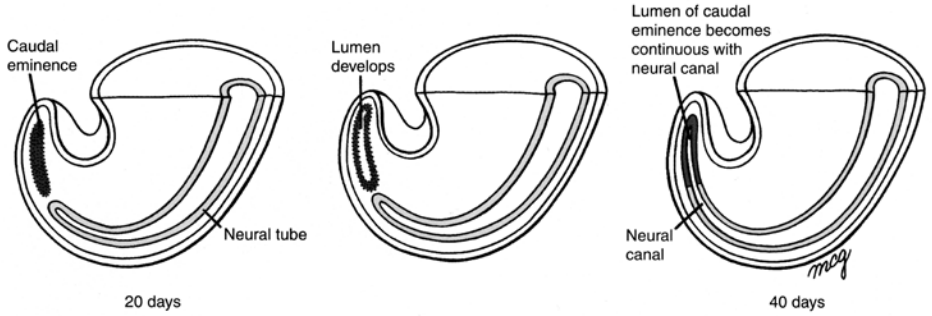

Fig. 2. Artist's illustration of formation of the neural tube inferior to the second sacral level by secondary neurulation. Mesoderm invading this region during gastrulation condenses into a solid rod called the caudal eminence, which later develops a lumen. At the end of the 6th week, this structure fuses with the neural tube. Reproduced with permission from Larsen WJ: **Human Embryology**, 3rd ed., Elsevier Science, 2001.

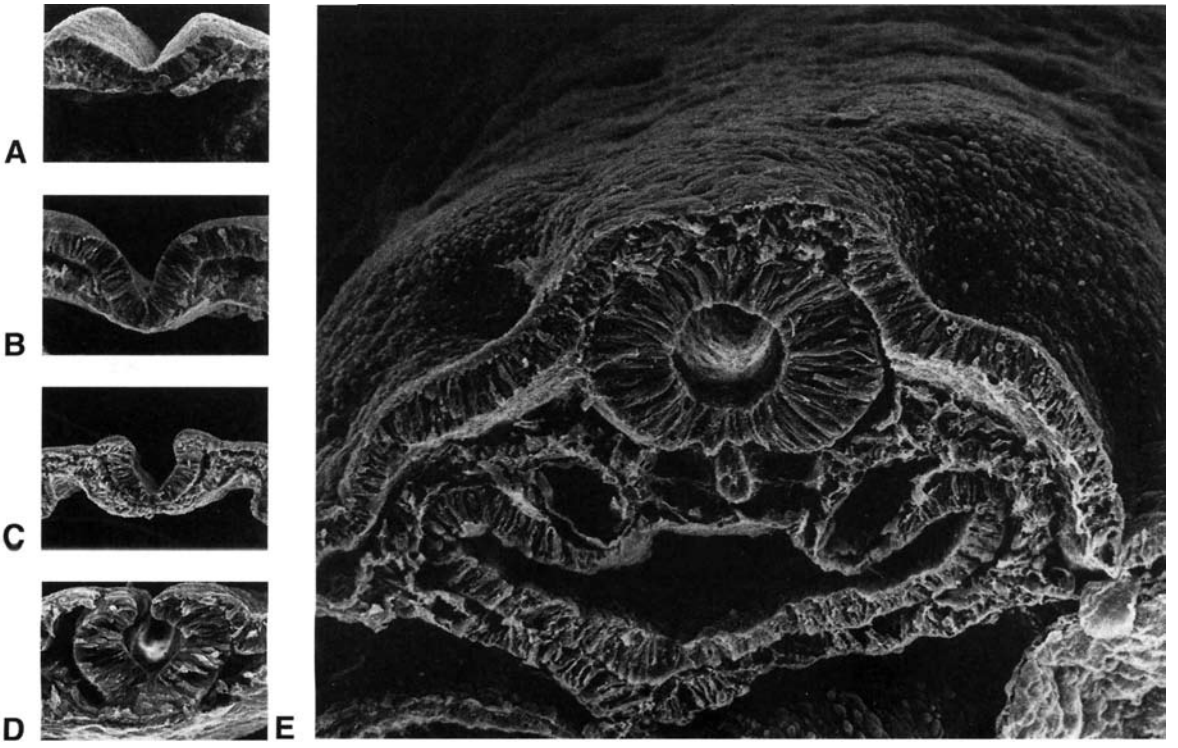

Fig. 3. Images showing neurulation. **A–C:** Neurulation begins in the occipitocervical region as the neural plate is thrown into the neural folds. **D:** The lateral edges of the neural folds meet in the midline and fuse while simultaneously detaching from the surface ectoderm. **D and E:** These detaching edges of the surface ectoderm then fuse with each other to enclose the neural tube completely. Reproduced with permission from Larsen WJ: **Human Embryology**, 3rd ed., Elsevier Science, 2001.

regression in motor function or bladder control or an arrest in achievement of developmental milestones may reveal important cues,[32] including gait difficulties, sensory deficits, development or progression of scoliosis, or foot deformities.[4,21,33] Sensory loss tends to occur in a nonsegmental distribution. Back and lower-extremity pain may be a presenting complaint in this age group.[26,31] 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.[4,26,31] In the adult with a known history of spina bifida, the clinical presentation is similar to that in adolescents with exacerbations of pain and sphincter dysfunction related to flexion and extension movements of the lumbosacral spine.[14] 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.[36] In the subset of adults who present without history of spina bifida, pain is the most common presentation, followed by weakness and urological dysfunction.[12,19,36] 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.[26,31]

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

Authors & Year	No. of Cadavers	Level of Conus Termination
Reimann & Anson, 1944	129	at or above L-2 vertebra in 95.5%
Barson, 1970	252 (infants)	above L1–2 disc space
Pinto et al., 2002	41	at or above L-2 vertebra in 95%

* All cadaveric studies.

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.[30,46] 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

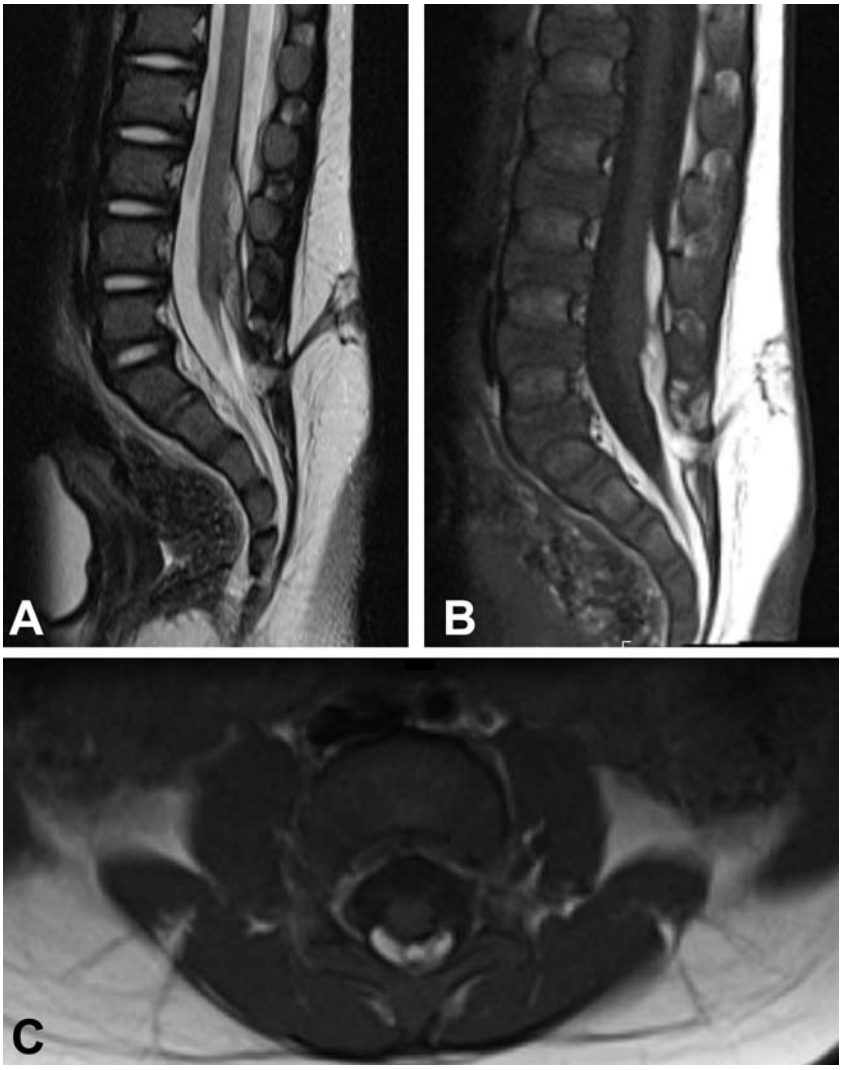

Fig. 4. Spinal MR imaging of a 5-month-old patient with a dermal sinus tract and intradural lipoma. **A:** Sagittal T2-weighted sequence demonstrating the conus medullaris at L-5 and the dermal sinus tract. **B:** Sagittal T1-weighted sequence shows the intradural lipoma and connection to the dermal sinus tract at the S1–2 level. The intradural lipoma is seen extending from L-3 to S-2. **C:** Axial T1-weighted sequence demonstrates the intradural lipoma.

clinical manifestations of deterioration.[43] 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.[10,11,30] 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.[3,34] Thickness of the filum terminale greater than 2 mm is considered abnormal in children, although this finding remains controversial.[34,50] 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

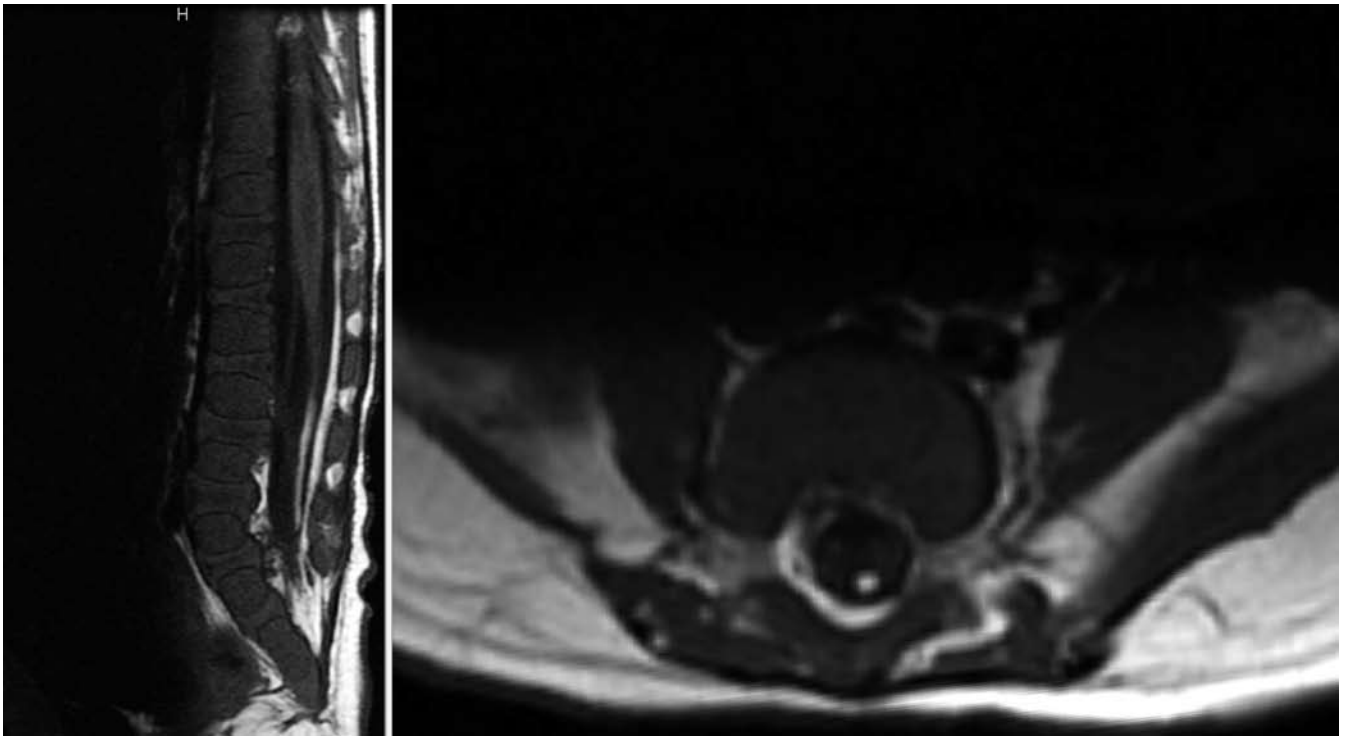

Fig. 5. Sagittal **(A)** and axial **(B)** T1-weighted MR images of 5-month-old patient. The conus medullaris terminates at the mid L-3 vertebral body, with marked thickening and fatty infiltration of the filum. The sagittal image demonstrates a syrinx that extends into the lower thoracic cord.

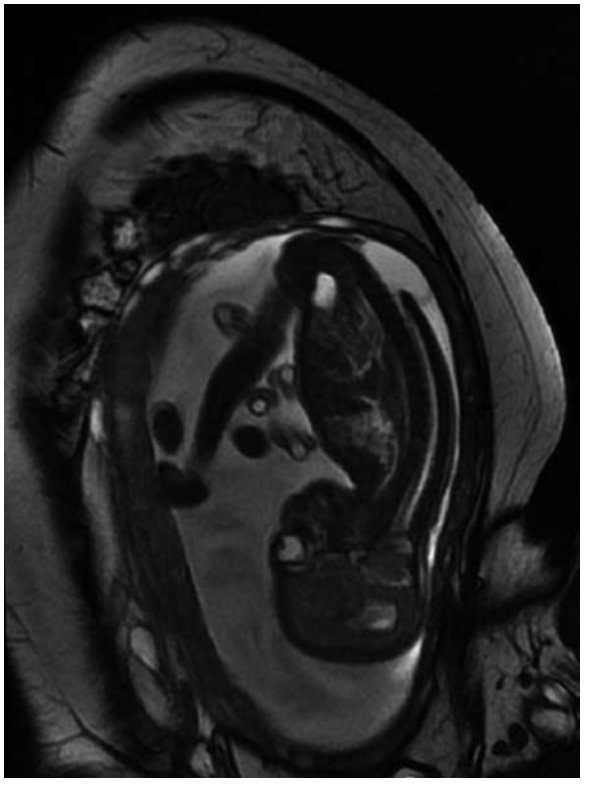

Fig. 6. Sagittal intrauterine MR imaging of a 22-week, 5-day-old fetus demonstrating a neural tube defect inferior to L-3. There is an absence of the posterior elements in the lumbar area. A large dorsal MMC measuring 3.7 × 1.4 cm with the neural placode in the sac is identified.

many as 2.8–32% of patients with MMC as a result of retethering as the vertebral column grows and lengthens (Table 2).[4,5,32,38,39,41] 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.[32,40] Phuong et al.[32] 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[9] 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[38] 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.[29] 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

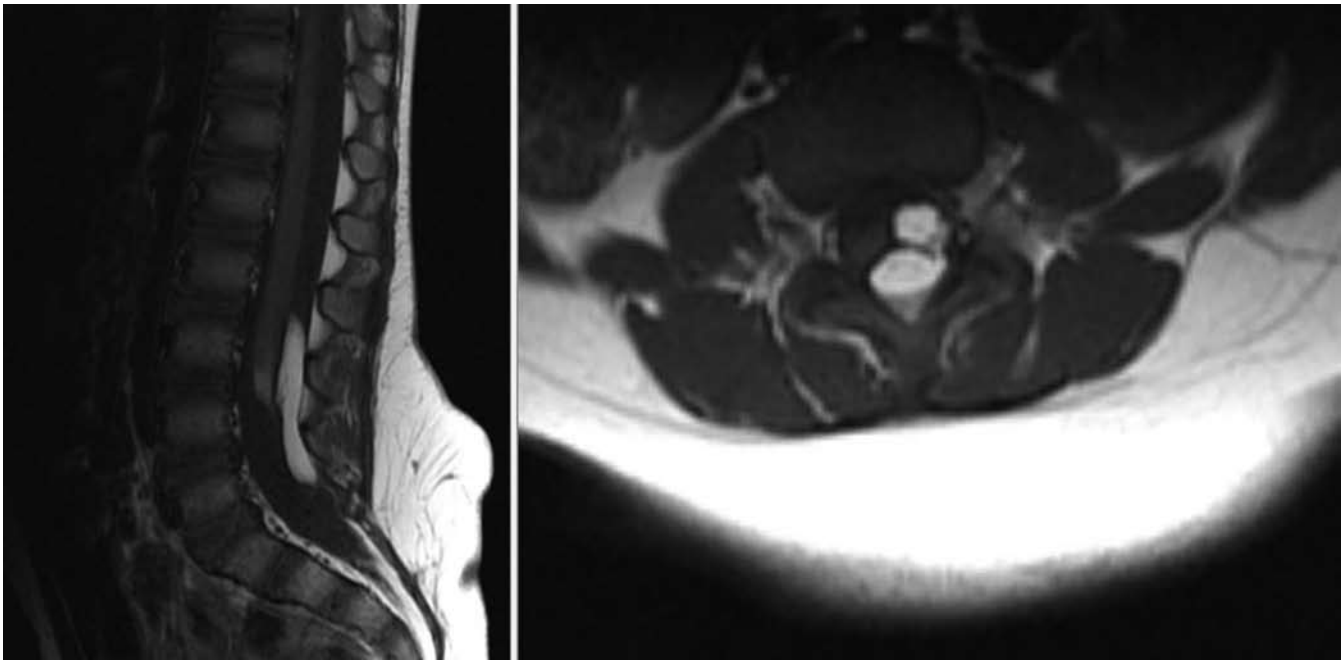

Fig. 7. Magnetic resonance imaging with T1-weighted sagittal **(A)** and axial **(B)** sequences of 2-year-old with lipomyelomeningocele. The conus medullaris terminates at L-4 and is tethered by an intraspinal lipoma measuring 4.5 cm in length. The intradural lipoma is connected with abnormal extradural fat. There is also extensive, abnormal subcutaneous fat at the gluteal cleft.

way to assess TCS as the majority of patients with MMC will have a low-lying conus on MR imaging but no related symptoms.[4,17] 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.[20,21] 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).[16,21] The most common manifestation is a subcutaneous mass, followed by skin dimples, hemangioma, hair patches, skin tags, and depigmented regions.[16,21] 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.[16] Koyanagi et al.[23] 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.[16,21] Kanev et al.,[21] 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.[21,23] Review of the literature and retrospective analyses of large case series has led some authors to advocate for detethering at the time of presentation regardless of age or neurological status.[20]

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

Authors & Year	No. of Patients	% Retethering After MMC Repair
Phoung et al., 2002	1435	2.8
Bowman et al., 2001	118	32
Shurtleff et al., 1997	654	13
Tamaki et al., 1988	60	15
Bowman et al., 2009	502	23
Selber & Dias, 1998	46	26

TABLE 3: Long-term follow-up of patients with lipomyelomeningocele/occult spinal dysraphism tethered cord

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

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.[12,25,28,31] 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.[12,25,28,31] Pang and Wilberger[31] 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[28] 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.[18,25,31,36,42] The pain is most often nondermatomal, 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.[31,47] 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[31] 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.[48,49] 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 often 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.[31] 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.[12] 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.[2] Little data exists about the natural history of adult TCS.[20,22,23,31]

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

Authors & Year	No. of Patients	Mean Age at Presentation	% w/ Pain	% w/ Motor Weakness	% w/ Bladder Dysfunction
Rajpal et al., 2007	61	36	56	79	34
Lee et al., 2006	59	43	73	78	71
Huttmann et al., 2001	56	34	77	57	70
van Leeuwen et al., 2001	57	41	74	44	67
Pang & Wilberger, 1982	23	39	78	65	56

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.[28] 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.[12,31] 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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Pathophysiology of adult tethered cord syndrome: review of the literature

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Object. Tethering of the spinal cord has been a recognized cause of neurological symptoms in pediatric patients and is increasingly being recognized as a cause of symptoms in adults as well. The pathophysiology surrounding spinal cord tethering has begun to be understood in the pediatric population but is still unclear in adult patients.

Methods. Using a PubMed database literature search, the authors reviewed the pathology and pathophysiology surrounding the tethered spinal cord, focusing particularly on the pathophysiology of adult tethered cord syndrome (TCS).

Results. Experimental data obtained in pediatric patients at surgery and in animal models indicate that spinal cord tethering causes a reduction in spinal cord blood flow and dysfunction of neuronal mitochondrial terminal oxidase. Retrospective analyses of patients undergoing surgery for adult TCS show that many adults developed symptoms following an event that could stretch the spinal cord, while others did not. Many patients also were found to have structural lesions in addition to a tethered spinal cord at diagnosis.

Conclusions. Both adult and pediatric TCSs are likely the result of a relative lack of blood flow to the spinal cord, causing dysfunction in mitochondrial oxidative phosphorylation. The likely reason the syndrome present later and differently in adults is that a secondary threshold of tension or a cumulative effect of repetitive, transient tension is placed on the cord before symptoms are recognized. *(10.3171/2010.3.FOCUS1080)*

Key Words • **adult tethered cord syndrome** • **pathophysiology** • **ischemia** • **spine surgery**

Tethered cord syndrome was originally described in 1976 by Hoffman et al.[7] after observing that the conus medullaris was affixed via an enlarged filum terminale to the sacrum in 31 children and that there was noticeable neurological improvement following release of the cord. Since that time, the term has encompassed many forms of occult spinal dysraphism in children who present with a constellation of neurological symptoms including motor, sensory, and urological complaints, as well as imaging findings of an affixed conus with a variety of structural pathologies including lipoma, tumor, fibrous tissue, and myelomeningocele, to name a few.[1,2,4,12,15,22]

Recently, it has become recognized that adults may also present with a variety of neurological complaints secondary to a tethered spinal cord.[1,6,9,10,15] Unlike children, adult symptomatology is slightly different, with pain being the most common presenting symptom (rare in children[4]), and this is followed by sensory disturbances, motor dysfunction and weakness, and urinary dysfunction.[1,2,13,15,18,20] Similar to the pediatric population, surgical intervention to release the adult tethered cord has proven to be successful in preventing further deterioration as well as sometimes improving symptoms and deficits. Thus, cord release has increasingly been accepted as a necessary intervention in symptomatic patients.[2,6,9,10,15]

The question as to what causes neurological dysfunction in patients with TCS has long been only theorized. Over the last 2 decades, there have been multiple advances in the understanding of the basic science and pathology behind the tethered spinal cord in animal models and in humans. These insights have begun to elucidate the pathophysiology behind this disorder.[16,21–23] However, what is still not clear is whether the adult tethered spinal cord and the pediatric tethered spinal cord share the same disease process and pathophysiology, or if they represent 2 different pathological processes, which could explain the different ages at presentation and different presenting symptoms.

Methods

A comprehensive literature search was performed using the PubMed database for all journal articles published until March 2010. Key words used in the search included "adult tethered cord syndrome," "pathophysiology," "pediatric," "outcomes," and "surgery"; terms were searched individually or in combination. The appropriate articles for our review were selected based on the scientific investigations surrounding the pathology and physi-

Abbreviations used in this paper: LDF = laser Doppler flowmetry; redox = reduction/oxidation; TCS = tethered cord syndrome.

ology of TCS in humans and animal models. In total, we found 4 papers addressing the basic science pathophysiology behind TCS, as well as many retrospective surgical analyses and reviews addressing the issue.

Results

Acute Model of Tethered Spinal Cord

Yamada et al.[22] examined the redox ratio of cytochrome a,a_3 using dual-wavelength reflection spectrophotometry as a surrogate marker for the metabolic function of neurons in human spinal cords at surgery and animal tethered cord models. Changes in the ability of a cell to undergo oxidative metabolism can be identified by changes in the redox ratio of this (cytochrome a,a_3) terminal oxidase of the mitochondrial electron transport chain used in oxidative phosphorylation of adenosine diphosphate to adenosine triphosphate. During a period of hypoxia, cytochrome a,a_3 becomes reduced and thus the redox ratio is increased and easily measured.

At the time of surgery in 7 pediatric patients with TCS, there was an abnormal unresponsiveness (no or only mild change in redox ratio) to transient induced hypoxia (via decreasing concentration of FiO_2) prior to tethered cord release. However, after the release, there was a trend toward normal redox changes of cytochrome a,a_3 following transient hypoxia (increase in redox ratio). This change suggested that mitochondria prior to release were highly reduced, and thus, hypoxic changes did not produce significant further reduction of cytochrome a,a_3, suggesting that there is a chronic inability of mitochondria to efficiently produce adenosine triphosphate in the tethered cord. The trend toward normal redox ratio changes following release suggests that there is potential for normal mitochondrial function following surgery.

These effects were further studied in an experimental cat model, in which a progressive amount of traction was placed on the filum terminale in a controlled setting, and reduction of cytochrome a,a_3 was determined. It was found that with the application of low and moderate amounts of traction, there was a marked trend toward cytochrome a,a_3 existing in the reduced state (thus indicating a state of relative ischemia), which was completely reversible after the traction was removed. When a large amount of traction was implemented, there was again significant reduction of cytochrome a,a_3; however, after removal of the traction there was incomplete resolution of the oxidative state of cytochrome a,a_3. This suggested that with a large degree of traction, irreversible damage to the spinal cord might occur secondary to the traction-induced mitochondrial dysfunction.

Chronic Model of Tethered Spinal Cord

Yamada et al.[23] performed additional experimentation using the cat model to measure redox ratio changes and observe behavior changes at various time points up to 9 months following the imposition of chronic, isometric traction on the filum terminale. They observed that the cats' hind limbs were weakened immediately following experimental tethering and that this weakness was worse in cats that had more traction applied to create the tethering. However, after several months' time, the cats adapted to the tethering and were then able to use their hind limbs naturally. This process took longer in the group in which more traction was applied than in the group with less traction. The redox ratios were measured at the time of sacrifice (9 months)—when all subjects were noted to have normal observed hind limb function—and were noted to be no different than controls. Light and electron microscopy of spinal cord specimens obtained in these animals were also noted to be no different than those acquired in controls.

Blood Flow Studies in Tethered Spinal Cord

Schneider et al.[16] used LDF to measure microcirculation of the spinal cord in 10 pediatric patients undergoing surgical tethered cord release. With detethering, it was noted that in all patients there was an increase in spinal cord blood flow as measured using LDF. In fact, on average, the spinal cord blood flow more than doubled. As a control, LDF was used to measure spinal cord blood flow in 5 patients undergoing dorsal rhizotomy. Blood flow to the spinal cord measured by LDF did not change in any of these patients at any point during the procedure and was similar to parameters measured in patients following tethered cord release.

Reduction of cytochrome a,a_3 through dual-wavelength reflectance spectrophotometry has also been examined by Yamada et al.[19,21] following aortic occlusion in cat models. They found that as blood pressure and oxygen tension declined, there was a marked increase in the level of reduced cytochrome a,a_3 (redox ratio increased). Additionally, interneuron potentials measured on the surface of the dorsolateral surface of the cord were markedly decreased after aortic occlusion. Both redox ratio and interneuron potentials recovered after blood flow was restored. However, if blood flow was interrupted for a longer period of time (15 minutes), there was only a partial recovery. This data provided a link between Yamada and colleagues' prior work with redox state of cytochrome a,a_3 and blood flow to the spinal cord.

Theorized Pathophysiology Based on Retrospective Analysis

The pathophysiology of adult TCS has been theorized by many groups based on their retrospective analyses of patients undergoing untethering surgery. Many of these theories take into account the aforementioned experimental data and extrapolate hypotheses as to how and why patients with adult TCS present in adulthood as opposed to childhood.

When Pang et al.[15] first described adult TCS, they theorized the pathophysiology behind it based on a variety of precipitating factors that led to a patient's presentation in 61% of their series. Three mechanisms were noted: 1) transient stretching of the spine, 2) mechanical constriction/narrowing of the spinal canal, and 3) spinal trauma, all in the presence of an already tightly tethered conus medullaris. In their series, examples of transient stretching included the lithotomy position during child-

birth, forced flexion of the hips following motor vehicle collision, prolonged sitting, and "missionary position" intercourse. Mechanisms causing spinal canal narrowing included heavy lifting, lumbar spondylosis/spondylolisthesis, and intervertebral disc herniation. Mechanisms of trauma included direct trauma to the lumbar spine as well as falling on the buttocks. Pang et al. theorized, based on the majority of their patients' presentations following such a precipitating factor, that the amount of traction determines age of symptom onset. Thus, the degree of tethering on the conus in many adult patients may not be enough to cause symptoms until there is a precipitating factor that increases traction directed to the conus. Following Pang and colleagues' initial description, many groups have noted similar precipitating factors in their patients with adult TCS,[1,2,6] even up to 70% of patients.[8]

Long before Pang and colleagues' description of symptom onset in ATCS,[15] Breig[3] demonstrated that flexion of the neck could cause sudden movement of the spinal cord. Repetitive tension resulting from various movements placed on a cord that is already tethered could, in theory, cause cumulative damage leading to adult-onset TCS.[6,13,15,20] Yamada and Losner[18] further categorized such movements that cause straightening of the lumbar spine and thus could potentially stretch a tethered spinal cord as the "three 'B' signs:" 1) difficulty sitting with legs crossed ("like Buddha"), 2) difficulty bending, and 3) difficulty holding a small amount of weight at the waist level (similar to the weight of a "baby").

Additionally, many patients who present with adult TCS have structural lesions that are tethering the cord and causing symptoms. These lesions include tumor, myelomeningocele, lipomyelomeningocele, fibrous tissue and arachnoid adhesion surrounding the filum terminale, and scar tissue from prior operation, to name a few.[5,9,10,14,17,18] Thus, it has been proposed that the adult patient with such an underlying pathology may present only after such pathology slowly progresses to the point that it significantly tethers (and therefore places traction upon) the cord.[18,20]

Adults may also present because of retethering from scar tissue formation following an initial operation for spinal dysraphism.[1,6,11] Retethering is considered a relatively common postoperative complication following correction of pediatric spinal dysraphism.[6,9] No studies were found analyzing the pathophysiology of retethering following operative intervention of pediatric TCS and spinal dysraphism.

Discussion

Since its first description nearly 2 decades ago,[15] adult TCS has become an increasingly recognized pathological entity encountered by neurosurgeons. Multiple retrospective analyses have been performed, suggesting that this disorder is perhaps more common than previously thought, may cause significant neurological dysfunction, and may be treated adequately with surgical cord release.[1,2,5,8,14,17] Common presenting symptoms,[1,2,5,6,8,9,14,16,17,20] indications for surgery,[6,8,13,18] outcomes following surgery,[2,5,6,8,10,13–15,17,18,20] and complications after surgery[2,8,10,14,17] have been well described in the adult TCS literature. However, the pathophysiology of adult-onset TCS is less clear. While there have been many advancements over the last 20 years to help explain the basic science physiology behind TCS, we do not know why a minority of patients with TCS do not present until adulthood. Furthermore, because both adult and pediatric TCS often present with different symptomatologies, it begs the question of whether the underlying pathology behind the 2 syndromes is different.

After the excellent work that Yamada et al.[21–23] have performed over the last 20 years, it seems that in both human and experimental animal models of TCS there is dysfunction in neuronal mitochondrial terminal oxidase in the electron transport chain. Such inability to use oxidative methods to efficiently produce adenosine triphosphate via oxidative phosphorylation likely causes cellular dysfunction that ultimately leads to cell death. Similar experiments proved that similar mitochondrial redox dysfunction was observed in experimental models in both an acute and chronic model of spinal cord tethering; however, the dysfunction eventually improved in the chronic model.[23] Although it was clear that this change in terminal oxidase redox state was caused by hypoxia, the cause of hypoxia in TCS could not be elucidated from the original experiments.[22] It was later shown that alteration in cytochrome redox state could be caused by experimental cessation of blood flow.[19] Furthermore, it has been shown by Schneider et al.,[16] using LDF, that there was a relative decrease in spinal cord blood flow in vivo in patients with pediatric TCS. Following cord release, blood flow normalized to control levels.

Thus, it seems that in children and animal models alike, tethering of the filum terminale causes a reduction in blood flow to the spinal cord, causing local tissue hypoxia that ultimately causes dysfunction of neuronal mitochondria, which can lead to cellular dysfunction and death. Furthermore, it seems apparent that the degree of traction on the spinal cord 1) correlates with cellular dysfunction, and 2) determines the permanence of this dysfunction after traction is released. One can extrapolate that if the filum is under too much tension, blood flow to the cord is not only reduced but is cut off altogether, causing neuronal cell death and permanent dysfunction that will not reverse after blood flow is restored following cord release. This also seems to explain how a patient who has had symptoms for a longer period of time may have a less likely chance of regaining neurological function following filum terminale release.

Following the observations made during retrospective analysis of adults with TCS, it seems that despite differences in presenting symptomatology, adults with a tethered cord have many of the same findings as children intraoperatively and radiologically. Thus, does adult-onset TCS represent the same pathophysiological process that is seen in pediatric TCS, or does the delayed presentation indicate a different underlying pathological process altogether?

Many authors argue that what determines when symptoms begin is actually how much tension is on the filum terminale.[2,15,18] This theory is consistent with the experimental data summarized above. Thus, adults who

are found to have TCS likely have always had a tethered spinal cord that has not been under significant tension. In children, symptoms develop as the child grows and the inflexible tethering of the filum is stretched and placed under tension. In this case, the degree of tethering is likely much greater than in adult patients with a tethered cord, and thus the necessary amount of tension placed on the cord is realized earlier in life when the child's spinal column reaches a certain size. This also would explain why adults with TCS often present after a precipitating event.[1,2,6,8,15] If the adult tethered cord is under just enough tension to not cause symptoms, any movement or lengthening of the spinal canal that would place slightly more traction on the already tethered cord may exceed the amount of tension that blood flow may overcome, thus producing hypoxia and symptoms. Thus, the patient with a tethered cord with only slight tension on the filum would be thrown over the "threshold" level of tension to reduce blood flow and cause symptoms if there was a precipitating event such as an acute disc herniation, for example.

This theory does not explain the existence of adult patients with TCS who do not present after a precipitating event. Rather, in these patients, it seems most likely that either 1) the underlying tethering process, such as fibrous tissue, scar tissue, tumor, or lipomyelomeningocele,[5,10,14,15,17,18] progresses to the point that it places enough tension on the cord to overcome blood flow, or 2) the normal daily movements of the neck (as Breig described[3]) and back place enough transient tension on the already tethered cord that they cumulatively produce enough ischemia periodically over the course of many years to begin to slowly cause symptoms.[6,13,15,20] In the first case, the underlying pathology progresses until the tension it is placing on the tethered cord exceeds the threshold limit to cause symptoms. In reality, adult TCS is likely explained by both the "threshold" tension model and the repetitive, cumulative-effect model described above.

Thus, it seems logical that adult-onset TCS is also caused by a reduction of blood flow to the spinal cord, as implicated in children with TCS and animal models, because such a reduction in blood flow is likely related to the degree of tension the cord is under. The overall, underlying pathophysiology in adult TCS appears to be the same as that in pediatric TCS. However, the presentation of adult TCS is different because the reduction of blood flow to the cord occurs over differing periods of time in adult TCS and pediatric TCS. This may very well explain some of the different presenting symptoms in adults and children with a tethered cord (namely, that adults are more likely than children to present with chronic pain).

Conclusions

Both adult and pediatric TCSs are likely caused by tension placed upon the filum terminale, causing a reduction in blood flow to the spinal cord that, in turn, affects cellular respiration and function. The delayed presentation of adult TCS is most likely the result of the following: 1) the fact that there is a threshold level of tension required for symptoms that is not reached until events later in life precipitate further tension on the cord in some patients, whereas in other patients 2) the low level of transient tension that is placed on the tethered cord with movement does not cause neuronal dysfunction until years of cumulative, repetitive movement reduce blood flow. Despite the similar underlying pathophysiology, such variations to achieve reduction in blood flow may explain some of the different presenting symptoms among adults and children with tethered spinal cords.

Disclosure

Dr. Park is a consultant for Medtronic and Depuy Spine.

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Electrophysiologically guided untethering of secondary tethered spinal cord syndrome

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Object. Many patients develop neurological symptoms related to spinal cord tethering after perinatal repair of myelomeningocele. This is referred to as secondary tethered cord syndrome (STCS). The authors describe their methodology and evaluate the intraoperative utility and postoperative outcomes of electrophysiologically guided untethering for STCS. In addition, the authors describe the use of electrophysiological guidance to identify an "autonomous placode" in the untethering of the cord in STCS.

Methods. The authors retrospectively identified 46 untethering procedures in 38 patients who had undergone perinatal myelomeningocele repair and in whom the index surgery was for tethered cord release at the site of the repair. In all cases, both passive (electromyography) and active (detection of compound muscle action potentials) electrophysiological monitoring was used. The proximity to neural elements was determined based on the current used; eliciting compound muscle action potentials with a ≤ 10-mA stimulation was assumed to represent direct neural stimulation. Clinical records were reviewed to evaluate the utility of electrophysiological guidance and patient outcomes.

Results. The median age at the time of untethering was 9.5 years (range 0.5–54 years). The median follow-up time was 42 months (range 3–172 months). Progressive bowel and bladder dysfunction, diagnosed either clinically or by cystometrogram, and low-back pain were the most common presenting symptoms. Intraoperative findings indicated that the most common causes of tethering were dense scar (76%) and a tethered placode (39%). Electrophysiological monitoring identified functional neural tissue near tethered elements and provided intraoperative guidance in all cases. In 41% of cases (19 cases), the untethering plan was noted to have been significantly influenced by intraoperative neurophysiological findings. Moreover, an autonomous placode was identified in 6 patients who were nonambulatory preoperatively and had presented with increasing pain and spasticity. In electrophysiologically silent areas, more aggressive dissection and untethering were possible. Symptoms of low-back pain, lower-extremity paresthesia, and lower-extremity spasticity were most likely to improve after untethering surgery (91, 88, and 82%, respectively). Sectioning above the electrophysiologically defined autonomous placode resulted in significant improvement in back pain and lower-extremity spasticity in 5 of 6 patients. There was 1 case of immediate postoperative neurological deterioration (fecal incontinence). All patients remained clinically stable or improved on long-term follow-up, except for 6 (16% of patients) who required a total of 7 additional procedures for recurrent symptoms (median time to repeat surgery 36 months). Complications were noted in 8 cases, including infections and CSF leaks.

Conclusions. Surgical untethering of STCS halts progression and often improves preoperative symptoms. Electrophysiological monitoring, using both a threshold-based interpretation system and continuous electromyography monitoring, provides an efficient, effective, and reliable method for intraoperative guidance, thereby limiting iatrogenic injury and providing a means to identify and untether autonomous placodes. Electrophysiological monitoring also allows for more aggressive dissection and untethering in functionally silent regions, possibly decreasing retethering rates. *(DOI: 10.3171/2010.3.FOCUS09299)*

Key Words • tethered spinal cord • secondary tethered cord syndrome • spinal cord untethering • intraoperative monitoring • myelomeningocele

Between 15 and 25% of patients who undergo perinatal repair of myelomeningoceles will develop neurological symptoms related to spinal cord tethering, or STCS.[3,19,23] Spinal cord tethering results in neuronal, metabolic, and vascular derangements, which can manifest clinically as pain, paresthesia, weakness, spasticity, or bladder and bowel dysfunction.[24] Studies of the natural history would suggest that these symptoms are progressive without intervention.[16] Surgical untethering is primarily aimed to prevent further neurological deterioration, and in some cases may allow for functional improvement. Untethering of the spinal cord after a previous myelomeningocele repair is, however, challenging because the structural and functional anatomy is distorted due to both developmental anomalies and scar from previous surgery. The placode and functional nerve roots can be difficult

Abbreviations used in this paper: CMAP = compound muscle action potential; CMG = cystometrogram; EMG = electromyography; STCS = secondary tethered cord syndrome.

to distinguish from scar, can be intimately involved with scar, or can be tethered themselves, placing the patient at significant risk of neurological deterioration from the untethering procedure. Measures are therefore necessary to minimize the risk of iatrogenic injury during untethering procedures.[3,8,12,19,20]

Previous reports, including a recent one by Al-Holou and colleagues,[1] have described the benefits of surgical untethering in this patient population, clearly documenting the benefits of intervention.[7] In this study, we again assess outcomes of untethering in this population. However, we focus on the role and contribution of electrophysiological guidance for surgical untethering. We have developed a systematic approach of using intraoperative neurophysiological mapping to guide repeat operations for spinal cord tethering in patients with myelomeningocele. We use neurophysiological monitoring intraoperatively to define the functionality of nerve roots and the placode and to guide surgical untethering. This allows determination of the proximity of functional neural elements for preservation, more aggressive untethering in electrophysiologically silent areas, and identification of and transection above autonomous placodes, which could not otherwise be done. We describe our experience using this technique, including an assessment of intraoperative utility and patient outcomes.

Methods

Patient Selection

We retrospectively reviewed all surgeries for spinal cord untethering over a 13-year period (1992–2005), identifying 101 consecutive surgeries performed using neurophysiological guidance. Patients were included in the current analysis if they met the following criteria: 1) they had undergone perinatal myelomeningocele repair; and 2) the index surgery was for release of tethered cord at the site of myelomeningocele repair. We identified 46 untethering procedures in 38 patients meeting these criteria (6 patients underwent 2 surgeries and 1 patient underwent 3 surgeries).

Patients were selected for surgery if they presented to neurosurgical attention with signs and symptoms consistent with spinal cord tethering (as described in Table 1 and the *Results* section) and MR imaging demonstrated radiographic evidence of tethering. All patients underwent preoperative EMG and CMG evaluation.

Operative Technique (Including Electrophysiological Mapping)

In all cases, we used both passive and active monitoring. Free-run EMG was used throughout each surgery to detect spontaneous discharges from efferent nerves due to intraoperative manipulation or stretch of functional neural elements. Active monitoring consisted of stimulating suspected functional tissue by using a bipolar electrode (5-mm interelectrode distance) and detecting CMAPs peripherally. Recording sites were selected based on preoperative EMG examination, preoperative clinical examination, and either the nerve roots in question or those at risk during surgery. Typical muscles recorded included biceps femoris, anterior tibialis, gastrocnemius, and anal sphincter. As reported previously, we use a threshold approach to determine the proximity to viable neural elements. The CMAPs elicited with ≤ 10 mA were interpreted as being a direct stimulation of the nerve.[15] The CMAPs elicited with 11–25 mA were interpreted as being near a functional nerve, probably with intervening tissue. If currents > 25 mA are needed to elicit a CMAP, we consider functional tissue to be remote and to be activated via spread through adjacent tissue. All surgical untetherings were performed by the senior author (J.A.J. Sr.). All intraoperative neurophysiological monitoring was performed by a neurologist specializing in neuromonitoring (L.H.P.).

TABLE 1: Presenting symptoms in 38 patients with SCTS

Presenting Symptoms	No. (% of 46 cases)
bladder/bowel dysfunction	30 (65)
low-back pain	23 (50)
lower-extremity weakness	20 (43)
lower-extremity spasticity	11 (24)
paresthesia	8 (17)

Outcomes Assessment

Patient charts, including operative reports, were reviewed in detail to assess the utility of neurophysiological guidance for surgical untethering. All patients were clinically evaluated immediately postoperatively and 3 months after surgery. Long-term follow-up was also obtained to determine if patients had stabilization or progression of symptoms after surgery. In addition, all patients underwent follow-up CMG evaluation to identify subtle changes in bladder function.

Results

Patient Profile

Thirty-eight patients underwent 46 untethering procedures for STCS (6 patients underwent 2 surgeries and 1 patient underwent 3 surgeries). Eighteen of the patients were male. The median age at the time of surgery was 9.5 years (range 6 months–54 years), representing a similar age distribution to that reported by Herman and colleagues[7] (Fig. 1). The median follow-up was 42 months (range 3–172 months). Eighteen patients had < 3 years of follow-up.

Most patients presented with multiple symptoms, as is common in tethered cord syndrome. Bowel and bladder dysfunction was the primary presenting symptom in nearly two-thirds of cases. Low-back pain was the second most frequent presenting symptom; it was found in 50% of cases. Presenting symptoms and corresponding frequencies are fully detailed in Table 1.

Operative Findings and Utility of Mapping

Electrophysiological monitoring identified functional neural tissue near tethered elements and provided intraoperative guidance in all cases. In 41% of cases (19 cases),

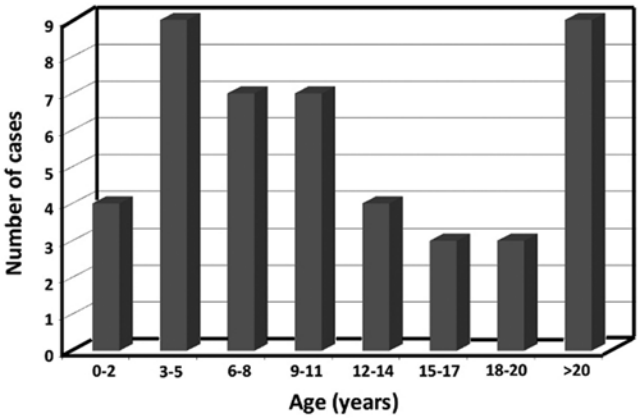

Fig. 1. Bar graph showing age distribution of patients who underwent operations for untethering of STCS. The median age at time of surgery was 9.5 years.

the untethering procedure was significantly influenced by intraoperative neurophysiological findings. For example, in 1 case, the surgeons noted that "further untethering may have caused injury" based on intraoperative neurophysiological findings. Conversely, more aggressive dissection and untethering were possible in electrophysiologically silent areas; the absence of identifiably functional tissue gave the primary surgeon the confidence to aggressively dissect and untether areas infiltrated with dense scar.

Multiple factors, even within an individual patient, can contribute to tethering of the spinal cord in STCS. In this series, the most common cause of tethering was dense scar (35 cases, 76%). A tethered placode was the next most common cause of cord tethering in this series (18 cases, 39%). The causes of tethering and relative frequencies are detailed in Table 2.

Autonomous Placode

We have previously described the concept of an autonomous placode.[15] It is defined as a placode which, when stimulated at its caudal extent with low currents (≤ 10 mA), results in marked lower-extremity CMAPs, but it produces no lower-extremity CMAPs when stimulated in a slightly more rostral position, signifying an intact efferent pathway that is completely disconnected from the functioning (and more rostral) spinal cord. Because of the functional disconnection from the normal spinal cord and because the placode is often a source of tethering (Table 2), identification of an autonomous placode and the functionally silent area between this placode and the normal spinal cord can be critical for successful untethering. Because this is a functionally defined entity, the autonomous placode cannot be identified visually and requires intraoperative neurophysiological monitoring for localization and confirmation.

An autonomous placode was identified in 6 patients, all of whom were nonambulatory preoperatively and had presented with increasing back pain and spasticity. In all of these patients, the functionally silent area superior to the autonomous placode and inferior to the normal spinal cord was sectioned. Five of the 6 patients experienced significant improvement in back pain and lower-extremity spasticity as a result of sectioning.

Symptomatic Outcomes

The goal of surgical intervention is preventing clinical progression. With one exception (discussed below), all symptoms were either stable or improved in all patients at the 3-month follow-up. Symptoms of low-back pain, lower-extremity paresthesia, and lower-extremity spasticity were most likely to improve after untethering surgery (91, 88, and 82%, respectively). Symptomatic outcomes at 3 months are detailed in Table 3. There was a single case of immediate postoperative neurological deterioration (fecal incontinence), deemed a surgical complication, which had not improved at follow-up. Long-term follow-up revealed stable clinical status in all patients relative to 3-month postoperative assessments.

A secondary measure of the efficacy of untethering is an evaluation of rates of repeat operation for persistent symptomatology. With a median follow-up time of 42 months, all patients experienced lasting clinical stability or improvement, except for 6 patients (16%) who required a total of 7 additional procedures for recurrent symptoms (similar to the original presentation), with a median time to repeat surgery of 36 months (range 3–119 months). The patient who underwent 2 additional untethering procedures had these interventions 32 and 36 months apart. Interestingly, in the patient who underwent early repeat surgery (at 3 months) an autonomous placode was identified during the first surgery, but the placode was not sectioned due to a question of persistent sensory function in the lower extremities. After repeat surgery and sectioning above the autonomous placode, the patient experienced significant improvement in her spasticity symptoms.

Surgical Complications

As noted above, neurological worsening was noted

TABLE 2: Tethering elements in 38 patients who underwent 46 procedures for spinal cord untethering

Tethering Element	No. (% of patients)
scar	35 (76)
placode	18 (39)
filum	16 (35)
lipomatous tissue	16 (35)
nerve roots	4 (9)

TABLE 3: Symptomatic outcomes at 3 months in 38 patients who underwent 46 procedures for spinal cord untethering

Symptom	% w/ Outcome (no. of cases)		
	Improved	Stable	Worse
bladder/bowel dysfunction*	20% (6)	77% (23)	3% (1)
low-back pain	91% (21)	9% (2)	0% (0)
lower-extremity weakness	35% (7)	65% (13)	0% (0)
lower-extremity spasticity	82% (9)	18% (2)	0% (0)
paresthesia	88% (7)	12% (1)	0% (0)

* Based on clinical or CMG evaluation.

in 1 patient, who had fecal incontinence after surgery. In addition, there were 8 cases of operative complications, including 2 infections (4%), 4 CSF leaks (9%), and 2 combined infection and CSF leaks (4%), all requiring operative intervention.

Discussion

Regardless of the cause of the condition, the goal of tethered cord surgery is to alleviate the neuronal, metabolic, and vascular derangements induced by spinal cord stretch.[24] In doing so, we hope to thwart further neurological deterioration, to reverse current neurological deficits, to avoid iatrogenic injury, and to prevent retethering. Achieving these goals requires aggressive and maximal untethering, which is complicated in STCS by prior repair of spinal dysraphism and disrupted anatomy of the caudal spine. Although others have reported the use of neurophysiological monitoring for tethered cord release, this is the largest series specifically analyzing the utility and outcomes of this technique in STCS.[1,13,14,17,22]

Secondary Tethered Cord Syndrome

Although radiographically demonstrated spinal cord tethering occurs in nearly all patients after repair of spinal dysraphisms, not all patients are symptomatic. The use of CMG diagnostics can be particularly helpful in identifying patients who are symptomatic from secondary tethered cord, and can be highly useful for operative decision making.[4] Although patients may present at any age, the most common age at presentation is between 5 and 9 years, concurrent with the period of rapid growth.[1,7] Because children may not always recognize bladder dysfunction, a CMG evaluation should be included in the standard assessment of STCS. It is imperative that symptomatic secondary tethered cords are recognized early because delayed recognition is associated with irreversible injury.[16]

Neurophysiological Monitoring for Cord Untethering

Multimodality neurophysiological monitoring for tethered cord release has been reported previously. For example, von Koch and colleagues[22] reported using neurophysiological monitoring for tethered cord release in 25 pediatric patients with a thickened filum and low-lying conus. Consistent with this report and as might be expected, they state that functional tissue can easily be differentiated from thickened filum based on stimulation thresholds, the latter requiring thresholds of nearly 100 times that of functional tissue. The same group reported similar results and utility of neurophysiological monitoring for tethered cord release in 15 adults with a diverse number of causes of tethered cord, including thick filum, spinal cord malformations, and low-lying conus.[17] Paradiso and colleagues[14] have also reported success with identifying viable tissue intraoperatively based on stimulation thresholds. Although the pathophysiological mechanisms of neurological deterioration are probably similar across causes for tethered cord syndromes, the techniques used for untethering cannot necessarily be extrapolated to patients with STCSs, in which the filum may not be readily visually identifiable, and in which little normal comparative anatomy is available.

In this series, a threshold approach to interpreting intraoperative stimulation provided a rapid means of assessing the in situ functional architecture of the secondary tethered cord. Neurophysiological monitoring in our series was effective in minimizing iatrogenic injury. In all cases, functional tissue was identified near the site of tethering. Moreover, 41% of cases were associated with an altered plan for untethering based on intraoperative findings. Whereas neurological deterioration has been reported in up to 9% of cases in other series of secondary tethered cord surgery regardless of the use of neurophysiological monitoring, neurological worsening in our series was limited to 1 surgery (2.2%), a rate similar to that reported in other series in which neurophysiological monitoring was used.[9,17] Having this rapid means of assessment is essential, considering that the two most common tethering elements are scar and placode, entities that are better defined functionally than visually.

Posttreatment Outcomes

Although the goal of spinal cord untethering is primarily to halt clinical progression, the majority of patients experience significant improvement, especially with respect to pain, paresthesia, and spasticity (see Table 3). Overall, approximately 75–90% of patients experience improvement or stabilization of symptoms.[1,7,9,12,18] Pain is most amenable to improvement; Al-Holou and colleagues[1] recently reported improvement in pain in 75% of patients.[9,10,12,20] Bowel and bladder function, on the other hand, are less likely to improve, with only 13–46% of patients reporting improved function (20% in this series).[1,7,17,22] In addition, we suspect that untethering contributed to the stabilization of bowel and bladder function in the other patients who presented with these symptoms.

In addition to neurological improvement, one of the goals of cord untethering is to prevent retethering or a repeat operation. Retethering requiring a repeat operation is estimated to occur in 20–30% of all patients, and in up to 50% of pediatric patients.[2,6,18] Techniques have been described to prevent retethering, including the use of intradural retention sutures and placement of implants (or dural substitutes) to reconstruct and maintain the intradural space.[21,25] We had to perform 7 repeat operations, one of which was needed due to incomplete untethering. Excluding this case, the rate of retethering requiring repeat operation was 15.4%, which is favorable relative to previous reports. Considering that the median time to the repeat untethering in this series was 36 months, and that untethering procedures have been reported in the literature several years after the index surgery, the reported retethering rate may be artificially low due to the number of patients in this series in whom the follow-up was less than 3 years.[18] Alternatively, more aggressive untethering with neurophysiological monitoring may have contributed to a decreased retethering rate.

Placode Transection

Tethering at the level of the placode is a common

finding and has been postulated to be due in part to the absence of a normal pial plane, and to the unavoidable contact of the placode with the reconstructed canal.[5] As we have previously noted, and report in additional cases in this paper, transection above the level of an autonomous placode is an efficacious treatment for paraplegic patients (for whom there is no risk of neurological deterioration) presenting with progressive spasticity. Although with some variation (for example, transection immediately rostral to the point of maximal placode adhesion), similar placode transection approaches have been previously described, with excellent results: nearly 100% pain and spasticity relief.[5,6,11,23] Although it has been suggested that transection might be reserved for those with recurrent retethering,[5] because of its efficacy (83% improvement), we advocate consideration of transection above the level of the autonomous placode at the time of initial untethering. Importantly, because the "autonomous placode" is functionally defined, it necessitates neurophysiological monitoring for identification.

Limitations of Monitoring

Although we believe that neurophysiological monitoring is critical to preserve function and to maximize untethering in STCS, we recognize the limitations. First, the success of motor root mapping is limited by the selection and placement of electrodes, which relies on a thorough preoperative clinical and electromyographic examination as well as a clear operative plan that recognizes which spinal levels may be at risk during surgery. Second, passive EMG monitoring for stretch-induced discharges has limited sensitivity; nerves may be stretched or injured without spontaneous discharges. It is therefore imperative not only to rely on passive monitoring, but also to use active mapping with a threshold-based interpretation system, providing a reliable and rapid assessment of functional architecture. Third, electrodiagnostic testing during surgery probably increases operative and anesthetic times. We believe that the time increase is modest when the test is routinely performed by experienced neurophysiologists and technicians, and that the benefits (for example, decreased iatrogenic injury) justify the potentially increased time of surgery and anesthesia.

This study is also limited by its design, being based on an institutional, retrospective, uncontrolled series. To determine whether this technique improves outcomes would technically require a prospective randomized trial. It is unlikely that this could be accomplished. We believe that intraoperative electrophysiological monitoring provides the surgeon with information that is otherwise unattainable, thereby making the surgery safer (preserving nerves) and more efficacious (identifying autonomous placodes).

Conclusions

Surgical untethering of secondary tethered cord syndrome is effective at halting symptomatic progression of bowel and bladder incontinence and weakness, and it improves preoperative symptoms of pain, spasticity, and paresthesia. Intraoperative neurophysiological monitoring, using both passive and active mapping and a threshold-based system for interpreting maps, provides an efficient, effective, and reliable method for intraoperative guidance, thereby limiting iatrogenic injury during untethering of STCS, and providing a means to identify autonomous placodes that can be detached from the tethered spinal cord. Moreover, using electrophysiological monitoring allows for more aggressive dissection and untethering in functionally silent regions, hopefully resulting in a decreased rate of retethering and repeat operation.

Disclosure

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Author contributions to the study and manuscript preparation include the following. Conception and design: Pouratian. Acquisition of data: Elias. Analysis and interpretation of data: Pouratian, Elias. Drafting the article: Pouratian. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Statistical analysis: Pouratian. Study supervision: Elias, JA Jane Jr, JA Jane Sr.

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Human motor evoked potential responses following spinal cord transection: an in vivo study

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Motor evoked potential (MEP) monitoring has been used increasingly in conjunction with somatosensory evoked potential monitoring to monitor neurological changes during complex spinal operations. No published report has demonstrated the effects of segmental spinal cord transection on MEP monitoring.

The authors describe the case of an 11-year-old girl with lumbar myelomeningocele and worsening thoracolumbar scoliosis who underwent a T11–L5 fusion and spinal transection to prevent tethering. Intraoperative MEP and somatosensory evoked potential monitoring were performed, and the spinal cord was transected in 4 quadrants. The MEPs were lost unilaterally as each anterior quadrant was sectioned.

This is the first reported case that demonstrates the link between spinal cord transection and MEP signaling characteristics. Furthermore, it demonstrates the relatively minor input of the ipsilateral ventral corticospinal tract in MEP physiology at the thoracolumbar junction. Finally, this study further supports the use of MEPs as a specific intraoperative neuromonitoring tool. *(10.3171/2010.3.FOCUS09201)*

Key Words • motor evoked potential • neurophysiological monitoring • spinal cord transection

Clinical neurophysiological monitoring, particularly SSEP monitoring, has increasingly become the standard of care for complex spinal operations, including scoliosis corrections or anterior spinal surgeries, as well as for intramedullary tumor resections. Although SSEPs in general can indicate global injury to the spinal cord, they have proven insufficient in many cases and fail to warn against postoperative neurological deficit, particularly in those cases in which damage is focally located in the anterior spinal cord.[1,5,8,16,18,41]

Motor evoked potentials were first used to measure spinal cord injury in animals in the 1970s, and they began to be monitored in humans in the 1980s.[2,14,19–21,25–29,32,33] Animal data have been able to demonstrate that loss of motor function and histopathology correlate well with permanent declines in MEP amplitudes during ischemia.[22] Perhaps most importantly, deliberate spinal cord ischemia associated with MEP declinations in animal models show preserved motor function after restoration of blood flow with associated MEP recovery.[22] Moreover, intraoperative studies in humans have shown that changes in MEPs correlate strongly with postoperative neurological status.[6,13,17,31,37] Taken together, these data suggest that MEPs offer an opportunity to measure the integrity of the motor pathways from cortex to specific muscles. Thus, the primary clinical purpose for neurophysiological studies is to alert surgeons to possible permanent injury so that therapeutic interventions can be made. In combination with cortical SSEPs, MEPs bring monitoring one step closer to predicting the functional results of an intervention.

The effects and importance of crossing corticospinal tracts are well documented in the literature.[15] However, transcranial magnetic stimulation in patients following stroke have also shown evidence of an ipsilateral corticospinal pathway that may be of importance in MEP monitoring.[39] This ipsilateral pathway, which has been demonstrated in rodents, cats, primates, and humans, may comprise up to 30% of corticospinal tract fibers.[30] Although the termination of these fibers remains uncertain, recent publications indicate that these ipsilateral projections may have a significant role in motor control.[15]

In summary, human data have not been published

Abbreviations used in this paper: MEP = motor evoked potential; SSEP = somatosensory evoked potential.

that demonstrate that MEPs directly assay the integrity of the corticospinal motor pathways. Additionally, data elucidating the effect of the ipsilateral ventral corticospinal tract on MEP recordings are lacking. Finally, no published reports have detailed changes in MEPs following documented human spinal cord transection.

Case Report

History and Presentation. This 11-year-old girl was born with a lumbar myelomeningocele that was repaired at birth. She subsequently had neither sensory nor useful motor function in her lower extremities and no bowel or bladder sphincter function. Her thoracolumbar scoliosis was increasing, and it was beginning to cause respiratory compromise and skin breakdown; thus, surgery was recommended.

Operation. The patient subsequently underwent a thoracolumbar fusion from T-11 to L-5 with anterior and posterior fixation. As part of this procedure, the spinal cord was transected to prevent tethering and loss of upper-extremity function as a result of curvature correction.

Monitoring. Neurophysiological monitoring was used to monitor both tibial nerve SSEPs and transcranial electrical MEPs. The SSEP stimulation was a 0.2-msec duration pulse delivered to the tibial, medial, and ulnar nerves at 2.35 Hz with an intensity of 15–30 mA. The SSEPs were recorded at the midline scalp (Cz′-Fz) and contralateral scalp (C3′-Fz, C4′-Fz). Transcranial electrical MEP stimulation was an anodal pulse delivered to the contralateral scalp (C3 or C4) with a 0.05-msec duration pulse in a train of 6 pulses at an intensity of 250 V. The MEPs were recorded from the thenar and abductor hallucis muscles (or tibialis anterior).

The spinal cord was transected in stages via posterior laminectomy. The dorsal column quadrants were transected first, followed by the right anterior quadrant and then the left anterior quadrant.

Monitoring Results. The following baseline neurophysiological observations were made. Stable ulnar and median nerve SSEPS were obtained throughout the procedure. Tibial SSEPs could not be recorded. Thenar MEPs were recorded at baseline and throughout the procedure. The abductor hallucis MEPs were recorded at baseline but with much reduced amplitudes, consistent with preexisting pathology.

When the posterior columns were transected, lower-extremity MEPs remained present (Fig. 1, transected quadrants 1 and 2). After transection of the right anterior quadrant, the right tibial MEPs disappeared, and the left tibial responses remained present (Fig. 1, transected quadrant 3). After spinal cord transection was completed with transection of left anterior quadrant (Fig. 1, transected quadrant 4), the left tibial MEP responses also disappeared (Fig. 2).

Discussion

This is the first reported case in which MEP responses have been demonstrated in spinal cord transection. It demonstrates that MEPs do not respond to dorsal column disruption. Furthermore, MEPs were able to distinguish transection of the right from the left anterior spinal cord. Finally, the case does not provide evidence of a clinically significant ipsilateral corticospinal tract effect given our MEP results.

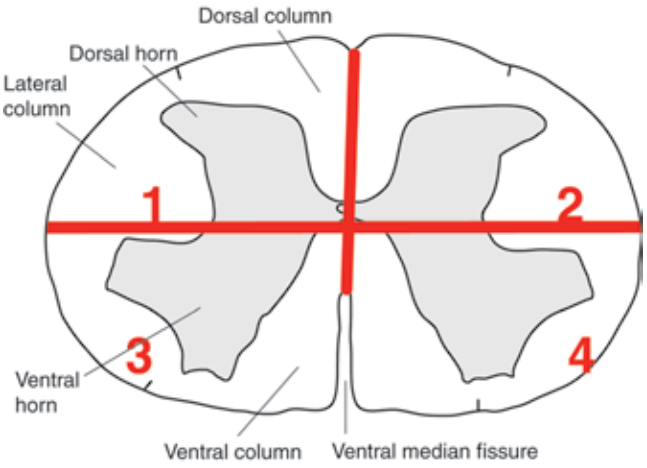

Fig. 1. Diagram of spinal cord cross-section and order of quadrant transection. The right posterior (1) and left posterior (2) quadrants were transected initially. The MEPs bilaterally remained intact. The right anterior quadrant (3) was transected, and the right lower-extremity MEPs disappeared. The left anterior quadrant (4) was transected last, and the left lower-extremity MEPs subsequently disappeared.

Despite the absence of functional motor activity in our patient's lower extremities, she did demonstrate markedly reduced but indeed intact corticospinal connectivity as measured by MEP monitoring that disappeared upon transection. This presence of signals in the setting of a clinical lack of function is likely due to long-standing deconditioning related to extremity disuse. As such, this case also illustrates the need for additional studies regarding the critical thresholds of MEP change since functional motor status was absent in the presence of MEPs prior to transection. These studies would be important as our data indicate that it might be possible to lose significant lower-extremity function from an intraoperative injury yet still have some MEP signals. Nevertheless, MEP monitoring should be regarded as a reliable proxy of corticospinal function in cases in which possible intraoperative injury to the anterior spinal cord is a concern.

The combination of MEP and SSEP monitoring was successful in this case. Motor evoked potentials can be recorded rapidly with brief stimulation, unlike SSEPs, which require signal averaging.[36] Furthermore, MEPs may be more sensitive to ischemia than SSEPs and less likely to deteriorate after midline myelotomy, unlike SSEPs.[4,5,7,10,12,24,34–36,40] Also, as demonstrated in this case, not only will MEPs often be present in neurologically compromised patients,[9] but they may also be able to detect subclinical deficits,[3,23,38] whereas SSEPs are significantly more likely to be absent in the normal and compromised neurological states. In a recent prospective trial, 38.7% of patients undergoing a variety of spinal procedures had either significantly diminished or absent tibial SSEPs. However, no patients with absent MEPs had useful SSEP signals.[5] In this same study, the addition of MEPs to the

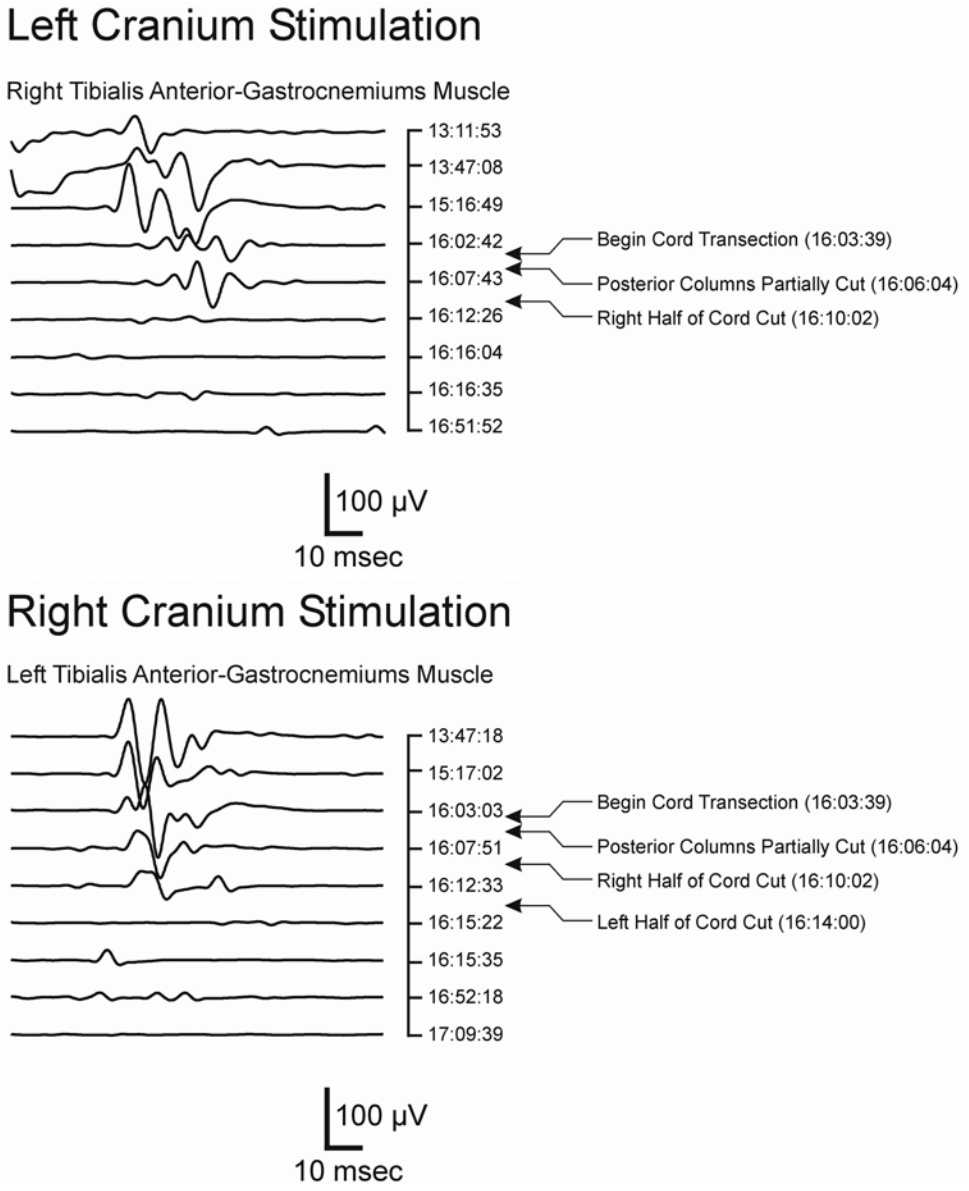

Fig. 2. Recordings of the bilateral tibial nerve–gastrocnemius muscle MEPs, demonstrating that MEPs remained after the posterior columns were transected. The potentials disappeared after the corresponding anterior quadrant was transected.

monitoring scheme meant that only 3.8% versus 26.9% could not be monitored during their surgery.

To achieve aggressive resections or corrections in complex spine and spinal cord surgeries while also minimizing the possibility of intraoperative neurological deficit requires reliable neurophysiological monitoring that accurately identifies transient injury in real time to operating surgeons. As discussed above, SSEPs alone are not consistently present in many patient populations with baseline neurological compromise. However, in multimodality intraoperative monitoring (MIOM) with MEPs, the number of patients who can be monitored increases and provides further diagnostic information that should be part of the surgical armamentarium. A study by Hsu et al.[11] demonstrated a low false-positive rate of 2.8% and a false-negative rate of 0%, suggesting that multimodality intraoperative monitoring represents the ideal confluence of intraoperative neurophysiological monitoring techniques available to protect patients from intraoperative injury.

Conclusions

This case demonstrates for the first time in a human that MEPs can directly predict unilateral loss of signal by transection of individual spinal cord quadrants. Therefore, MEPs can be considered as an additional tool for monitoring possible intraoperative spinal cord injury.

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: all authors.

Acquisition of data: all authors. Analysis and interpretation of data: Ramakrishna, Nair, Kinney, Slimp, Ko, Avellino. Drafting the article: Ramakrishna, Nair. Critically revising the article: Ramakrishna.

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Management of recurrent adult tethered cord syndrome

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Recurrent tethered cord syndrome (TCS) can lead to significant progressive disability in adults. The diagnosis of TCS is made with a high degree of clinical suspicion. In the adult population, many patients receive inadequate care unless they are seen at a multidisciplinary clinic. Successful detethering procedures require careful intradural dissection and meticulous wound and dural closure. With multiple revision procedures, vertebral column shortening has become an appropriate alternative to surgical detethering. *(DOI: 10.3171/2010.3.FOCUS1073)*

Key Words • tethered cord syndrome • vertebral column resection • spina bifida

Tethered cord syndrome is a condition that appears most commonly in the pediatric population. It results from stretching of the spinal cord between 2 fixation points, and treatment entails untethering the spinal cord. Results can vary depending on the time period between presentation and the initiation of treatment. If treated early, patients can recover from their neurological deficits. Chronic or excessive stretching of the spinal cord can lead to permanent disability. Currently the literature suggests that metabolic derangements and alterations in oxidative metabolism contribute to the reversible symptoms associated with TCS.[18] Once neuronal damage occurs, however, partial neurological deficits can persist despite untethering.

For some patients, TCS can present de novo in adulthood or can impart recurrent clinical manifestations that extend into adulthood. Adult TCS may be neglected without the multidisciplinary care provided by a pediatric spina bifida clinic familiar with the natural history, diagnosis, and management of TCS. Given the fact that a majority of patients with spina bifida now live into adulthood, it is increasingly important to diagnose and appropriately treat TCS in adults. A lack of understanding or enthusiasm in treating adult TCS can lead to significant deterioration and permanent disability.

Abbreviations used in this paper: EMG = electromyography; SSEP = somatosensory evoked potential; TCS = tethered cord syndrome; VCR = vertebral column resection.

Epidemiology

The incidence of neural tube defects is 0.17–6.39 per 1000 live births worldwide.[2] There has been a decline in incidence over the past 20 years largely due to folic acid supplementation, termination of pregnancy after prenatal diagnosis, and other unknown factors. At centers experienced with myelomeningocele, the 20- to 25-year mortality rate is 24%.[1] Significant improvements over the past couple of decades in the care of patients with myelomeningocele at centers for excellence have led to a growing population of these patients reaching adulthood. However, spinal cord tethering is a problem in this population. It occurs in 23% of patients between 1 month and 23.3 years after surgery. Twenty-nine percent of these cases occur in patients who have undergone > 1 tethered cord release procedure.[3]

Clinical Presentation

In dealing with patients with TCS, a multidisciplinary team consisting of neurosurgeons, urologists, physiatrists, neurologists, orthopedic surgeons, and physical therapists is preferred. Patients with symptomatic TCS present with back pain, worsening motor weakness, or new bowel or bladder symptoms. Of these presenting symptoms, pain is the most common. Back pain occurs in 73% of patients presenting with TCS, whereas leg pain or sciatica is the presenting symptom in 56% of patients.[11] Muscular weakness and bladder dysfunction occur as the present-

ing symptom in 78% and 71% of patients, respectively.[11] Adults with a tethered cord tend to present in a delayed fashion compared with their pediatric counterparts.[12] Adults may be in denial of any significant symptoms and unwilling to see their practitioner, and thus delaying their diagnosis and increasing their chances of disability. Progressive scoliosis, foot deformity, leg length discrepancy, or muscular atrophy diagnosed by a multidisciplinary team can also suggest TCS. Untreated, TCS can progress in 27.5%, 40%, and 60% of cases at 1, 2, and 5 years after diagnosis, respectively.[16]

Diagnostic Workup

The determination of TCS is a clinical diagnosis. Potential shunt malfunction should be assessed in any patient with a ventriculoperitoneal shunt and undergoing evaluation for TCS before any treatment of a tethered cord. Head CT scanning and/or a shunt tap can be performed to determine shunt malfunction. Baseline manual motor test results from the physical therapist can be recorded and monitored on routine visits to the clinic. Routine urodynamic tests can detect any subtle changes in bladder function. Standing scoliosis radiographs should be obtained to monitor the progression of scoliotic curves as progressive scoliosis can indicate tethering. Once the diagnosis of TCS is made in conjunction with these tests, imaging is performed to identify potential areas of adhesions. Although MR imaging can visualize a low-lying cord, fatty filum terminale, or lipomyelomeningocele for the initial presentation of TCS, it is less useful for localizing adhesions. Comparing the location of the conus on previous MR images can help to support the diagnosis of TCS. Computed tomography myelography has been found to be more helpful in detailing the regions of adhesions, which prevent the flow of contrast through the thecal sac. More importantly, this imaging procedure provides an understanding of the bony anatomy that will be useful during exposure. Electrophysiological tests, such as SSEP recording and EMG, used in conjunction with imaging can also provide useful information. Somatosensory evoked potentials can provide information regarding the severity and extent of tethering.[17] Somatosensory evoked potentials obtained preoperatively can be compared with postoperative measurements to confirm the benefits of surgery.[9]

Surgical Management

The surgical management of tethered cord remains controversial. Although treatment in a patient with TCS and evidence of neurological decline is straightforward, the care of patients who are asymptomatic despite their condition remains controversial. As previously stated, the natural history of TCS suggests a progressive decline when managed conservatively. Thus, in a symptomatic patient, any change in routine urodynamic or manual motor testing should be closely monitored.

With surgery, the initial approach is aimed at dealing with the local pathology. Over time, however, recurrent tethering can produce symptoms. In the adult population, the rate of retethering has been quoted to be as high as 25%.[5]

Lee et al.[11] found that surgical untethering can improve back and leg pain in 78% and 83% of patients, respectively. However, motor weakness will stabilize or improve in only 27% and 64% of patients, respectively. Sensory deficits remained unchanged in 50% of patients. Urological abnormalities improved in 50% of patients undergoing untethering and remained stable in 45%.

Surgical Untethering

When obtaining informed consent regarding any surgical untethering procedure, it is important to discuss all scenarios that may be encountered. Dural closures may be particularly difficult, and a CSF diversion procedure, such as a lumbar drain or external ventricular drain, may be required. A gluteal flap may be considered if significant difficulties are expected when closing the wound.

The patient is positioned prone after intubation. If any instrumentation is planned, devices are placed on a Jackson table. Any previous surgical incisions are marked out accordingly. A wide area is prepped laterally and inferiorly to include the buttocks and lateral thigh in the event that a gluteal flap is needed. Dissection is performed along the old incision site. It is important to create a single dissection plane deep and near the spine to preserve the subcutaneous tissue for closure. Ideally, dissection should be initiated along the superior or inferior margin of interest where normal bony anatomy is present to prevent entering the intradural spaces without recognition. Under these circumstances CT myelography is particularly helpful to isolate where there may be a defect in the lamina. Once the scar tissue overlying the dura as well as surrounding bony margins are identified, intradural exposure is performed. It is important to note on imaging if the spinal cord is adherent to the dura posteriorly given that entry into the intradural space can cause injury to the spinal cord. Using a 15-blade scalpel, a midline incision in the dura is made over the region of the suspected adhesions as demonstrated on CT myelography or MR imaging. A Woodson dissector is helpful intradurally to dissect a plane for sharp dissection or opening the dura. Edges of the dura are tacked up to the surrounding tissue by using a 4-0 nonabsorbable suture. A microscope is brought in for better visualization. From here, the Rhoton instruments, microscissors, and an arachnoid knife are helpful in providing safe lysis of adhesions. On occasion, the CO_2 laser may be helpful as a means of the subtle lysis of adhesions when the risk of injuring or pulling the cord with sharp or blunt dissection is significant.[4] When bipolar electrocautery is used, Isocool (Codman/Johnson & Johnson Professional) or nonsticking bipolar tips are used as well. The lysis of adhesions should continue until the cord is visualized to fall freely from its attachments.

Lumbosacral nerve roots should be identified prior to microsurgical untethering. Microsurgical techniques should focus on the careful lysis of adhesions and the removal of significant pathology leading to tethering. Such techniques are performed in conjunction with intraoperative monitoring to prevent any significant morbidity from occurring.

During evaluation, careful attention should be di-

rected at all lumbosacral nerve roots including the anal sphincter. Common modalities used to monitor include SSEP recording, spontaneous EMG, and triggered EMG. Somatosensory evoked potentials offer high specificity and low sensitivity for detecting damage to the nervous system. Conversely, continuous EMG provides high sensitivity and low specificity in detecting new neurological deficits.[15] Thus, neurological monitoring with SSEP recording and EMG provides high specificity and sensitivity in detecting new neurological deficits. With untethering of the spinal cord, SSEPs have been shown to improve.[9] Spontaneous EMG is used to detect inadvertent manipulation of nerve roots. Triggered EMG allows for the stimulation of tissues for the purpose of differentiating important neural structures from adhesions or scar tissue when performing a dissection.

After the lysis of adhesions is complete, closure of the dura is achieved with a running 4-0 locking stitch. Placing DuraGen (Integra LifeSciences) subdurally at the time of closure can be considered as it has been shown to reduce postoperative fibrosis.[7] A Valsalva maneuver is performed by an anesthesiologist to ensure no CSF leakage from the closed durotomy site. A dural sealant can be applied to the dural edges if leakage occurs around the margins. In certain circumstances in which primary dural closure is not achieved, a dural substitute can be used to close the wound. Subcutaneous wound closure is then performed. Note that a primary closure is desired. A subfascial Jackson Pratt drain can be left in place if necessary. When a drain is inserted, no or minimal bulb suction is applied without promoting CSF leakage into drain. In patients with a history of posterior operations, subcutaneous wound closure can be difficult. Achieving a tight fascia closure is ideal in the setting of a closed durotomy, and this can be done with a nonabsorbable suture instead of Vicryl sutures. After fascial closure, the subcutaneous tissue may be tense especially in regions where there is a significant amount of scar tissue. Fascial release both above and below the plane may provide adequate release of tension for closure of the subcutaneous suprafascial tissue. In certain circumstances in which there is not enough suprafascial tissue to provide for complete closure, plastic surgeons may be needed to perform a gluteal flap.

With multiple untethering procedures, arachnoid scarring around the nerve roots can lead to potential neurological deficits that may be difficult to treat. There is the risk of damaging spinal nerve roots while performing the dissection. This risk increases with each subsequent untethering operation. Multiple surgeries also increase the risk of wound dehiscence and wound infection.

Postoperative Management

The most common complication of surgical untethering is CSF leakage. The patient should be kept flat for 3–5 days to allow the durotomy site to heal. If it is believed that a CSF diversion procedure is needed, a decision needs to be made regarding the placement of a lumbar drain versus an external ventricular drain. For patients with myelomeningocele and shunted hydrocephalus, a functional shunt may need no additional intervention. Given a low-lying cord in most circumstances, a lumbar drain should be placed under fluoroscopic guidance. However, in a small portion of patients without a VP shunt in which a lumbar drain cannot be safely placed or if placement of the lumbar drain involves entering the surgical bed, an external ventricular drain is preferable. An MR image is obtained within 48 hours to establish a baseline position of the spinal cord.

Vertebral Column Shortening

Patients who have recurrent symptoms despite multiple detethering procedures may be candidates for a vertebral column shortening procedure. Vertebral column resection and pedicle subtraction osteotomy are 2 powerful techniques used in the correction of spinal deformity. These 3 column osteotomies allow the spine to be manipulated in both the coronal and sagittal planes. Both techniques effectively shorten the vertebral column.

In 1995 Kokubun[10] described a case report of spinal osteotomy for vertebral column shortening in a patient with a low-lying conus medullaris. Grande et al.[6] used a cadaveric model to examine the effect of vertebral column shortening at the thoracolumbar junction on the spinal cord, lumbosacral nerve root, and filum tension. They found that shortening the vertebral column by 15–25 mm significantly reduces tension on the spinal cord, nerve roots, and filum terminale. To achieve this same effect by a traditional detethering procedure would require the release of more than 90% of the neural elements.

Since Kokubun's original report, 2 other papers have been published in the English language describing vertebral column shortening procedures for the treatment of tethered cord.[8,13] All procedures were performed at the thoracolumbar junction (4 at L-1 and 1 at T-12). There are several advantages in performing a VCR at these levels. Candidates for this procedure have often undergone several detethering procedures in the lumbosacral region. Typically, both intra- and extradural scar tissues are present, which can buckle during closure of the osteotomy and cause neurological injury.[14] Vertebral column resection also avoids the reopening of friable dura, which poses a challenge to watertight closure and leads to CSF leakage. The thoracolumbar junction has minimal curvature, which makes approaching and closing the osteotomy technically easier. Furthermore, VCR at the thoracolumbar junction minimizes the loss of motion that would occur if the procedure were performed at lower lumbar levels.

Miyakoshi and colleagues'[13] 3 cases were initially treated for TCS. Because of VCR's technical demands and relatively limited track record, it is our practice to reserve this resection as an option in patients with multiple retetherings who have been treated with traditional detethering. We also consider VCR in patients in whom traditional detethering has failed or the risk of neurological decline with further detethering is thought to be elevated. Further experience will determine optimal indications for VCR. The procedure has been highly successful in a limited number of patients thus far and may become a more common operation for TCS in the future.

Illustrative Case

History and Examination. A 37-year-old woman with a history of spina bifida and 4 previous tethered cord releases presented with the chief symptoms of back and bilateral leg pain, which occurred immediately after a child ran into her knocking her to the ground. Her symptoms progressively worsened with increased pain, numbness, and decreased gait and urinary function. Her surgical history was also notable for intrathecal pain pump placement at the time of the last detethering procedure. Neurological examination revealed 4/5 strength on left dorsiflexion, plantar flexion, and extensor hallucis longus muscle function. Sensory examination revealed numbness in L-5 and S-1 dermatomes with absent Achilles reflexes. She stood in neutral coronal and sagittal alignment and slightly favored her left leg on gait testing.

A review of radiographic images revealed multilevel midline defects in her lumbar spine consistent with laminectomy defects. Magnetic resonance imaging suggested ventral tethering of the cord at the L5–S1 level and dorsally at the L3–4 level with a degenerated disc at the L4–5 level (Fig. 1). Standing scoliosis radiographs showed normal coronal and sagittal alignment; no motion was noted on lumbar flexion and extension radiographs.

After a thorough discussion of the risks and benefits of a traditional tethered cord release versus VCR, the patient elected to undergo a T-12 VCR with a T10–L2 posterior spinal instrumentation and fusion.

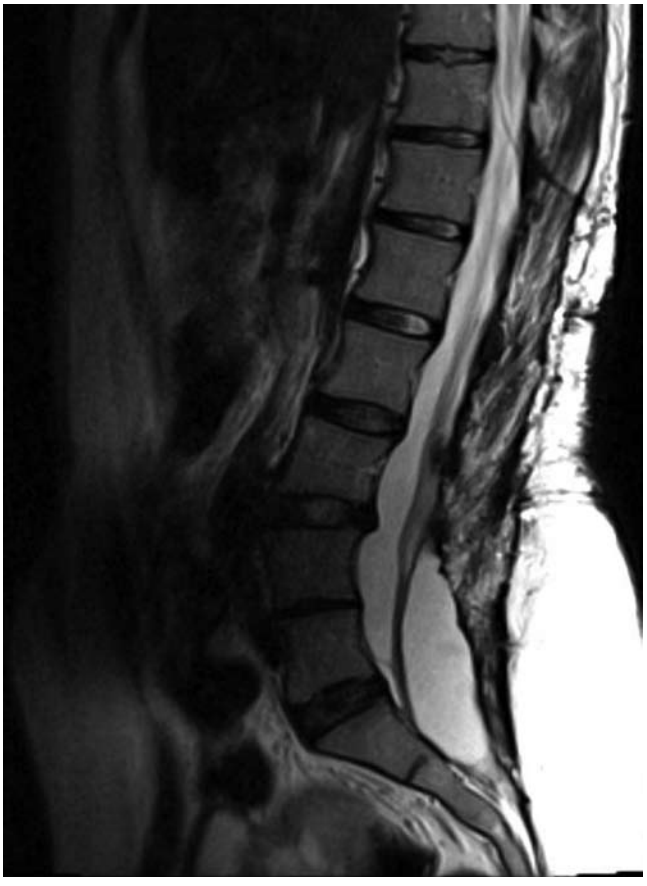

Fig. 1. Magnetic resonance image of the lumbar spine revealing a low-lying conus and ventral tethering of the cord at the L5–S1 level.

Surgical Technique. After inducing general endotracheal anesthesia, the patient was placed in a Mayfield head holder and carefully positioned prone on a Jackson frame. (As this patient population can have shunted hydrocephalus, take care not to injure any shunt apparatus with the Mayfield fixation.) All pressure points were carefully padded. After prepping and draping in a standard fashion, a midline incision was made. Her intrathecal catheter was identified and preserved during the operation. Subperiosteal dissection was performed from T10–L2. Radiographic verification of all levels was obtained. Pedicle screws were placed from T-10 to L-2 using a freehand technique. A T-12 and partial T-11 laminectomy was performed while saving bone for an autograft. The pedicles were then resected flush with the vertebral body. The T-12 rib was identified and the rib head was resected. The T11–12 disc space was identified. At this stage, 1 cm of the cancellous vertebral body was resected bilaterally using a drill, curettes, and pituitary rongeurs, while saving cancellous bone for use as an autograft. Temporary rods were placed for column stability while performing the osteotomy. A T11–12 discectomy and T-12 superior endplate resection was performed. The defect measured 20 mm at this point and was believed to be sufficient to achieve the surgical goals. The osteotomy was confirmed to be symmetric on both sides. Lateral vertebral body walls were resected to match the defect in the cancellous bone. A posterior vertebral body wall impactor was positioned ventral to the dura and used to disengage the posterior vertebral body wall, including the T11–12 disc. The posterior wall was impacted anteriorly into the osteotomy defect and was carefully removed. A Woodson elevator confirmed the neural elements were free of residual bone and disc material. The osteotomy was then closed using temporary rods, with careful attention paid to the amount of dural buckling. Somatosensory and motor evoked potentials were monitored throughout the osteotomy closure. No electrophysiological changes were noted. Permanent rods were then placed, and the osteotomy was inspected and confirmed to be bone on bone. Intraoperative CT scanning confirmed appropriate placement of the pedicle screws and bone-on-bone contact of the osteotomy. Posterior arthrodesis was performed. A large amount of locally harvested autograft from the osteotomy acted as the primary graft material. A subfascial drain was placed and the wound was closed in standard fashion.

Postoperative Course. Postoperatively, the patient noted improvement in her neurological status. Her leg pain and urological function returned to baseline. Standing scoliosis radiographs demonstrated bone-on-bone contact for the osteotomy and solid fixation of spinal instrumentation (Fig. 2).

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

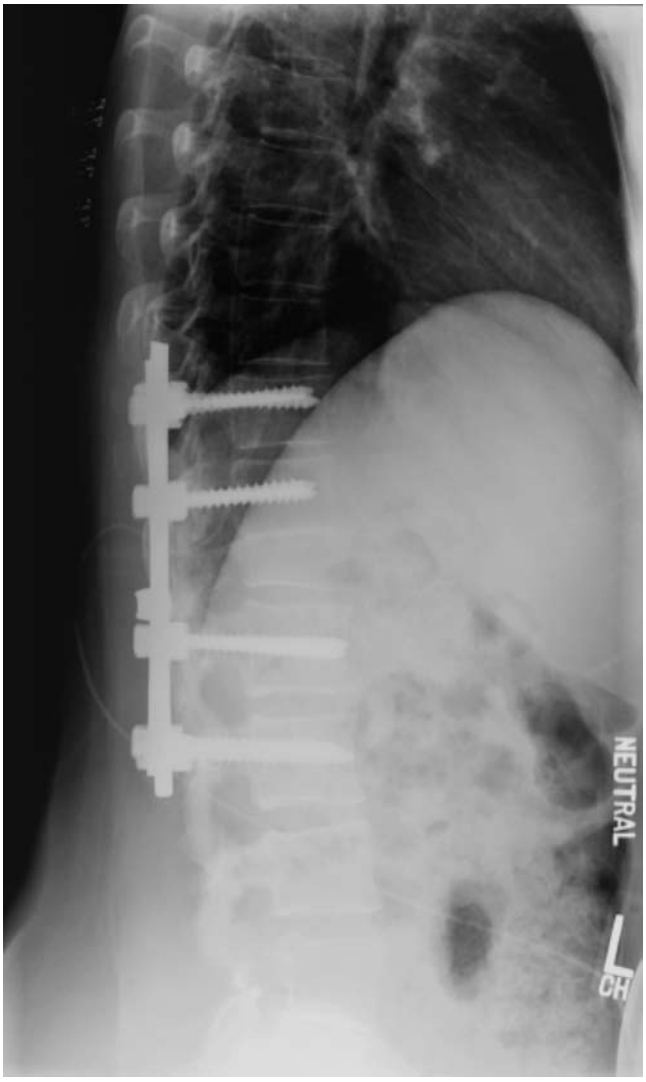

Fig. 2. Lateral plain radiograph showing bone-on-bone contact in the region of the T-12 VCR with posterior segmental instrumentation.

include the following. Conception and design: Shih. Drafting the article: Shih, Halpin, Liu. Critically revising the article: Koski, Ganju. Reviewed final version of the manuscript and approved it for submission: Koski.

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Posterior vertebral column subtraction osteotomy for the treatment of tethered cord syndrome: review of the literature and clinical outcomes of all cases reported to date

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Tethered cord syndrome (TCS) is a debilitating condition of progressive neurological decline caused by pathological, longitudinal traction on the spinal cord. Surgical detethering of the involved neural structures is the classic method of treatment for lumbosacral TCS, although symptomatic retethering has been reported in 5%–50% of patients following initial release. Subsequent operations in patients with complex lumbosacral dysraphic lesions are fraught with difficulty, and improvements in neurological function are modest while the risk of complications is high. In 1995, Kokubun described an alternative spine-shortening procedure for the management of TCS. Conducted via a single posterior approach, the operation relies on spinal column shortening to relieve indirectly the tension placed on the tethered neural elements. In a cadaveric model of TCS, Grande and colleagues further demonstrated that a 15–25-mm thoracolumbar subtraction osteotomy effectively reduces spinal cord, lumbosacral nerve root, and filum terminale tension. Despite its theoretical appeal, only 18 reports of the use of posterior vertebral column subtraction osteotomy for TCS treatment have been published since its original description. In this review, the authors analyze the relevant clinical characteristics, operative data, and postoperative outcomes of all 18 reported cases and review the role of posterior vertebral column subtraction osteotomy in the surgical management of primary and recurrent TCS. *(DOI: 10.3171/2010.4.FOCUS1070)*

Key Words • tethered cord syndrome • posterior vertebral column subtraction osteotomy • review

Lumbosacral TCS describes a constellation of clinical signs and symptoms caused by abnormal longitudinal traction on the caudal end of the spinal cord (Fig. 1).[55] The condition results from a wide array of congenital and acquired pathological processes, the majority of which act to anchor the spinal cord within the lumbosacral region of the spinal canal.[1,13] Common presenting neurological signs and symptoms are representative of the lumbosacral spinal cord levels affected: lower-extremity motor and sensory deficits; low-back, perineal, and lower-extremity pain; urinary and bowel incontinence; and sexual dysfunction.[41] Since progressive neurological, orthopedic, and urological complications can arise in the absence of surgical intervention, prompt correction of the responsible lesion is necessary.[1,7] Regardless of the procedure employed, the aim of surgery is to free the spinal cord and neural elements from tension to restore neurological function and prevent further deterioration. The traditional surgical treatment for TCS is a detethering procedure, although recurrent TCS develops in 5%–50% of patients following this initial operation.[2,10,14,21,31,33,35,38,44,54] Due in part to scar formation and arachnoid adhesions, surgery for recurrent TCS is difficult and patients are at risk for developing further neurological injury, CSF leakage, pseudomeningocele, and wound complications.[14,21,33,45] Recognizing

Abbreviations used in this paper: PVCSO = posterior vertebral column subtraction osteotomy; rhBMP-2 = recombinant human bone morphogenetic protein–2; TCS = tethered cord syndrome; VB = vertebral body; VCSO = vertebral column subtraction osteotomy.

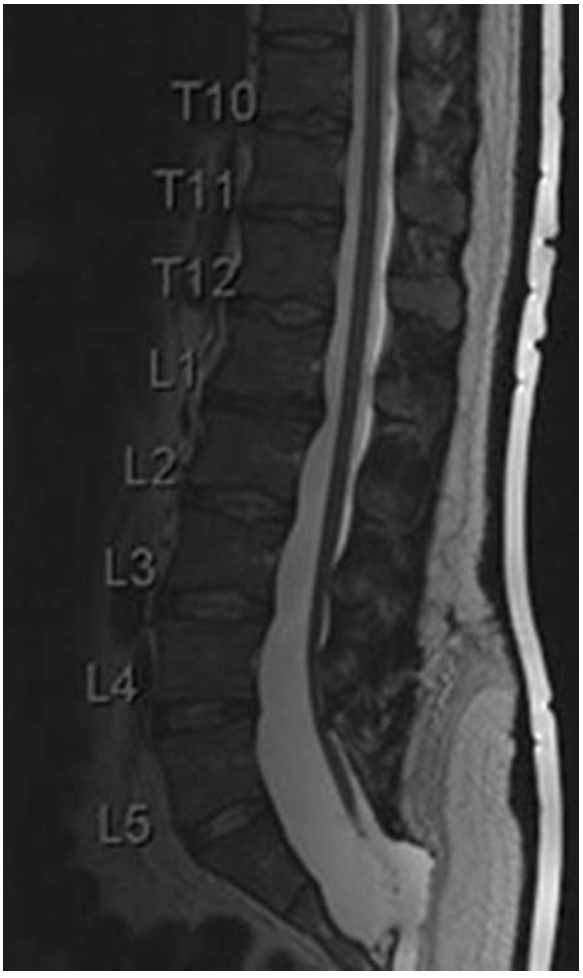

Fig. 1. Preoperative sagittal T2-weighted MR image demonstrating a low-lying conus medullaris and a recurrent tethered cord at L4–5 in a patient who had undergone previous multiple detethering surgeries. Reproduced with permission from Hsieh PC, et al: **J Neurosurg Spine 10:**278–286, 2009.

the drawbacks of these traditional detethering operations, Kokubun et al.[28,29] pioneered a spine-shortening procedure to relieve the longitudinal tension placed on the tethered neural elements without violating the dura. The operation is conducted via a posterior approach and a vertebral osteotomy is performed to facilitate spinal column shortening (Figs. 2 and 3). A detailed search of the neurosurgical and orthopedic literature reveals that posterior vertebral column subtraction osteotomy (PVCSO) has been used in 18 patients with TCS since its initial description. In this review, we provide an analysis of the relevant clinical characteristics, operative details, and postoperative outcomes in these patients and assess the role of PVCSO in the treatment of primary and recurrent TCS.

Methods

A PubMed (National Library of Medicine) search was performed to identify all articles pertaining to the use of spinal column shortening for the treatment of primary or recurrent lumbosacral TCS. Surgical series describing neurological outcomes were analyzed in detail and reference lists were reviewed for additional articles not identified in the original PubMed search. In the event that patient populations overlapped,[28,51] the series with the larger cohort of patients was selected for further review. Pertinent clinical characteristics extracted from each report included patient age and sex, presenting neurological complaints, etiology of TCS, additional spinal pathology, and prior corrective procedures. Regarding operative data and postoperative outcomes, osteotomy level, length of spinal column shortening, duration of follow-up, recovered neurological functions, and residual neurological deficits were recorded.

Articles originally published in Japanese were translated to English by a professional translation service.

Surgical Technique and Operative Details

The following describes our approach to the PVCSO for a patient with primary or recurrent TCS without additional spinal column pathology.[16] For optimal reduction of tension on the conus medullaris, filum terminale, and spinal cord, PVCSO is performed in the lower thoracic or upper lumbar levels.[12] The specific osteotomy level, extent of bony resection, and degree of instrumentation should be determined on a patient-by-patient basis.

The patient is placed in the prone position on a Jackson Spinal Table. Neurophysiological monitoring, including somatosensory evoked potentials, motor evoked potentials, and free-run electromyography, is performed.[42] A midline incision is made to expose the spine 2 levels rostral and 2 levels caudal to the planned PVCSO site (that is, T-12). We prefer to perform the osteotomy at the thoracolumbar junction. With the need to stabilize the spine 2 levels above and below to achieve immediate postoperative spinal stability for early mobilization, a T-11 or T-12 osteotomy allows one to spare more mobile segments in the lumbar spine. In addition, the regional spinal profile is neutral on both the sagittal and coronal plane. Therefore, a single-plane correctional maneuver to shorten the spinal column can avoid potential long-term decompensation of global spinal balance.

Following wide exposure of the T-12 segment, bilateral pedicle screws are implanted at T-10, T-11, L-1, and L-2, allowing for the placement of a relatively short yet biomechanically stable construct. To expose the lateral T-12 VB, the bilateral rib complexes, including the rib head and neck, and bilateral transverse processes at T-12 are removed (Fig. 2). Careful subperiosteal dissection along the lateral VB and lateral T-12 pedicle is performed until the anterior VB is visualized. A T11–12 discectomy is then carried out posterolaterally and the T-12 pedicles are removed entirely using a combination of rongeur and high-speed drilling. Prior to initiating the osteotomy, temporary rods are placed onto the pedicle screws to prevent translation of the spine while the posterior VB is osteotomized (Fig. 3). An approximate 20-mm T-12 VB osteotomy is created using a high-speed drill, beginning with the lateral walls and then moving in a posteroanterior fashion. The anterior VB wall is resected to the anterior longitudinal ligament, which is preserved; the posterior longitudinal ligament is sacrificed during removal of the posterior VB wall. Throughout the procedure, care

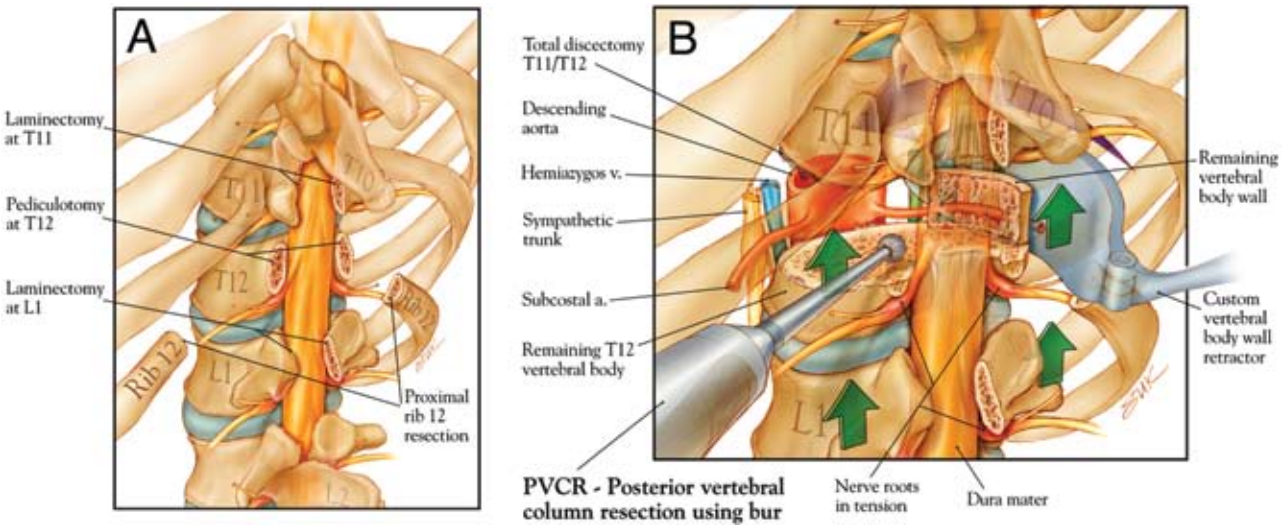

Fig. 2. Artist's illustrations of the PVCSO technique. **A:** Posterior illustration showing a T11–L1 laminectomy and resection of the proximal T-12 rib complex **B:** Posterior illustration of a T-12 PVCSO in which a high-speed drill is used to remove the T11–12 disc. a. = artery; v. = vein. Reproduced with permission from Hsieh PC, et al: **J Neurosurg Spine 10:**278–286, 2009.

is taken to avoid placing any unnecessary pressure on the thecal sac to prevent iatrogenic spinal cord injury.

Following completion of the VB resection, the temporary rods are replaced with 2 short segmental rods connected in parallel with domino connectors to facilitate safe, incremental closure of the osteotomy. Fusion across the 2 surfaces of the osteotomy can be achieved by placement of local autograft or iliac crest autograft. Alternatively, rhBMP-2 can be used to enhance bone fusion.[8,9,46] The use of rhBMP-2 is an off-label application of this product. However, clinical studies have shown that rhBMP-2 results in high rate of fusion in multisegment spinal fusions for spinal deformity without the morbidity associated with iliac crest autograft harvest. Nevertheless, the cost and complications related to the rhBMP-2, including heterotopic bone growth and osteolysis, must be taken into account. The cephalad rod is secured onto the T-10 and T11 pedicle screws while the caudal rod is secured onto the L-1 and L-2 screws. The approximate 20-mm gap created by the T11–12 discectomy and T-12 osteotomy is now closed with serial compression through the domino connectors; temporary rods are replaced with permanent rods once final spinal column alignment is achieved (Fig. 3). Posterolateral arthrodesis is performed and the wound is closed in anatomical layers.

Many patients in whom PVCSO is indicated have undergone prior spinal column operations. In these cases, the PVCSO should be performed away from the prior operative site, as scar formation and arachnoid adhesions can place the neural structures at risk for injury during spinal shortening.[40]

Tethered Cord Syndrome: Pathophysiology, Clinical Presentation, and Treatment Options

Tethered cord syndrome is multifold in its etiology, yet what constitutes a tethered spinal cord has been the subject of much debate due in part to a limited understanding of the underlying pathophysiology. When Hoffman et al.[15] published their account of 31 cases of TCS in 1976, only those patients with radiographic evidence of a low-lying conus medullaris secondary to a thick filum terminale were included in the analysis.[1,58] Patients with lipomyelomeningoceles, myelomeningoceles, diastematomyelia, or other space-occupying lesions of the lumbosacral region were excluded from the study, despite Lichtenstein's observations in 1940 that lumbosacral masses can pathologically tether the caudal spinal cord.[58] In 1981, Yamada et al.[61] established experimentally in human and animal models of TCS that excessive stretching of the spinal neural elements impairs normal neuronal oxidative metabolism and overall electrophysiological functioning, therein providing a metabolic cause to a functional neurological effect. Their investigations further demonstrated that surgical release of the tethered neural structures restores oxidative metabolism with subsequent restoration of neurological function, a finding neurosurgeons had observed for many years despite little knowledge of the molecular pathophysiology.[55] With a greater understanding of the subtle molecular defects present in TCS, the definition of a tethered spinal cord would gradually come to encompass the spectrum of lesions initially excluded from Hoffman and colleagues' analysis.[1,56,59]

The presenting signs and symptoms of TCS are almost as diverse as its causes, and although commonalities exist, the clinical presentation varies by age.[41,60] Infants and children classically present with foot and spinal deformity and often have cutaneous manifestations of spinal dysraphism, such as lumbosacral capillary hemangiomas or tufts of hair. Urinary symptoms are common and manifest as excessive dribbling and delayed toilet training. Lower-extremity motor deficits typically result in gait abnormalities. Neurological symptoms are aggravated by growth spurts, which place excessive tension on the tethered spinal cord.[7,42] In contrast to children, adults present more frequently with complaints of pain in the low back, anus and perineum, and lower extremities. Motor deficits

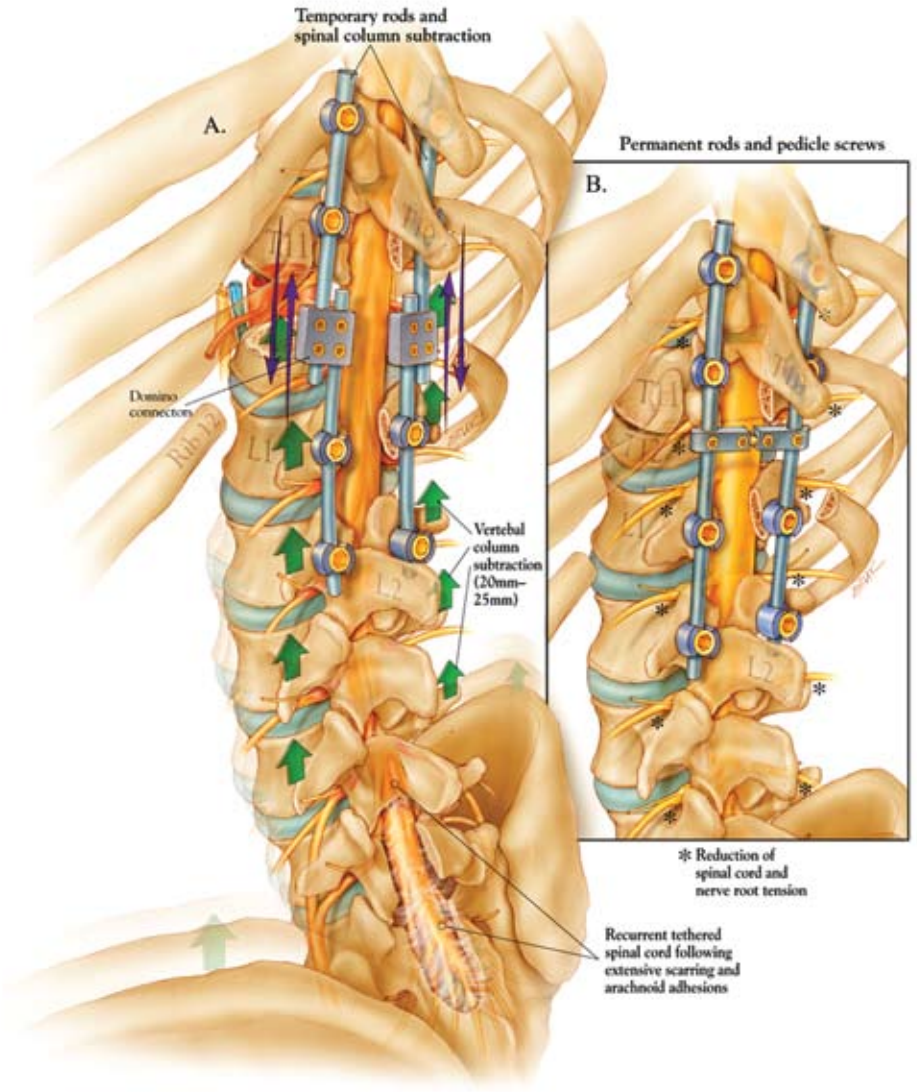

Fig. 3. Artist's illustrations of the surgical technique involved in osteotomy closure. **A:** Closure of the PVCSO is accomplished through careful serial compressions across the domino connectors. Note that closure of the osteotomy results in overall shortening of the spinal column and relaxation of distal spinal cord, nerve root, and terminal filum tension. **B:** The final instrumented spinal construct following closure of the PVCSO. Reproduced with permission from Hsieh PC, et al: **J Neurosurg Spine 10:**278–286, 2009.

manifest as pure lower-extremity weakness. As with children, urological symptoms are common and frequency, urgency, and incontinence are typical complaints. Factors that precipitate symptom onset include trauma, disc herniation, lumbar spondylosis, spinal canal stenosis, and excessive lumbar flexion or extension.[13,20,41,57]

In children and adults, surgical management of TCS is warranted at the first sign of neurological impairment.[31] The classic operation is a detethering procedure, in which the tethered neural structures are meticulously freed from their attachments, and this is followed by correction or removal of any associated pathology. In most cases, the filum terminale is severed or pathological adhesions around the distal spinal cord are detethered to reduce tension on the caudal spinal cord and associated neural elements. To avoid postoperative CSF leakage and pseudomeningocele formation, care is taken to close the dura in a watertight fashion.[1,31,32] Most patients experience an improvement in their preoperative neurological symptoms following first-time release; however, favorable recoveries decline with additional release operations.[33,53] In severe cases of recurrent TCS, Blount et al.[4] have advocated for complete spinal cord transection as a definitive measure of preserving more rostral neurological function. They have argued that traditional detethering procedures leave the freed neural structures approximated too closely to the incised dura, which allows for repeated retethering during the normal wound healing process. The procedure is reserved for selected cases in which the spinal cord and filum terminale are nonfunctional and in which the main goal of surgery is not to restore neurological function but to prevent further decline. Despite these available surgical options, a

nontrivial percentage of patients still suffer progressive neurological deterioration from recurrent TCS.

Posterior Vertebral Column Subtraction Osteotomy

Spine-shortening procedures were first used for the correction of various deformities of the spinal column.[3,5,22,47–50] The operations were initially conducted via dual anterior and posterior approaches and consisted of either wedge osteotomies or VB resection for spinal column shortening. Following critical advances in the technology of spine instrumentation, a single posterior approach for spine-shortening operations was developed.[29,47–49] Classic indications for spinal column shortening include severe scoliosis, kyphosis, and VB fractures.[19,23,29,43] In 1995, Kokubun[28] described the use of a spine-shortening procedure performed via a posterior approach for the treatment of TCS in 2 patients. The operation was conducted in lieu of a traditional detethering procedure as a means of relieving pathological tension on the tethered spinal cord and nerve roots, and was the first report of an entirely extradural procedure for TCS treatment. Although only 1 patient experienced complete neurological recovery, the initial results were promising and provided cause for further investigation into this novel technique. Subsequently, using a cadaveric model of TCS, Grande et al.[12] demonstrated that greater than 90% of the neural elements require release by traditional surgical detethering to achieve the same decrease in tension on the spinal cord, nerve roots, and filum terminale as a 15–25-mm VCSO at the thoracolumbar junction. Since efficient release of the tension placed on the tethered neural structures is necessary for improved oxidative metabolism and electrophysiological functioning,[61] PVCSO may be a preferable alternative to traditional detethering operations (Figs. 4 and 5).

Case Series and Outcome Data

Table 1 summarizes the relevant clinical characteristics of the patients described in previously published case reports that provided significant clinical data. There are currently a total of 18 patients who underwent PVCSO for TCS treatment with significant clinical data provided in the previous publications.[16,22,28,34,37,39,51,52] The mean age at the time of surgery was 40 years (range 12–57 years). Prior to PVCSO, 5 patients (28%) underwent traditional detethering operations and 5 patients (28%) underwent excision of the lumbosacral mass. Lumbosacral lipomyelomeningocele (61%) and lumbosacral lipoma (28%) were the most common causes of TCS. One patient (6%) had a VB fracture and 1 patient (6%) suffered from severe kyphoscoliosis with an intramedullary syrinx in addition to a tethered spinal cord. Lumbosacral spina bifida was identified in 5 patients (28%). Presenting neurological symptoms included urinary dysfunction (94%), lower-extremity sensory disturbances (89%), lower-extremity motor disturbances and/or gait abnormalities (78%), low-back and/or lower-extremity pain (72%), bowel incontinence (11%), and sexual dysfunction (11%). In 2 instances, the decision to perform PVCSO was motivated by marked worsening of the patient's neurological status following a traditional detethering procedure;[34,39] in these cases, the duration to PVCSO was 3 months. In these patients, the primary goal of PVCSO was reportedly to prevent further neurological deterioration rather than to offer functional recovery.

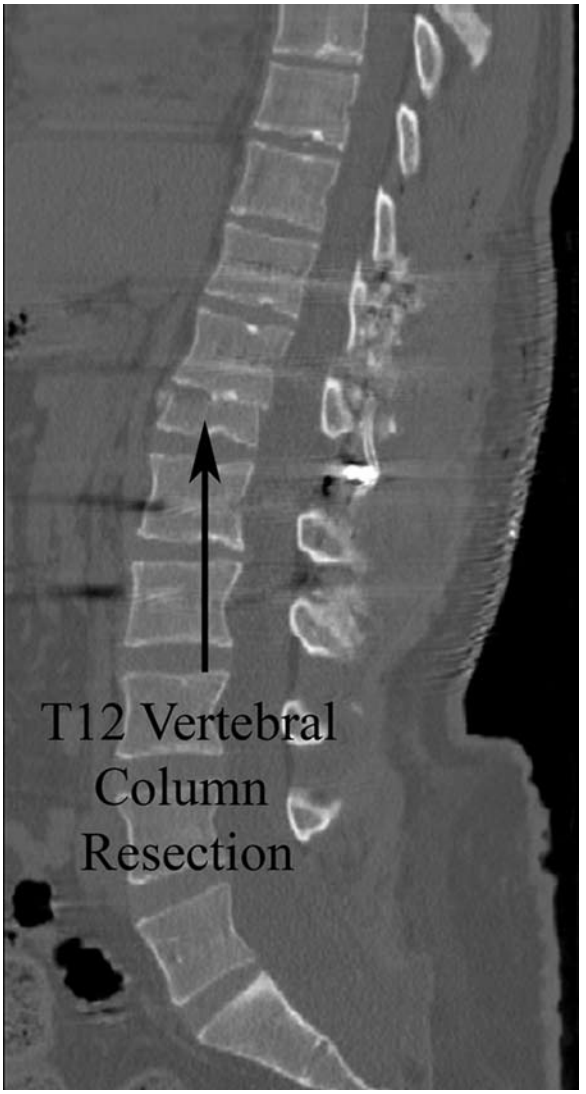

Fig. 4. Postoperative sagittal CT reconstruction demonstrating a T-12 PVCSO and closure of the osteotomy in anatomical alignment along the thoracolumbar junction. Reproduced with permission from Hsieh PC, et al: **J Neurosurg Spine 10:**278–286, 2009.

Pertinent operative data and postoperative outcomes are summarized in Table 2. No intraoperative or immediate postoperative complications were reported. The vertebral osteotomy was performed at L-1 in 14 patients (78%), at T-12 in 2 patients (11%), and at T-9 and L-2 in 1 patient each (6%). The length of spinal column shortening was reported for 16 patients (mean length 20.5 mm). The mean follow-up duration was 3 years (range 6 months–10 years). Urinary dysfunction (17 cases) was the most common residual deficit, as only 3 patients (18%) noted a full recovery of function. Two (12%) of 17 patients with urological symptoms reported improvements, although deficits remained at the time of last follow-up. Twelve (86%) of 16 patients with lower-extremity motor dysfunction improved and 8 (50%) noted complete resolution of preoperative lower-extremity sensory symptoms. Nine patients

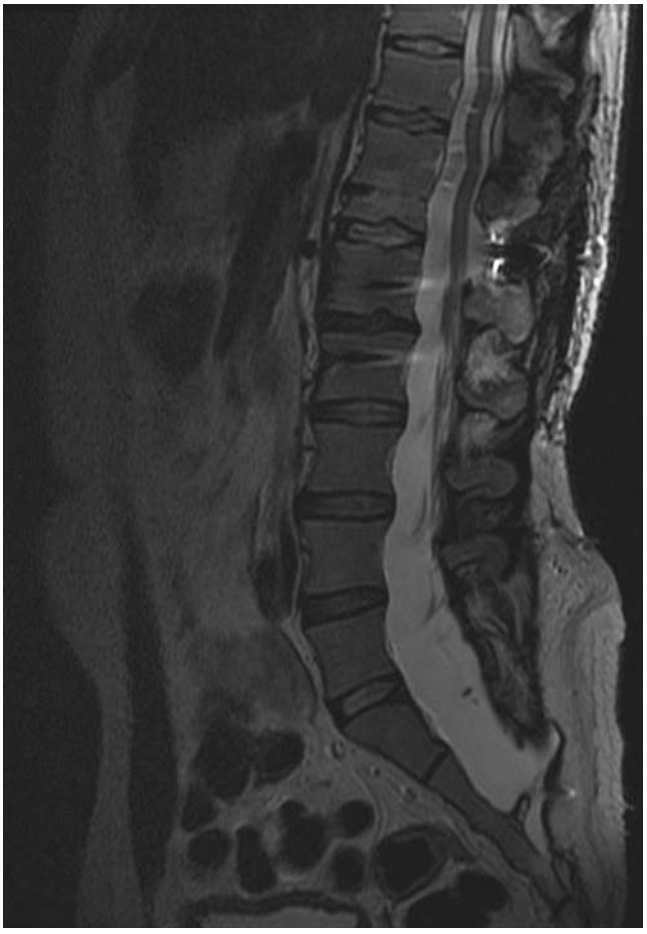

Fig. 5. Postoperative sagittal T2-weighted MR image obtained 1 year following PSVCO for TCS, showing relaxation of the distal spinal cord and cauda equina without evidence of spinal cord kinking. There is bone-on-bone approximation and fusion at the PVCSO site without subluxation or abnormal regional spinal alignment. Reproduced with permission from Hsieh PC, et al: **J Neurosurg Spine 10:**278–286, 2009.

(75%) reported immediate low-back or lower-extremity pain relief following PVCSO. Two patients (100%) reported improved sexual functioning, although only 1 regained complete erectile capabilities. Two patients (11%) experienced little symptomatic or functional benefit from PVCSO. In these cases, however, the main aim of surgery was to prevent further worsening of the neurological condition, as each had experienced new-onset neurological dysfunction following a detethering operation shortly before undergoing spinal column shortening. Where reported, complete bone union at the osteotomy site was noted in all cases at the time of last follow-up.[16,22,28,37,39,51]

Discussion

The surgical management of both primary and recurrent lumbosacral TCS is complex, yet without intervention patients will suffer progressive neurological, orthopedic, and urological decline.[1,21,33] Several studies have evaluated the symptomatic and functional outcome of adults undergoing detethering procedures for primary lumbosacral TCS and collectively indicate a 38.5%–61% improvement in urinary symptoms, a 44%–70% improvement in weakness/motor disturbances, and an 81%–91% improvement in pain following surgical detethering.[11,17,20,31,41] Despite these encouraging outcomes, a reported 5%–50% of patients eventually experience symptomatic retethering, and patients who undergo multiple detethering operations do not share this same degree of success.[31,33] Our review of the available outcome data in patients who have undergone PVCSO indicate that 28% exhibited improved urological symptoms, 86% recovered complete motor functioning, and 75% were relieved of debilitating back or lower-extremity pain. Although many factors preclude a direct comparison of the symptomatic and functional outcomes following traditional surgical detethering compared with PVCSO for TCS treatment, the outcomes, grossly, are similar. Regardless, we believe that the principal benefits of PVCSO over detethering operations will be in preserving the symptomatic and functional neurological improvements with lasting results and potentially obviating the need for future reoperations. However, longer follow-up results are needed from the PVCSO series to support this belief.

In the treatment of TCS, there are several advantages of PVCSO over detethering operations. First, direct manipulation of the neural structures is not required. Because traditional detethering procedures mandate careful dissection of the tethered spinal cord and affected neural elements away from the sites of adhesion, the risk of direct neurological injury is high. Operations for recurrent TCS may be additionally complicated by extensive scar formation and arachnoid adhesions,[30] further elevating the likelihood of iatrogenic neurological injury. Second, because PVCSO is an entirely extradural operation, the risks associated with postoperative CSF leakage and pseudomeningocele formation can be largely avoided. Third, PVCSO reduces traction on the affected neural elements indirectly (Fig. 5). Given adequate bone union and a stable instrumented construct, the beneficial effects of spinal column shortening should remain permanent. As a result, patients may be able to expect sustained improvements in their neurological symptoms, and the risks and costs associated with reoperations may be avoided. These advantages are particularly relevant in patients who have undergone multiple intradural detethering procedures without success. It is our belief and practice that patients are considered for PVCSO only in recurrent cases of TCS without local mass effects or findings that are amendable to standard surgical detethering procedures. Nevertheless, our current literature review has shown that, in the Japanese literature, 5 patients have undergone primary PVCSO treatment for lumbosacral lipoma or lipomyelomeningocele with favorable outcomes. Future clinical studies with longer clinical follow-up periods will be necessary to determine if PVCSO can be as effective as the traditional detethering surgery in the primary surgical treatment for patients with TCS.

Despite the appeal of PVCSO for the treatment of primary and recurrent TCS, there are several theoretical disadvantages. First, as a result of extensive dissection and spinal manipulation, iatrogenic neurological injury may

TABLE 1: Summary of patient data and clinical characteristics*

Authors & Year	Age (yrs), Sex	Symptoms†	Duration (yrs)	Etiology of TCS‡	Prior Ops: Type, No. of Ops	Prior Ops: Interval to PVCSO¶	Additional Pathology
Nakamura et al., 1998	55, F	M; S; U	2; 3; 2 mos	lumbosacral lipoma	detethering, 1	3 mos	spina bifida, L-5
Ueno et al., 2001	50, M	E, P, S, U	4	lumbosacral lipoma	NA	NA	spina bifida, L-5
Kanno et al., 2008	57, F	S, U; M	4; 1	lumbosacral lipoma	NA	NA	VB fracture, T-12; spina bifida, L4–S1
Hsieh et al., 2009	34, M	E, P, S, U	20	lumbosacral lipomeningocele	mass excision, 1; detethering, 3	10 yrs	none
	19, M	M, S; B, P, U	15; 4	lumbosacral lipomyelomeningocele	detethering & mass excision, 2; detethering, 3	1 yr	none
Matsumoto et al., 2009	12, F	B, P, S, U; kypho-scoliosis	3 mos; 2	lumbosacral lipoma	mass excision, 1; detethering, 1	3 mos	81° kyphosis, T3–12; 88° scoliosis, T5–L1; intramedullary syrinx, ~T6–L5
Miyakoshi et al., 2009	22, M	P; M, U	7; 2	low-lying conus medullaris	NA	NA	spina bifida, S2
	27, F	P, S	10	sacral lipoma	NA	NA	none
	13, M	M; P, U	3; 3 mos	lumbosacral lipomyelomeningocele	NA	NA	spina bifida, L5–sacrum

Authors & Year	No. of Cases	Mean Age, Range (yrs)	Symptoms (no. of cases)†	Mean Duration (yrs)	Etiology of TCS‡	Prior Ops: No. of Patients	Prior Ops: Type, No. of Ops
Tanaka et al., 2005	9	28, 15–53	M (9), P (6), S (9), U (9)	6	lumbosacral lipomyelomeningo-cele	2; 1	mass excision, 1; detethering, 4

* B = bowel incontinence; E = erectile/sexual dysfunction; M = motor disturbance; NA = not available; P = pain; S = sensory disturbance; U = urinary dysfunction.

† The reported duration indicates the approximate time from symptom onset to PVCSO; the actual duration and degree of neurological dysfunction were variable in patients who underwent prior procedures.

‡ All cases were reportedly secondary to low-lying conus medullaris syndrome rather than a documented thickened filum terminale.

¶ Interval to PVCSO refers to the time period between the patient's last operation and PVCSO.

TABLE 2: Operative data and clinical outcomes*

Authors & Year	Age (yrs), Sex	Level of Osteotomy	Spinal Shortening (mm)	FU (yrs)	Recovered Functions	Residual Deficits
Nakamura et al., 1998	55, F	L-2	19	2	none	M, S, U
Ueno et al., 2001	50, M	L-1	20	0.5	E, P	S, U
Kanno et al., 2008	57, F	T-12	22	2	M, S	U
Hsieh et al., 2009	34, M	T-12	25	1.25	E, M, P, S, U	E†, U†
	19, M	L-1	17	1.5	B, M, P, S, U	U†
Matsumoto et al., 2009	12, F	T-9	21	1.67	none‡	B, P, U
Miyakoshi et al., 2009	22, M	L-1	20	5	M, P, U	none
	27, F	L-1	NA	4	P, S	none
	13, M	L-1	NA	3	M, P, U	none

Authors & Year	No. of Cases	Mean Age (range) in Yrs	Level of Osteotomy	Mean Spinal Shortening (range) in mm	Mean FU (range) in Yrs	Recovered Functions (no.)	Residual Deficits (no.)¶
Tanaka et al., 2005	9	28 (15–53)	L-1	20 (14–23)	3 (1–10)	M (7), P (4), S (4), U (1)	M (2), P (2), S (5), U (8)

* FU = follow-up.
† Although deficits remain, these patients did note a slight improvement in neurological function postoperatively.
‡ Despite no improvement in neurological function, the patient's kyphoscoliosis was effectively corrected by PVCSO without new neurological injury.
¶ In one patient progressive paralysis began developing 4 months postoperatively.

arise from excessive nerve root traction, dural violation, or exacerbation of prior spinal deformities.[18,24] In an analysis of the risk of neurological injury following lumbar pedicle subtraction osteotomy, reported by Buchowski et al.,[6] 3% of patients experienced permanent new-onset neurological deficits. Second, because the osteotomy involves all 3 spinal columns, the spine is highly unstable following PVCSO, and translation, subluxation, or misalignment can occur if the gap closure is inadequate, or if pseudarthrosis develops. Despite the use of an instrumented construct for spinal stabilization, pseudarthrosis has been found in 17%–24% of cases involving multisegmental, instrumented fusion.[25–27] Such complications increase the risk of instrumentation failure, neurological injury, spinal deformity, and chronic back pain. The complication rate associated with a similar technique of pedicle subtraction osteotomy in deformity revisions can be as high as 50%, but much of those complications are related to medical complications resulting from performing high-risk complex spinal surgeries in a more elderly population. Third, instrumented fusion of the spinal column is required with PVCSO. To ensure biomechanical stability, our protocol is to provide fixation 2 levels rostral and caudal to the osteotomy, necessitating a 4-level fusion. A multisegmented, instrumented fusion predisposes the patient to future disease in adjacent segments.[36] Fourth, vertebral column resection requires extensive paraspinal dissection and the amount of perioperative blood loss is not trivial. This can be minimized with careful subperiosteal dissection along the VB wall, which reduces injury to segmental vessels. Despite these potential drawbacks and the inherent technical demands of the operation, the procedure can be performed safely and with acceptable surgical risks at experienced centers.

Our analysis of data obtained in 18 patients reported on in the literature indicates that PVCSO is a viable alternative to traditional detethering operations for the management of primary and recurrent TCS. Future outcome studies with larger sample sizes and long-term follow-up are needed to further clarify the risks and benefits of the procedure and to identify specific patient groups most likely to benefit from PVCSO.

Conclusions

Tethered cord syndrome is a progressively debilitating neurological condition caused by abnormal longitudinal traction on the caudal spinal cord. The classic treatment is surgical detethering, although rates of symptomatic retethering range from 5%–50%. With subsequent operations, surgical complications increase while favorable neurological outcomes decrease. The present analysis indicates that PVCSO is a safe and effective alternative to traditional detethering operations for the treatment of primary and recurrent TCS. Not only does the procedure avoid the risks inherent in any intradural operation, but cadaveric studies also indicate that extensive surgical detethering is necessary to achieve results similar to those from a 15–25-mm thoracolumbar PVCSO. Because the use of PVCSO for TCS treatment is in its infancy, future studies with a larger number of patients and longer follow-

up periods are needed to verify its efficacy and identify patients most likely to benefit from this novel approach.

Disclosure

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Minimally invasive tethered cord release in adults: a comparison of open and mini-open approaches

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Object. Symptomatic tethered cord and associated anomalies such as diastematomyelia rarely present during adulthood but can cause significant pain as well as motor, sensory, and bladder dysfunction. As with children, studies have shown that surgical detethering may provide improvement in pain and neurological deficits. Typical surgical management involves an open laminectomy, sectioning of the filum terminale, and exploration of the split cord malformation. Such open approaches, however, cause significant paraspinous muscle trauma and scarring. Recent advances in minimally invasive techniques allow for access to the spine and thecal sac while minimizing associated muscular trauma. The authors present a comparison of open versus minimally invasive surgery to treat adult tethered cord syndrome.

Methods. Six adult patients underwent surgical release of a tethered spinal cord (2 of them also had diastematomyelia). The mean age of the patients was 47.78 years (range 31–64 years). All medical records and images were retrospectively reviewed. Three of the patients underwent traditional open laminectomies for detethering (open group) while the other 3 patients underwent minimally invasive (mini-open) spinal cord detethering. The length of the incision, length of stay, estimated blood loss, and complications were compared between the 2 groups.

Results. All 6 patients had tethered spinal cords, and 1 patient in each group had diastematomyelia. The mean estimated blood loss during surgery (300 ml in the open group vs 167 ml in the mini-open group, $p = 0.313$) and the mean length of stay (7 days in the open group vs 6.3 days in the mini-open group, $p = 0.718$) were similar between the 2 groups. The incision length was half as long in the mini-open group versus the open group. However, 1 patient in the mini-open group developed a postoperative pseudomeningocele requiring surgical revision, whereas the open group had no revision surgeries.

Conclusions. Cases of symptomatic diastematomyelia and tethered cord in adults can be safely and effectively explored through a mini-open approach. In this small case series, the authors did find that the mini-open group had an incision that was 50% smaller than the open group, but they did not find a significant clinical difference between the groups. *(DOI: 10.3171/2010.3.FOCUS1077)*

Key Words • **minimally invasive approach** • **diastematomyelia** • **tethered cord** • **laminectomy**

The onset of symptomatic TCS in combination with diastematomyelia during adulthood is rare.[13,21] Patients typically present with a variety of symptoms including low-back pain, leg pain, lower-extremity weakness, sensory deficits, bladder dysfunction, and/or sexual dysfunction.[1,5,8–10,14] Just as in cases of TCS in children, there is evidence that surgical detethering leads to improvement in symptoms.[1,3–5,7,8,10,14,20] A wide lumbosacral laminectomy is the standard approach used to explore the split cord malformation and to section the filum terminale. We present a comparison of the use of a minimally invasive (mini-open) approach with an expandable tubular retractor versus an open approach to treat 6 adults (3 in each group) with TCS (with and without diastematomyelia).

Abbreviations used in this paper: EBL = estimated blood loss; LOS = length of stay; TCS = tethered cord syndrome.

Methods

This retrospective study was approved by the University of California, San Francisco, Committee on Human Research. Six adult patients were evaluated for symptomatic TCS. The mean age was 47.7 years (range 31–64 years). All medical records and images were retrospectively reviewed.

Three of the patients underwent traditional open laminectomies for detethering (open group) while the remaining 3 patients underwent a minimally invasive approach using an expandable tubular retractor (mini-open group). The choice of the surgical approach was selected by the surgeon, and the patients were not randomized. The mean ages of the 2 groups were comparable (43.3 years in the open group and 52 years in the mini-open group, $p = 0.535$). One patient in the minimally invasive group had undergone a detethering operation as a child,

TABLE 1: Summary of patient data*

Case No.	Age (yrs)	EBL	LOS (days)	FU (mos)	Approach	Diagnosis	Specific MRI findings
1	31	200	8	53	open	TC	fatty filum; no lipoma; scoliosis
2	36	500	9	20	open	diastematomyelia w/ TC, after previous detethering operation as a child	diastematomyelia w/ fibrous band btwn hemicords; scoliosis; no lipoma
3	63	200	4	12	open	TC, after previous lipoma resection & previous detethering as a child	intradural lipoma; fatty filum
mean	43.3	300	7				
4†	37	100	7	3	mini-open	TC	fatty filum; no lipoma
5	55	200	7	15	mini-open	diastematomyelia w/ TC	diastematomyelia w/ fibrous band btwn hemicords; syringomyelia w/in conus & distal spinal cord; subcutaneous lumbosacral lipoma
6	64	200	5	12	mini-open	lipoma w/ TC	intradural lipoma attached to conus
mean	52	167	6.3				

* The mean age, EBL, and LOS of the 2 groups were not significantly different ($p > 0.05$). Abbreviations: FU = follow-up; TC = tethered cord.
† The patient in Case 4 had a postoperative pseudomeningocele requiring revision surgery.

and we performed a revision detethering of her cord. Two patients in the open group also had prior detethering surgeries in the past, and we performed revision detethering of the cord. One patient in each group had a diastematomyelia in addition to a tethered cord.

The incision length, EBL, LOS, and the postoperative complication rates were compared between the 2 groups.

Results

The mean EBL was not statistically different between groups (300 ml in the open group vs 167 ml in the mini-open group, $p = 0.313$). The mean LOS was likewise not statistically different between groups (7 days for the open group vs 6.3 days in the mini-open group, $p = 0.718$). The incision length was half the size in the minimally invasive group (4–5 cm) compared with the open group. One patient in the mini-open group developed a pseudomeningocele, requiring readmission for surgical revision while no patients in the open group suffered surgical complications (Table 1).

Illustrative Case

History and Examination. This 55-year-old woman had a 6-month history of continuous low-back and bilateral leg pain. These symptoms began without any precipitating factors. She denied any significant motor weakness, sensory deficits, or bowel/bladder dysfunction. Her surgical history was notable for a remote history of repair of a left clubfoot. She also reported that she had undergone ligation of a sacral meningeal cyst via an open lumbosacral laminectomy approach 12 years earlier. On a visual analog scale for pain (0–10, with 10 being worst), she reported a score of 10 for both back and leg pain.

The patient had a body mass index of 29.2. She was noted to have weakness of the left ankle plantar flexors and dorsiflexors. She had difficulty with tandem gait, and her ankle reflexes were absent. Her neurological examination was otherwise intact with normal strength, sensation, and reflexes in the remainder of her lower extremities. She also had evidence of a subcutaneous lumbosacral lipoma. Lumbar MR imaging revealed evidence of a tethered cord with a small syrinx at T12–L1 and diastematomyelia with a dominant right hemicord beginning at L-1 and extending to the L2–3 disc level (Fig. 1).

Operation. Under fluoroscopic guidance, a 4–5-cm midline skin incision was made from the L1–2 disc level to the L2–3 disc level (Video 1).

Video 1. Minimally invasive tethered cord release and exploration of diastomyelia in an adult. Click here to view with Windows Media Player. Click here to view with Quicktime.

The incision was extended through the fascia using monopolar electrocautery. The L-2 spinous process was removed with a rongeur. Serial tubular dilators were used to dilate the paraspinal muscles at the L-2 level, and a single expandable tubular retractor (Pipeline, DePuy Spine, Inc.) was placed. This retractor allowed exposure from the inferior aspect of the L-1 lamina to the superior aspect of the L-3 lamina. The operating microscope was then brought into the field, and a midline laminectomy of the entire L-2 lamina was performed. The dura was exposed and opened in the midline. Exploration revealed a split cord malformation at the level of L-1 (Fig. 2). The right hemicord was larger than the left and had a thickened caudal extension extending into the filum. The left hemicord rejoined the right hemicord after approximately 3 cm and had no separate filum. Several nerve roots arose from the right hemicord, and intraoperative electromyography was used to distinguish the nerve roots from the filum. Once identified, the filum was coagulated and divided. Exploration of the split cord malformation revealed a fibrous connective tissue band separating the 2 hemicords. This tissue was dissected free. The dura was closed, and a subfascial drain was placed (connected to a gravity bag). The incision was closed in layers. The total operative time was 208 minutes, and the EBL was 200 ml.

Postoperative Course. The patient maintained full

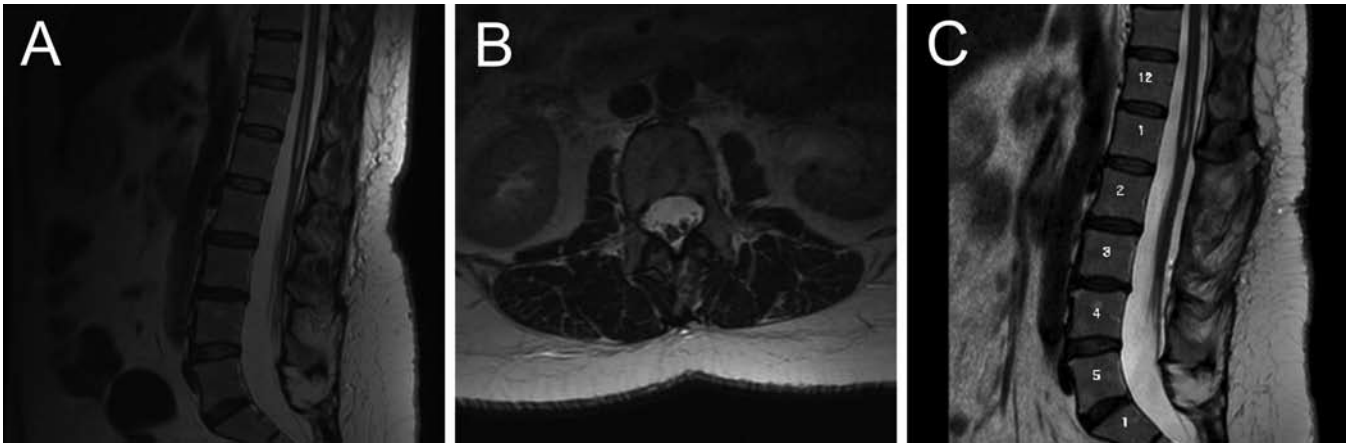

Fig. 1. A and B: Preoperative sagittal and axial T2-weighted MR images showing a tethered cord and T12–L1 syrinx **(A)** and diastematomyelia **(B)**. **C:** Postoperative sagittal T2-weighted image with fat suppression showing the complete L-2 laminectomy and detethered cord.

strength and intact sensation throughout her lower extremities. However, due to output of clear fluid from her subfascial drain, we suspected that her dural repair was not watertight. Because of this, a lumbar subarachnoid drain was placed on postoperative Day 2. Her lumbar drain was clamped after 72 hours of drainage, and it was subsequently removed after no evidence of further leakage. The patient was mobilized and discharged home on postoperative Day 7 in good condition. A postoperative MR image showed typical postoperative changes.

At the 15-month follow-up, the patient reported improvement in her preoperative symptoms, with mildly improved low-back and leg pain as well as increasing strength in her feet. She was able to heel- and toe-walk with only mild difficulty. She continued to have difficulty with tandem walking, however, which was thought to be due to her history of bilateral club feet. Her most recent postoperative visual analog scale pain score was 8 for both back and leg pain.

Discussion

Typical detethering procedures involve making a wide laminectomy for adequate exposure of the thecal sac and underlying neural elements. The muscle dissection associated with this approach can be extensive and can lead to significant blood loss and postoperative scarring. Mayer et al.[11] reported paraspinous muscle atrophy and decreased trunk strength after spinal surgery, while Sihvonen et al.[17] correlated instances of "failed back syndrome" with paraspinous muscle denervation and atrophy. They postulated that such iatrogenic paraspinous muscle injury could lead to increased biomechanical strain postoperatively. Minimally invasive approaches to the lumbar spine reduce the amount of muscle dissection and trauma. Studies have shown decreased markers of tissue injury, such as C-reactive protein, interleukin-6, and creatine-phosphokinase, when comparing minimally invasive lumbar decompression techniques with open approaches.[6,15,16]

Previous studies of surgical detethering in adult patients with a tethered cord showed that pain is the most effectively treated symptom with success rates of 48%–100%.[1,3,5,7,8,10,20] Other deficits, such as motor weakness and sphincter dysfunction, are more difficult to treat. In our illustrative case, the patient's presenting complaint was low-back and leg pain, and she was found to have distal lower-extremity weakness on preoperative examination. Her pain was mildly improved postoperatively.

Tubular retractor systems have been effectively used for intradural spinal procedures, including repair of a spinal dural arteriovenous fistula[2] and resection of intradural tumors.[12,19] These reports have shown excellent outcomes with minimal complications. Tredway et al.[18] also reported the use of a tubular retractor system for detethering of the spinal cord in 3 patients. In each case, they performed an inferior L-4 and a superior L-5 laminotomy that allowed adequate exposure for dissection of the filum and nerve roots, electromyography, and dural closure. They

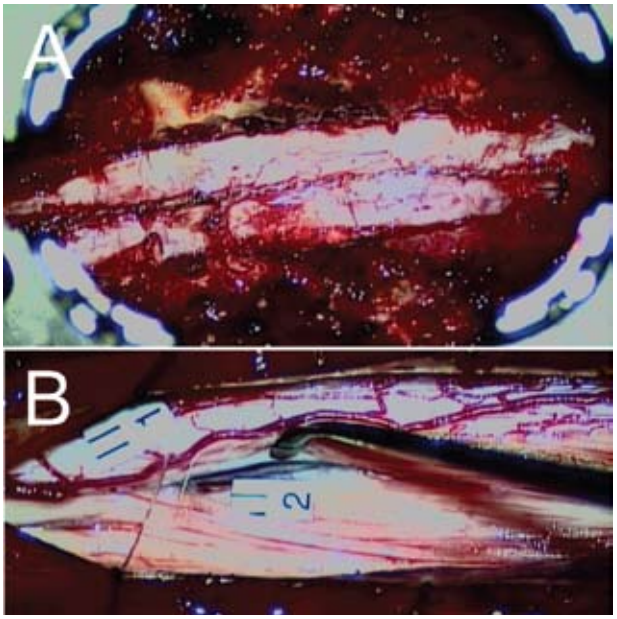

Fig. 2. Intraoperative photographs. **A:** Intraoperative view of the lumbar tethered spinal cord through the expandable retractor system. **B:** The larger right hemicord (labeled 1) can be distinguished from the smaller left hemicord (labeled 2).

reported no complications, and all 3 patients remained symptom-free at follow-up.

Our report is the first to demonstrate the use of a mini-open approach with an expandable tubular retractor to explore diastematomyelia in adults and release a tethered cord. The use of this mini-open approach allowed for a reduction in the size of the incision to 4–5 cm in a patient with a body mass index of 29.2.

Furthermore, this is the first study to compare open and minimally invasive (mini-open) tethered cord release. With such a small cohort, statistical comparisons are not ideal. Interestingly, we did not find a significant difference in intraoperative blood loss or LOS when we compared the 2 surgical approaches. The minimally invasive group had a higher complication rate as 1 patient had to have revision surgery for pseudomeningocele repair. Cerebrospinal fluid leaks have been reported after traditional open detethering procedures in the past at a rate of 5%–15%.[10,14] We have found, however, that closing the dura through a tubular retractor is technically more difficult than closing the dura in an open case. The tubular retractor limits the angles of approach with the needle driver.

The main advantage of performing this surgery through a mini-open transspinous approach is to reduce the length of the incision and the related scar tissue. However, we cannot demonstrate a clinical difference when we compared the minimally invasive approach with the open approach to treat tethered spinal cords in adults.

Conclusions

This is the first report of the use of minimally invasive techniques for both detethering of the spinal cord and exploration of an associated diastematomyelia, and to compare the results with open cases. Symptomatic diastematomyelia and tethered cord in adults can be safely and effectively explored through either an open or a minimally invasive (mini-open) approach. The length of the incision is reduced by 50% with the use of the mini-open approach, but there does not appear to be a clinically significant difference in the blood loss or LOS when comparing these approaches.

Disclosure

Dr. Mummaneni is a consultant for DePuy Spine and Medtronic. He receives financial support from DePuy Spine (royalty) and Medtronic (grant).

Author contributions to the study and manuscript preparation include the following. Conception and design: Gupta, Mummaneni. Acquisition of data: Wu, Potts. Analysis and interpretation of data: Wu, Potts. Drafting the article: Potts. Critically revising the article: Wu, Potts, Mummaneni. Reviewed final version of the manuscript and approved it for submission: Wu, Potts, Mummaneni. Statistical analysis: Wu, Potts. Study supervision: Mummaneni.

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A patient with myelomeningocele: is untethering necessary prior to scoliosis correction?

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Object. Tethering of the spinal cord is thought to increase the chance of neurological injury when scoliosis correction is undertaken. All patients with myelomeningocele (MM) are radiographically tethered, and untethering procedures carry significant morbidity risks including worsening neurological function and wound complications. No guidelines exist as regards untethering in patients with MM prior to scoliosis correction surgery. The authors' aim in this study was to evaluate their experience in patients with MM who were not untethered before scoliosis correction.

Methods. Seventeen patients with MM were retrospectively identified and 1) had no evidence of a clinically symptomatic tethered cord, 2) had undergone spinal fusion for scoliosis correction, and 3) had not been untethered for at least 1 year prior to surgery. The minimum follow-up after fusion was 2 years. Charts and radiographs were reviewed for neurological or shunt complications in the perioperative period.

Results. The average age of the patients was 12.4 years, and the following neurological levels were affected: T-12 and above, 7 patients; L-1/L-2, 6 patients; L-3, 2 patients; and L-4, 2 patients. All were radiographically tethered as confirmed on MR imaging. Fourteen of the patients (82%) had a ventriculoperitoneal shunt. The mean Cobb angle was corrected from 82° to 35°, for a 57% correction. All patients underwent neuromonitoring of their upper extremities, and some underwent lower extremity monitoring as well. Postoperatively, no patient experienced a new cranial nerve palsy, shunt malfunction, change in urological function, or upper extremity weakness/sensory loss. One patient had transient lower extremity weakness, which returned to baseline within 1 month of surgery.

Conclusions. The study results suggested that spinal cord untethering may be unnecessary in patients with MM who are undergoing scoliosis corrective surgery and do not present with clinical symptoms of a tethered cord, even though tethering is radiographically demonstrated. *(DOI: 10.3171/2010.3.FOCUS1072)*

Key Words • **myelomeningocele** • **neurological injury** • **tethered cord** • **posterior spine fusion** • **scoliosis**

Scoliosis often develops in patients with myelomeningocele.[3,9,16] Risk factors for the development of scoliosis in these patients include motor level, ambulatory status, and last intact laminar arch, with scoliosis developing in almost 100% of patients with a thoracic motor level.[9,16] In addition, virtually all patients with MM radiographically demonstrate a tethered spinal cord,[6,13] although only 10–30% exhibit symptoms.[1,2,15] This occurs due to a difficulty in closing the dura mater around the neural placode to keep it bathed in CSF and prevent adherence to the dura.

A tethered spinal cord can predispose patients to neurological injury when they undergo surgical correction of scoliosis.[5,12,17] Theoretically, this occurs due to excessive traction on a tethered spinal cord. Symptoms can include those attributed to shunt malfunction, cranial nerve palsies, upper extremity weakness, and worsening lower extremity function. However, untethering the spinal cord in patients with MM also carries some risk of morbidity including worsening neurological function and wound problems.[2,6] No reports in the literature address whether untethering is necessary in an asymptomatic patient with MM who presents for scoliosis surgery. Practice patterns vary from institution to institution with some advocating untethering of all patients and others never untethering prior to scoliosis correction. In the present report, we describe our experience with patients with MM who did not undergo untethering prior to their scoliosis surgery.

Methods

After obtaining institutional review board approval we performed a retrospective review of all patients with MM who had undergone surgery for scoliosis correction at our institution. None of the patients had been untethered for 1 year prior to their surgical procedure. Patient charts and radiographs were reviewed for the major coronal Cobb angle, sagittal kyphosis and lordosis, sagittal and coronal balance, and presence of a shunt. Intraoperative and postoperative complications were noted with

Abbreviation used in this paper: MM = myelomeningocele.

TABLE 1: Summary of patient characteristics

total no. patients	17
M	7
F	10
mean age at surgery in yrs (range)	12.4 (10–17)
mean length of follow-up in yrs (range)	3.3 (2–8)
patients w/ shunt (%)	14 (82)

particular attention devoted to shunt issues and neurological function including the brainstem, a change in urological function, and upper and/or lower extremity function.

Results

Patient Demographics

Seventeen patients—10 females and 7 males—with an MM who underwent surgery for scoliosis were identified. None of these patients presented with symptoms suggesting symptomatic tethered cord, and none had been tethered in the year preceding their scoliosis surgery. The average age was 12.4 years (range 10–17 years). The affected motor levels were thoracic (7 patients), L-1 or L-2 (6 patients), L-3 (2 patients), and L-4 (2 patients; Table 1). The major curve was lumbar in 6 patients (apex L-1/L-2 disc to L-4), thoracolumbar in 5 patients (apex T-12 to L-1), and thoracic in 6 patients (apex T-2 to T-11/T-12 disc). Fourteen patients underwent a T-2 to pelvis fusion. The remaining 3 patients underwent fusion of the following levels: T10–L4 (2 patients) and T11–L4 (1 patient).

Scoliosis Correction

The preoperative Cobb angle averaged 82° (range 56°–120°), and it measured 35° (range 20°–55°) at the last follow-up, for a 57% correction (Fig. 1). Maximal kyphosis averaged 50° (range 20°–110°) preoperatively, and improved to 33° (range 12°–66°) postoperatively (Table 2).

Neurological Complications

No patient experienced a shunt-related issue postoperatively. In addition, no new cranial nerve palsies, change in urological function, or upper extremity neurological deterioration occurred. One patient, whose MM motor level was at L-4, had moderate right quadriceps muscle weakness postoperatively, which improved to baseline function over 1 month. The etiology of the weakness was unclear as a postoperative CT was unremarkable. Although neurological complications were sparse, wound problems occurred in 4 (23%) of 17 patients, a rate similar to others reported in the literature.[8,14]

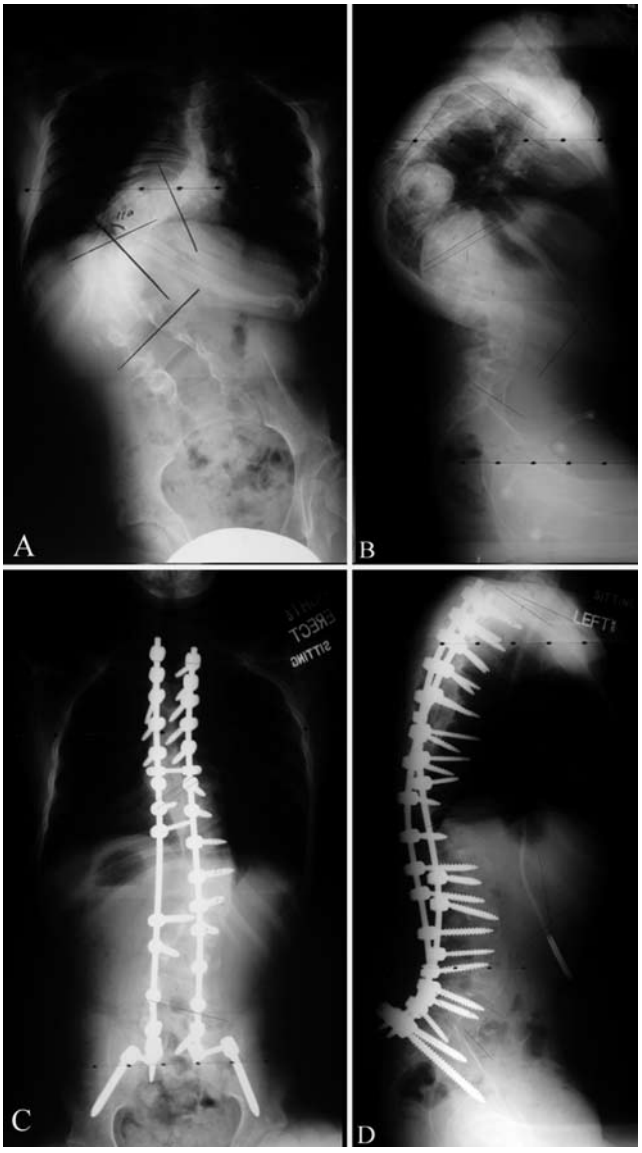

Fig. 1. Preoperative posteroanterior **(A)** and lateral **(B)** full spine radiographs obtained in a 15-year-old boy with an L-2 level myelomeningocele, showing a > 100° scoliosis curvature. The boy underwent a T-2 to pelvis posterior spine fusion without a prior untethering procedure. Postoperative posteroanterior **(C)** and lateral **(D)** radiographs showing good correction.

Discussion

Our results suggest that untethering the spinal cord in patients with MM who do not present with symptoms of a tethered cord may be unnecessary before surgical correction of their scoliosis. In our series of 17 patients, significant correction of scoliosis was achieved, with only 1 patient sustaining a temporary lower extremity deficit. No shunt problems, upper extremity weakness, or brainstem dysfunction was observed in the perioperative period. The main limitations of our study are its retrospective nature and relatively small sample size.

Virtually all patients with MM demonstrate radiographic tethering.[6,13] Theoretically, the spinal column lengthening that occurs with scoliosis correction can result in neurological deterioration. Little has been published on whether patients should undergo untethering prior to correction of their scoliosis. Al-Holou et al.[1] commented on prophylactic untethering in patients with MM undergoing scoliosis surgery. Similarly, in a subsection of their paper, Bowman et al.[2] reported on 10 children with severe sco-

TABLE 2: Deformity correction*

Deformity	Cobb Angle (°)	
	Preop	Most Recent Follow-Up
major coronal curve	82 (56–120)	35 (20–55)
kyphosis	50 (20–110)	33 (12–66)

* Values are presented as means with ranges in parentheses.

liosis who "underwent untethering immediately prior to spinal fusion to prevent neurological decline secondary to possible spinal column lengthening." At our institution we do not untether patients with MM who are undergoing scoliosis correction surgery, as long as they do not present with symptoms suggestive of a tethered cord.

Symptoms suggestive of a tethered cord in these patients include pain, weakness, gait abnormality, lower extremity hip and foot deformity, urological changes, and rapidly progressive scoliosis.[2,6] After untethering, improvements are commonly seen in pain and often in weakness, gait, and urological changes.[1,2] Results of untethering in patients with a rapidly progressive scoliosis appear to be best when the curve is < 40°–45° and in patients whose motor level is below T-12.[2,7,10,11] Pierz et al.[10] reported on 21 patients with MM and rapidly progressive scoliosis who underwent untethering. None of the patients with curves > 40° or with an affected thoracic level demonstrated any benefit. Reigel et al.[11] examined 216 patients with MM who were untethered and found none of those with thoracic level MM stabilized.

Although none of our patients experienced shunt malfunction in the early postoperative period, this complication is potentially devastating in this population. In their series of 77 patients with MM undergoing scoliosis surgery, Geiger et al.[4] described 4 patients who presented with acute postoperative shunt failure, with 1 death. The mechanism for acute shunt failure may include blood products entering the CSF and blocking the shunt, or shunt breakage from the change in alignment of the torso after scoliosis correction. Heavy narcotics for postoperative management should be used with caution to ensure adequate neurological assessment.

Conclusions

Virtually all patients with MM have a tethered spinal cord. Our results suggest that in a patient with MM who is asymptomatic, one may not need to untether the spinal cord prior to scoliosis surgery. These findings should be considered preliminary as the number of patients in our study was relatively small.

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: Samdani, Betz. Acquisition of data: Samdani, Sagoo, Cahill, Clements. Analysis and interpretation of data: Samdani, Fine, Sagoo, Shah. Drafting the article: all authors. Critically revising the article: Samdani, Fine, Shah. Reviewed final version of the manuscript and approved it for submission: Samdani, Fine, Sagoo, Cahill, Clements, Betz. Study supervision: Betz.

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Spinal cord traction, vascular compromise, hypoxia, and metabolic derangements in the pathophysiology of tethered cord syndrome

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Object. The definition of tethered cord syndrome (TCS) relies mainly on radiological criteria and clinical picture. The presence of a thickened filum terminale and a low-lying conus medullaris in symptomatic patients is indicative of TCS. The radiological definition of TCS does not take into account cases that involve a normal-lying conus medullaris exhibiting symptoms of the disease.

Methods. The authors performed a MEDLINE search using the terms "tethered cord" and "pathophysiology." The search returned a total of 134 studies. The studies were further filtered to identify mostly basic research studies in animal models or studies related to the biomechanics of the filum terminale and spinal cord.

Results. Spinal cord traction and the loss of filum terminale elasticity are the triggers that start a cascade of events occurring at the metabolic and vascular levels leading to symptoms of the disease. Traction on the caudal cord results in decreased blood flow causing metabolic derangements that culminate in motor, sensory, and urinary neurological deficits. The untethering operation restores blood flow and reverses the clinical picture in most symptomatic cases.

Conclusions. Although classically defined as a disease of a low-lying conus medullaris, the pathophysiology of TCS is much more complex and is dependent on a structural abnormality, with concomitant altered metabolic and vascular sequelae. Given the complex mechanisms underlying TCS, it is not surprising that the radiological criteria do not adequately address all presentations of the disease. *(10.3171/2010.3.FOCUS1085)*

Key Words • **filum terminale** • **metabolism** • **pathophysiology** • **tethered cord** • **traction**

Tethered cord syndrome belongs to a group of abnormalities under the term OSD.[5] Although the incidence and prevalence of OSD and primary TCS are not well established, the advent of better neuroimaging studies and improved clinical awareness has led to an increase in the number of diagnosed cases.

Tethered cord syndrome is a clinical entity presenting with neurological symptoms, urological dysfunction, and orthopedic sequelae caused by congenital or secondary factors leading to the tethering of the distal part of the spinal cord. The presence of a thickened filum terminale with a diameter greater than 2 mm and/or conus medullaris located below the level of L1–2 (Fig. 1) are the proposed radiological diagnostic criteria in cases presenting with symptoms.[1,2,9,20,30,31]

Interestingly, the literature cites a significant number of cases that do not meet the proposed criteria for filum terminale thickness of the and conus medullaris level yet the patients present clinically with symptoms of TCS.[13,17,24,25,29,32,33,36] The resection of the filum terminale, in these patients, leads to resolution of the clinical symptoms. These observations suggest that the radiographic criteria for TCS alone are inadequate for the diagnosis of TCS and must be combined with the clinical presentation to clinch the diagnosis. Thus, a better understanding of the pathophysiology of TCS could improve our ability to diagnose TCS in cases in which the radiographic criteria are not met.[36]

Primary and Secondary Causes

Tethered cord syndrome can be congenital or acquired. In congenital TCS or primary TCS, the tethering of the cord is a developmental phenomenon associated with multiple other congenital anomalies. Secondary tethered cord syndrome occurs in cases in which the normal anatomy of the caudal spinal cord is disrupted by infections, fibrosis, or tumor.[2]

Several congenital anomalies are associated with TCS. Tethered cord syndrome can occur in cases either associated with OSD or nonoccult spinal dysraphism. Occult spinal dysraphism associated with a tethered cord is often present in the form of meningocele, diastematomyelia, caudal agenesis, or dysgenesis syndromes.[2] A

Abbreviations used in this paper: OSD = occult spinal dysraphism; TCS = tethered cord syndrome.

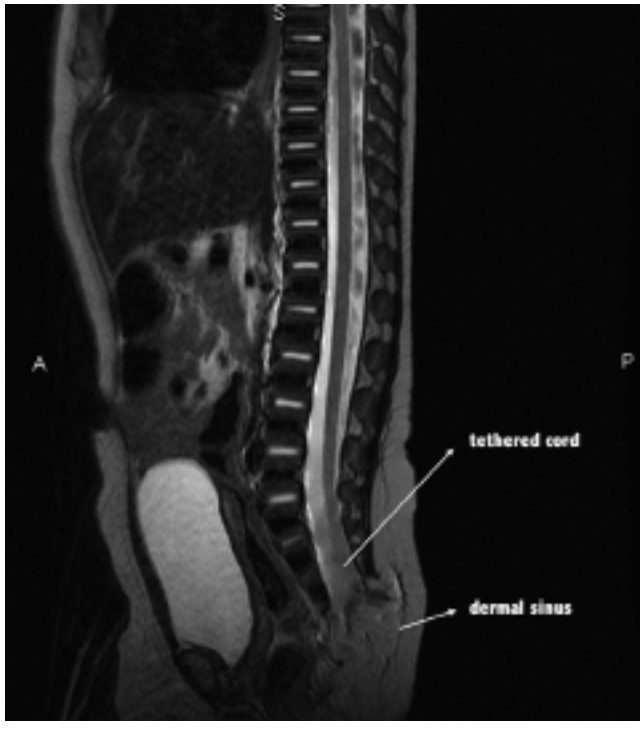

Fig. 1. Sagittal T2-weighted MR image obtained in a 9-week-old girl with a tethered cord lying at the coccyx, associated with a spinal cord lipoma and a dermal sinus.

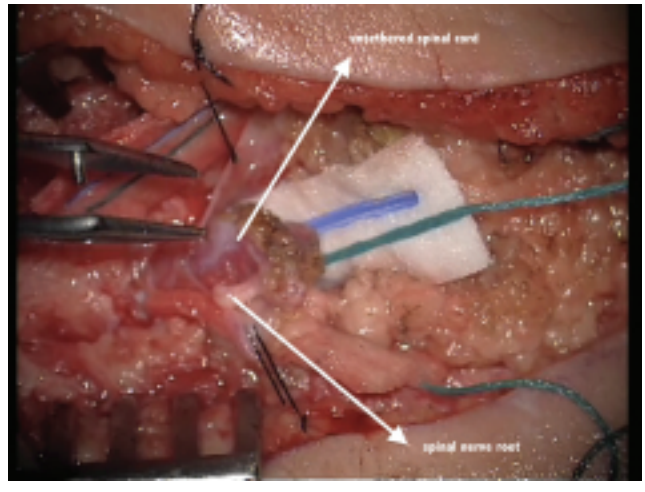

Fig. 2. Intraoperative photograph showing the caudal spinal cord in a 9-week-old girl after untethering. A spinal nerve root can also be observed.

subtype of meningocele called "meningocele manqué" is also a cause of TCS. Meningocele manqué is a form of spontaneously healed meningocele resulting in the formation of dorsal bands. These dorsal bands promote spinal cord tethering. Tethered cord syndrome has also been described in association with a fatty filum terminale, imperforate anus, or a dermal sinus (Figs. 1 and 2). Myelomeningocele is a form of nonoccult spinal dysraphism that is associated with tethered cord and TCS. The tethering of the cord in this case results from the attachment of the spinal cord to the dura mater or the surface ectoderm. Rare, complex, developmental syndromes presenting with anomalies in different systems like the OEIS syndrome (omphalocele, extrophy of the cloaca, an imperforate anus, spinal malformations with tethered cord) and the VATER association (vertebral anomalies, imperforate anus, tracheoesophageal fistula and renal-radial anomalies) can be associated with TCS.[2,15] The Currarino triad (anorectal malformation, presacral mass, and sacral bone abnormalities) is also a syndrome that is associated with tethered cord.[7] Interestingly, genetic studies suggest that TCS may be genetically transmissible.[3,16,26] More recently a link between the *TBX1* gene, 22q11.2 deletion, and trisomy 21 with TCS has been described.[3] Other genetic abnormalities and syndromes like trisomy 13q32, trisomy 8, neurofibromatosis-1, Klippel-Feil syndrome, FG syndrome (shortness of stature, large head, congenital hypotonia, delayed motor and speech development, and a characteristic combination of minor anomalies, malformations, and functional disturbances of the CNS, and gastrointestinal system), Klippel-Trenaunay-Weber syndrome, Dandy-Walker anomaly, and Fuhrmann syndrome have also been reported in connection to TCS.[3,18] The background of the presentation of TCS in conjunction with genetic abnormalities and syndromes indicates a possible genetic link that plays a key role in the development of the syndrome or its pathophysiology.[3,18]

Elasticity of the Spinal Cord

In their original series describing TCS, Hoffman et al.[9] suggested that traction and elasticity of the spinal cord were fundamental factors underlying the pathophysiology of the disease. The most caudal part of the spinal cord exhibits the greatest elongation when traction forces are craniocaudally applied; thus, the conus medullaris is the region of spinal cord that is most vulnerable to traction.[28] The elongation of the filum terminale follows a linear relation with the applied traction force. The spinal cord elongates in a nonlinear fashion as more weight is applied. Interestingly, the spinal cord regions above the attachment of the lowest pair of dentate ligaments do not elongate. This fits the clinical observation that neurological deficits are identified at the level of the caudal spinal cord.[28,36]

Spinal cord tension can also be applied transversely at the craniocaudal axis by the flexion of the torso reminiscent of TCS.[4,8] This clinical picture is common in patients with occult TCS involved in sporting activities like rowing or cycling.[27] In cases in which a spinal mass is present in combination with a tethered cord, local compression of the spinal cord by the mass and flexion of the torso increase the intensity of the symptoms.[8] Similarly in cases of spinal cord malformation in which a ventral septum is present, local pressure on the cord can exacerbate the symptomatology of TCS.[10,11,19]

Filum Terminale

The biomechanical properties of a tethered filum

terminale exhibit a viscoelastic behavior.[5,20–22,25,28,36] The addition of more traction leads to further linear extension of the filum. The connection between the conus medullaris and filum terminale permits an adaptive movement of this section of the spinal cord during spinal flexion and extension.[5,20–22,25,28,36] Changes in the tissue components and anatomy of the filum terminale promoted by fibrous scarring, widening, or the coexistence of a tumor, such as a lipoma, or the presence of anorectal or other lumbosacral abnormalities, such as a myelomeningocele, alter the filum's viscoelastic property and even lead to its loss. Then, the tethered filum terminale becomes an ineffective regulator of the movement of the conus medullaris and acts as an anchor, limiting its movement and contributing to the application of traction forces that lead to neurological impairment.[5,20–22,25,28,36] A key observation in TCS, a low-lying conus, is attributed to the changes that filum terminale has undergone due to developmental anomalies or acquired, secondary causes like infections, scarring, and tumors.[2] Interestingly, Selçuki and Coşkun[24] and Selçuki and colleagues[25] have argued that TCS with a normal-lying conus can be observed in cases in which the elasticity of the filum terminale is lost due to fibrosis.[24,25]

Spinal Cord Tension and Blood Flow

Increase in the traction forces on the spinal cord reduces blood flow to the cord.[6,36] Dolan and colleagues[6] used tracers, autoradiography, and a distraction apparatus in cats to assess the spinal blood flow under various mechanical tension scenarios. Spinal evoked potentials were also used in parallel to monitor the electrophysiology of the neurons. The authors observed severe spinal cord ischemia in the group of the experiments with the longest applied distraction. Traction experiments showed that the blood flow to the gray matter was affected more and spinal evoked potentials demonstrated patterns consistent with ischemia. These results suggest that 2 factors, local distraction and vascular compromise, contribute to the pathophysiology of TCS.[12,14,36,40] A decrease in the diameter of the lumen of spinal vessels, due to traction, substantially reduced the total spinal blood flow, causing local ischemic insult.[6,12,14,36,40] If detethering of the cord is performed within 2–8 weeks of the initial insult, blood flow and spinal evoked potentials may return to normal. Longer delays may result in irreversible changes.[12,14]

The utilization of intraoperative blood flow monitoring in tethered cord surgery further strengthened the notion that ischemia is also a key contributor to the development of the disease. Schneider et al.[23] used laser Doppler flowmetry and identified a significant increase in the blood flow at the caudal spinal cord after untethering operations.

Role of Traction on Hypoxia and Impaired Oxidative Metabolism

The critical observation that traction of the caudal spinal cord is the cornerstone in the TCS development led to extended studies that tried to identify the effect of spinal cord traction at the macroscopic, microscopic, and biochemical level.

Yamada and colleagues[37,40] used reflection spectrophotometry and traction experiments combined with anoxia to study the metabolic status of tethered spinal cord. They demonstrated that a tethered cord in humans and animal models is associated with metabolic abnormalities at the level of cytochrome α,α_3 in interneurons.[37,40] Cytochrome α,α_3 is the terminal oxidase enzyme at the respiratory chain in mitochondria, and its reduction/oxygenation ratio is associated with changes in the availability of oxygen or metabolic demands in mitochondria.[37,40] Increasing traction in the spinal cord is associated with increased reduction of the cytochrome α,α_3. High reduction levels of cytochrome α,α_3 are associated with decreased metabolic demand, decreased oxygen availability, and spinal cord hypoxia and ischemia.[40] Mitochondria in nerve cells with high biochemical reduction states show a decreased production of ATP (adenosine triphosphate). This change in metabolic energy storage and utilization capacity could reflect the impairment of function of nerve cells in the spinal cord.

The aforementioned results indicate that traction-induced hypoxia results in metabolic changes and energy depletion of neurons in the tethered spinal cord.[40] Veins, arteries, and capillaries lose their lumen diameter as they are under mechanical forces, and thus the blood flow to the spinal cord is impaired, leading to decreased energy production and high reduction states of the cytochrome α,α_3 in neuronal mitochondria. The degree of the metabolic impairment is correlated with the severity of the neurological symptoms.[33,35,36,38,39]

Interestingly, the surgical untethering of the spinal cord is followed by the return of the metabolic status in the cytochrome α,α_3 of the mitochondria from a high reduced state to a re-oxidation state reflecting the return to normal.[40] This phenomenon was observed in cases with mild or moderate metabolic redox changes while cases with severe reduction/oxygenation disruption did not yield adequate results. The return to the normal metabolic state of the spinal cord was accompanied by neurological improvement in a period ranging from 2 weeks to 2–4 months. In severe cases, only partial neurological improvement was observed, with limited metabolic shift from a predominantly high reduction state of the cytochrome to states with more oxidation. The aforementioned results were also confirmed with studies that used 2-deoxyglucose as a measure of metabolic activity.[33–40]

A More Functional Definition of TCS

The classic definition of the TCS involves the presence of a thickened filum terminale and/or a low-lying conus medullaris in a patient with neurological deficits. Currently, a more accepted diagnosis of TCS is defined as a pathological fixation of the spinal cord in an abnormally lying position. The data, derived from the pathophysiology of the syndrome, indicate that mechanical tension of the caudal spinal cord, vascular compromise, and hypoxia result in metabolic derangements and neurological impairment. Although the radiological evidence of a low-

lying conus are the key factors in the diagnosis of TCS, a clinical picture consistent with TCS can also be present in a group of patients—accounting for 14–18% of various published series—with a normal anatomical position of the conus.[13,17,24,25,29,32,33,36] In these cases the presence of a tight, inelastic filum terminale is the key factor that leads to the development of the syndrome.[25] Selçuki and Coşkun[24] found that urodynamic studies are more reliable than somatosensory evoked potentials for predicting that a tight filum terminale is probably the cause of the clinical presentation. This group of patients exhibit a significant response to the surgical untethering that varies from 47–67% in the published series.[13,17,24,25,29,32,33,36]

Results obtained in the group of patients with TCS symptoms and a normal-lying conus indicate that the approach to the diagnosis and treatment of TCS should not rely only on an anatomical definition and criteria alone but should also take into account the underlying pathophysiology. An inelastic filum terminale that has lost the ability to act as a buffer to traction forces applied at the caudal spinal cord is the cause in most of these cases and it is a part of the pathophysiology of the syndrome that must be clarified to further understand when surgery should be performed in cases involving a normally positioned conus. The existence of neurological, urological, and orthopedic signs and symptoms supports an aggressive approach because about 50% of these patients improve significantly following detethering surgery.

Conclusions

The pathophysiology of TCS involves the existence of traction and the loss of the elasticity of the filum terminale as well as ischemic insult to the cord. The extension of the spinal cord, the metabolic abnormalities observed, and the decrease in spinal cord blood flow in TCS lead to neurological deficits. Further research should be conducted at the biomechanical, biochemical, and molecular levels to be able approach the specific activated pathways. The identification of the threshold level of spinal cord damage that defines the clinical picture of TCS as reversible or irreversible should also be clarified. The indications that specific genes participate in the evolution of the syndrome should be further studied to unveil the mode of inheritance in familial cases and identify key genes in the pathophysiology of the syndrome.

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: Filippidis, Kalani. Analysis and interpretation of data: Filippidis, Kalani. Drafting the article: Filippidis, Kalani. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Study supervision: Rekate, Theodore.

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Idiopathic ventral spinal cord herniation: a rare presentation of tethered cord

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Idiopathic ventral spinal cord herniation is a rare condition that has been increasingly reported in the last decade. The natural history and optimal management have yet to be defined. Therefore, debate exists regarding the pathogenesis and surgical management of this condition. The purpose of this review article is to further educate neurosurgeons about the surgical techniques and outcomes associated with treating this rare and often misdiagnosed condition. *(10.3171/2010.3.FOCUS1089)*

Key Words • **idiopathic spinal cord herniation** • **tethered spinal cord** • **thoracic spine**

Idiopathic ventral spinal cord herniation is a rare condition that primarily affects the thoracic spinal cord. Originally described by Wortzman et al.[48] in 1974 after these authors discovered the spinal cord protruding through a dural defect when performing a thoracotomy for disc herniation, 128 cases have since been reported. With increased awareness among physicians and the widespread availability of MR imaging, more cases have been identified in the last decade. Despite this increase, many questions still exist regarding the pathogenesis, natural history, and surgical outcomes. Without a clear understanding of the natural history of this condition, the optimal management strategy for these patients remains controversial.

Although the origin of the dural defect is unknown, many theories have been postulated over the years, largely based on observations made during surgery. Variations in the type of dural defects include a full-thickness dural defect, a defect in the inner layer of duplicated dura mater, and an epidural cyst or pseudomeningocele. It has been proposed that the ventral dural defect causes a tethering of the spinal cord that progressively worsens as a result of persistent CSF pulsations and negative epidural pressures.[27] This tethering results in progressive neurological symptoms, which are often misdiagnosed.

In this article, we review the clinical presentation, pathogenesis, radiographic imaging characteristics, and surgical results in patients with this rare condition.

Clinical Presentation

Patients usually present with progressive myelopathy and signs of either Brown-Séquard syndrome or spastic paraparesis months to years prior to surgical intervention. The interval between initial onset of symptoms and surgical intervention ranges from 6 months to 36 years, with a mean of 5.2 years.[14] Approximately 66% of reported cases present with Brown-Séquard syndrome, 30% with symmetrical spastic paraparesis, 4% with pure sensory deficits, and 1% with isolated motor deficits.[34] Bowel and bladder dysfunction are reported in 7%.[34] The age of patients affected is between 21 and 78 years, with a mean age of 51 years.[34,36] Twice as many women are affected as men.[10]

Pathogenesis

Most idiopathic spinal cord herniations occur ventrally in the thoracic spine. Although many theories exist, the pathogenesis of this condition is unknown. It has been hypothesized that the thoracic spine is primarily involved, due to the normal kyphosis of the thoracic spine, the ventral position of the spinal cord within the midthoracic region, the spinal cord's physiological ventral motion due to cardiac pulsations, and the biomechanical impact of flexion and extension movements on the thoracic spinal cord.[34] Considering these factors, idiopathic herniations often develop through ventral or ventrolateral dural defects in the upper or midthoracic region. A review of the literature reveals that T4–5 is the most common level.[14] A ventral dural defect, either congenital or acquired, is the proposed basis for this entity.

Congenital Defect

Some authors have proposed the possibility of a congenital dural defect that allows for gradual herniation of

the spinal cord over time. Theories about this congenital defect include a preexisting ventral pseudomeningocele, a meningeal diverticulum, or an extradural arachnoid cyst.[47] Both Wortzman et al.[48] and Masuzawa et al.[24] suggested that herniation results from a congenital ventral meningocele. No other congenital bony spinal or neural deformities were reported in these cases. Some authors have questioned how a congenital defect can lead to symptoms in late adulthood.[23]

Another theory advocated by some authors is congenital duplication of the ventral dura mater, with a defect in the inner layer.[25,30,31,33] Nakazawa et al.[30] suggested that a cavity caused by CSF pulsations forms between the layers of the dura, leading to herniation of the spinal cord and strangulation at the neck of the hernia. Opponents of this theory question the mechanism for disruption of this inner dural layer and the reason for fenestration.[17]

This theory is based on operative findings and was previously challenged for lack of radiological and pathological evidence. However, in 2008 Ishida et al.[19] reported a case in which the duplicated dura ventral to the spinal cord was identified prior to surgery by using high-resolution 3D MR imaging. In a recent review of the literature, 16 of 89 cases reportedly had duplication of the dura.[10] Duplication was always found to be ventral to the spinal cord, with the defect consistently in the inner layer. Pathological examination of the inner layer of dura mater was performed in 6 cases, showing similar histological findings to that of dorsal dura, without any evidence of inflammation.[25,30,31,39,43,44]

Dorsal arachnoid cysts are also associated with ventral spinal cord herniation. Isu et al.[20] hypothesized that the pressure of the dorsal arachnoid cyst causes thinning of the ventral dura until a tear occurs. The spinal cord then comes in contact with the dural defect, and CSF pulsations over time result in herniation. Resection of the cyst alone, however, does not result in spinal cord untethering or improvement of symptoms. Patients in whom cyst resection was performed initially have required additional surgery to untether the cord and to prevent further neurological decline.[10,38]

Acquired Defect

The proposed pathogenesis of acquired defects includes damage to the ventral dura mater by inflammation,[28] remote spinal trauma,[40,46] vertebral body defect,[29] and thoracic disc herniation.[26] Hausmann and Moseley[17] theorized that in some cases, thoracic disc herniation may damage the ventral dura, allowing the cord to herniate through the defect. Several other authors have also described thoracic disc herniation producing an adjacent dural defect, with subsequent cord herniation at the level of the vertebral body and not at the disc level.[1,4,10,17,26] These patients are thought to have a worse prognosis.[4] Erosion of the dura by a herniated and calcified disc has been observed just caudal to the level of spinal cord tethering.[23,32] Although the theory of a thoracic disc causing ventral dural weakening with subsequent cord herniation is described often, the case reported by Miyaguchi et al.[26] is the only one in the literature in which a herniated disc was visualized through the ventral dural defect at surgery. It is proposed that the thoracic disc injures the ventral dural surface with repeated flexion and extension, and predisposes it to further damage.

Pathophysiological Features of Tethering

Borges et al.[7] theorized that tethering of the spinal cord on the side of the herniation results in unilateral damage of the lateral funiculus, and that symptoms of spinal cord herniation are a result of tension on this structure. The lateral position of the lateral spinothalamic tract within the lateral funiculus predisposes its axons to dysfunction secondary to tension induced by the tethering. As the tension progresses, the corticospinal tracts become involved, resulting in progressive weakness and spasticity.

Another theory regarding the progressive nature of symptoms is ischemia due to distortion or involvement of anterior spinal vessels due to tethering.[47] This vascular mechanism may also explain the lack of improvement in some patients following untethering of the spinal cord.

Diagnostic Imaging

Magnetic resonance imaging often demonstrates ventral displacement of the thoracic spinal cord and enlargement of the dorsal subarachnoid space. A ventral C-shaped kink is typically seen on sagittal imaging (Fig. 1). However, this condition is often misdiagnosed and misinterpreted as a dorsal arachnoid cyst.[10,20,30,38,45] Atrophy of the spinal cord with signal change can also be seen, potentially confusing this condition for astrocytoma, disc herniation, extradural compression, and transverse myelitis.[13,17,46]

Misdiagnosis may lead to erroneous surgery for decompression and resection, with no clinical improvement.[20,38] Several authors have reported clinical worsening following biopsy of the herniated spinal cord, which may appear abnormal compared with normal spinal cord tissue.[7,17,24,38]

Computed tomography myelography has been used for better evaluation of the relationship of CSF to the spinal cord, particularly in differentiating the widened dorsal subarachnoid space with a dorsal arachnoid cyst. In cases of idiopathic ventral herniation, CT myelography demonstrates no filling defect dorsal to the spinal cord or retention of contrast agent ventral to the dura mater. In contrast, arachnoid cysts demonstrate an intradural filling defect[41] (Fig. 2).

The MR imaging signal intensity of arachnoid cysts is similar to that of CSF on both T1- and T2-weighted images, and the cyst walls are usually not visible. Phase-contrast MR imaging may help establish the presence or absence of a dorsal arachnoid cyst by showing reduced CSF pulsations in the cyst. This modality is also valuable in evaluating absence of pulsatile flow ventral to the spinal cord at the level of the herniation. This absence is another valuable sign of tethered cord.[8]

Untethering of the spinal cord is clearly shown on postoperative imaging, with restoration of spinal cord

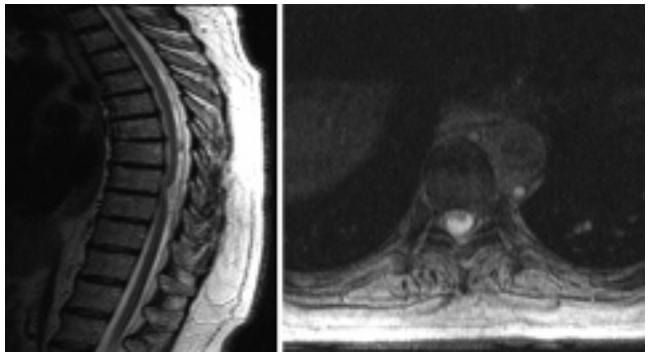

Fig. 1. Preoperative MR imaging studies obtained in the thoracic spine of a 71-year-old woman who presented with Brown-Séquard syndrome and progressive myelopathy. **Left:** A sagittal T2-weighted image demonstrates a ventral spinal cord herniation at the level of T8–9. **Right:** An axial T2-weighted image shows ventral displacement of the thoracic spinal cord and a widened dorsal subarachnoid space.

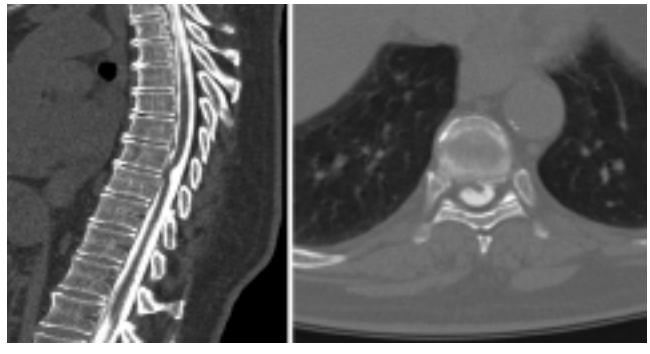

Fig. 2. Preoperative CT myelography studies of the same patient showing no contrast ventral to the spinal cord at T8–9. **Left:** Sagittal image demonstrating contrast enhancement dorsal to the herniated spinal cord. This makes a diagnosis of arachnoid cyst less likely. **Right:** An axial image demonstrating the ventral herniation at the level of the disc space.

alignment and evidence of CSF ventral to the cord (Fig. 3).

Surgery and Techniques

The optimal treatment strategy for idiopathic ventral spinal cord herniation has not yet been determined because the natural history of this rare condition is still unclear. In general, treatment consists of either conservative management or surgery. Conservative management has been proposed for patients without significant motor deficits or progressive myelopathy.[1,2,17,23] Most reported series are surgical case discussions, and only a few authors have presented results of conservative management in these patients.[1,2,17,23,37]

In one of the largest series, Massicotte et al.[23] reported on 8 patients, 4 of whom did not have surgery. These 4 patients were observed for up to 8 years after presentation and none developed progressive neurological symptoms. In each of these nonsurgical patients, neither weakness nor spasticity were evident at presentation. Similarly, Ammar et al.[2] reported on 2 patients who presented without weakness or spasticity and were followed for 15 months in one and 5 years in the other, without progression of symptoms. Senturk et al.[37] followed a 38-year-old woman with progressive thoracic back pain without neurological deficits for 6 months without progression of symptoms. Hausmann and Moseley[17] reported on 2 patients who presented with spasticity and weakness, but did not undergo surgery. No further follow-up information was provided. Given the lack of long-term follow-up, the classification of patients who would benefit from nonsurgical management is not yet known.

Surgery is typically recommended for patients with motor deficits or progressive neurological symptoms. Both dorsal and ventral surgical approaches have been used to treat ventral spinal cord herniations. Only Borges et al.[7] and Wortzman et al.[48] have used a ventral approach to treat these lesions. All other reports have used a strictly dorsal thoracic approach with laminectomy. Although the dorsal approach allows for a wide exposure of the spinal cord, it often does not provide adequate ventral exposure to untether the spinal cord and repair the dural defect safely. To avoid retracting the spinal cord, the dentate ligaments and arachnoid adhesions are usually released to mobilize it.[9,15]

To obtain greater exposure of the ventral dura, costotransversectomy and transpedicular approaches have also been described.[9,15,17,42] Gwinn and Henderson[15] reported on 3 patients who underwent a unilateral posterolateral transpedicular approach for repair of the dural defect

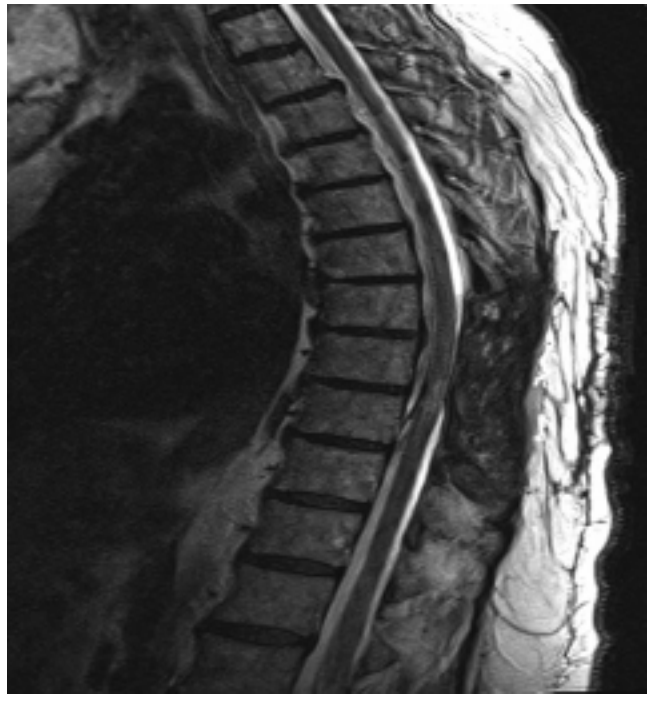

Fig. 3. Postoperative MR imaging study of the thoracic spine demonstrating realignment of the spinal cord within the canal and restoration of CSF ventral to the spinal cord. The patient showed neurological improvement at the 2-year follow-up.

with bovine pericardia. All patients were clinically improved at 3 months, without recurrence of tethering. In a modification of the transpedicular approach, Chaichana et al.[9] reported a bilateral transpedicular approach followed by unilateral removal of a pedicle and transverse process to provide a sufficiently wide bilateral exposure of the ventral dural defect. A Gore-Tex graft was sutured to the ventral dura for repair of the dural defect. At 6-week follow-up, the patient was improved clinically, with no radiographic evidence of ventral herniation.

Although techniques may differ among authors, release of the tethered spinal cord and repositioning of the cord to a normal anatomical position is generally agreed on as the main goal of surgery. To prevent reherniation, several different strategies have been advocated. These include enlargement of the ventral dural defect,[21,25,30,33,39,44,45] primary closure of the defect with sutures,[7,18,22] and insertion of a ventral patch for duraplasty.[4,6,11,13,23]

In a recent literature review, Saito et al.[33] found that approximately 20% of patients with primary ventral dural closure worsened clinically after surgery, compared with 10% of patients with dural patching or widening of the dural defect. This difference is attributed to the difficulty of suturing the ventral dura from a strictly dorsal approach without excessively retracting the spinal cord. Many authors now prefer using a dural patch to seal the defect, because it minimizes spinal cord manipulation and requires less direct visualization.

Duraplasty was first described by Masuzawa et al.[24] for a case in which a fascial flap was sutured to the ventral dural opening. Subsequently, a number of different grafting materials have been used to obliterate the defect, including muscle, fascia, fat, bovine pericardium, Teflon, and Gore-Tex.[2,3,5,12,15,23,27,28,35,38,48]

Because of the sporadic nature of this condition and limited long-term data, there is no evidence demonstrating superiority of one particular graft material or operative strategy in preventing the recurrence of tethering.

Clinical Outcomes

Because the number of surgically treated patients is limited, there is little evidence to predict which ones would benefit from surgery and when it would be most effective. Relying on case reports, which have significant variability in patient symptomatology and time to presentation, extensive reviews of the literature have only been able to make observations regarding the experience of surgeons to date.[10,14,34] In general, surgery is usually followed by stabilization or improvement in neurological symptoms, but postoperative worsening has been reported. In one of the largest published series,[16] 10 patients were followed for 6–110 months after surgery. Four patients showed motor improvement, 2 had pain relief, and 1 experienced improvement in sensory function. Three patients were unchanged overall.

Although most patients improve neurologically after surgery, its long-term benefits are not clear. The range of follow-up is variable in reported cases, from weeks to years. In the longest reported follow-up, Selviaridis et al.[36] reported a recurrence of ventral spinal cord herniation and clinical myelopathy after 10 years. The patient initially improved neurologically after the first surgery, then developed progressive paraparesis and urinary incontinence years later. During the first surgery, Surgicel was packed into the dural defect after untethering. On repeat surgery, a larger segment of the spinal cord was found to be herniated through a larger dural defect. Other cases of recurrence have been reported, ranging from 18 months to 3 years after surgery.[2,29]

Overall, patients tend to improve after surgery. Numerous case reports have demonstrated improvement in clinical outcomes for surgically treated patients. In a recent review of the literature, 73% of all patients improved, 20% experienced no change, and 7% deteriorated after surgery.[34]

When examined for differences based on presenting neurological signs, the outcomes for patients who initially presented with Brown-Séquard syndrome were better than for patients with spastic paraparesis. Improvement was reported in 56 (76.7%) of 73 patients with Brown-Séquard, and in 9 (47.3%) of 19 with spasticity.[34]

In the most comprehensive review of the literature, Groen et al.[14] performed a meta-analysis to identify factors that affect postoperative outcome. In their analysis, Brown-Séquard syndrome and release of the herniated spinal cord were independent factors associated with favorable postoperative outcomes. With regard to management of the ventral dural defect, widening of the defect was associated with the highest prevalence of postoperative motor function improvement when compared with the application of a ventral dural patch. Persistent ventral displacement or realignment of the spinal cord on postoperative MR imaging was not found to correlate with outcome. However, only 56% of reported patients underwent postoperative imaging. Despite the meta-analysis, it is evident that the management of this condition needs to be individualized for each patient, because definitive conclusions cannot be made from a heterogeneous sampling of case reports.

Conclusions

Idiopathic ventral spinal cord herniation and tethering is a treatable cause of myelopathy that is often misdiagnosed. An MR imaging study that shows obliteration of the ventral CSF space with ventral focal deformity of the cord in the thoracic region raises a high index of suspicion for this condition. Surgical correction involves untethering the spinal cord and repairing the dural defect. Surgery may lead to clinical improvement in select cases, despite a long history of myelopathy.

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: both authors. Acquisition of data: both authors. Analysis and interpretation of data: both

authors. Drafting the article: both authors. Critically revising the article: both authors. Reviewed final version of the manuscript and approved it for submission: both authors. Administrative/technical/material support: Krishnaney. Study supervision: Krishnaney.

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Reversal of longstanding neurological deficits after a late release of tethered spinal cord

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The importance of early surgery for tethered cord syndrome in the pediatric population is well established. Optimal treatment and prognosis of tethered cord in adults, on the other hand, is less clear. Some advocate a conservative approach in asymptomatic patients, while others recommend early detethering in all patients. For symptomatic patients, however, there is a consensus in favor of early surgery to prevent progression of neurological deficit. Many studies have reported cessation of neurological decline or reversal of recently acquired neurological deficits in patients with adult tethered cord syndrome. There are limited data in the literature about late surgery for the treatment of tethered spinal cords when the neurological deficits are longstanding. We report on a 37-year-old woman who demonstrated dramatic neurological improvement after surgical release of a tethered spinal cord more than 20 years after the onset of progressive neurological deficits. *(DOI: 10.3171/2010.3.FOCUS1078)*

Key Words • spinal cord • adult tethered cord • late detethering • neurological improvement

Early surgery for TCS in the pediatric population is standard;[5,19] optimal treatment and prognosis of TCS in adults is, however, unclear. Some advocate a conservative approach in asymptomatic patients, while others recommend early detethering even in all patients.[5,17,19,24] For symptomatic patients, however, there is a consensus advocating early surgery to prevent progression of neurological deficit.[5,13,14,16,17,21,24] Many studies have reported cessation of progressive neurological decline or even reversal of recently acquired neurological deficits in adult patients with TCS.[1,4,11,15] There are limited data in the literature on the outcome of late surgery when neurological deficits are long standing and presumed to be irreversible. We report on a 37-year-old woman who experienced dramatic neurological improvement after surgical release of a tethered spinal cord more than 20 years after the onset of progressive neurological deficits.

Case Report

History and Presentation. This 37-year-old Caucasian woman, in whom a diagnosis of Charcot-Marie-Tooth (CMT) hereditary neuropathy was established, was referred for evaluation of longstanding and progressive neurological deficits including incontinence of urine.

Her early motor development was notable only for a delay in walking until 18 months of age, but subsequent motor development and function appeared normal until high school. At 16 years of age, the patient was referred for orthopedic and podiatric care. She underwent corrective surgery for a toe deformity, but no diagnosis was offered. By 20 years of age, she had bilateral foot drop. She received a diagnosis of CMT hereditary neuropathy and was treated with bilateral ankle foot orthoses. At age 25, she underwent multiple surgical procedures including tendon transfers, Achilles tendon lengthening, and wedge osteotomies.

Urinary incontinence, including nocturnal enuresis, began at age 20 years and progressed in severity despite treatment with anticholinergic medications and a sling procedure. Other problems included progressive numbness over the plantar aspect of both feet, decreased sensation over the left buttock, perianal numbness, and a feeling of being "off balance." At age 32 years, MR imaging of the brain and entire spine was performed to rule out multiple sclerosis. The only abnormality noted in the report was a tethered cord. The patient was not informed of this finding, and no further evaluation or treatment was advised.

At 36 years of age, nerve conduction studies in the

Abbreviations used in this paper: CMT = Charcot-Marie-Tooth; EMG = electromyography; TCS = tethered cord syndrome.

lower limbs showed evidence of axonal neuropathy, which was suggestive of CMT type 2. In light of her clinical presentation, particularly the urinary incontinence and lack of upper extremity involvement, the diagnosis of CMT type 2 was questioned and she was referred to our institution for management of her progressive neurological deficit and further evaluation.

Examination. On physical examination, the patient was 5 feet 6 inches tall and weighed 170 pounds. Examination of the skin demonstrated a 0.7-cm freckle left of the midline in the gluteal cleft. No dermal sinuses or hair tufts were visible. There was a hyperlordotic lumbar curve, and no pain was elicited with back extension maneuvers. Motor examination using the Medical Research Council scale revealed weakness only in the lower limbs: the iliopsoas and quadriceps were 5/5 bilaterally, ankle dorsiflexors 0/5 on the right and 3/5 on the left. The plantar flexors and extensor hallucis longus muscles were 0/5 bilaterally. The patient's tendon reflexes were hypoactive at the knee and absent at the ankle. Her calf diameters were both 34 cm. In the left S-1 distribution sensory examination was diminished to pinprick and light touch. No deficits were detected in either upper or in the right lower limb. Joint position sense and vibration sensation were decreased on the left as well. The Romberg test was positive. The patient had a "steppage" gait and inability to walk on her toes or heels.

Urodynamic studies with bladder EMG revealed neurogenic bladder dysfunction and a small bladder capacity. There was detrusor overactivity and possible intermittent detrusor sphincter dyssynergia with normal upper tracts. There was significant cough-induced bladder overactivity with evidence of stress incontinence and nonrelaxation with voiding.

A CT scan demonstrated a Grade I isthmic spondylolisthesis at the L5–S1 level. Magnetic resonance imaging showed the spinal cord to be tethered at S1–2; it terminated in a lipoma at the S-2 level (Fig. 1). Comparison with the previously obtained spine MR images showed no appreciable change. Electromyography of the lower extremities showed evidence of chronic bilateral lumbosacral radiculopathy at L5–S1. Nerve conduction velocity was abnormal only in the lower limbs, with markedly diminished amplitude of compound muscle action potentials and no response to stimulation of the left tibial nerve. Left sural amplitude was diminished as well.

The clinical, electrophysiological, and radiological features were consistent with and attributable to a diagnosis of symptomatic tethered spinal cord.

Operation and Postoperative Course. Surgical treatment involved S1–2 laminectomies, intradural exploration with untethering of the spinal cord, sectioning of the fatty filum terminale with intraoperative EMG monitoring, and subtotal resection of the intradural lipoma (Fig. 2). The extent of the lipoma resection was dictated by intraoperative EMG findings. Pathological examination showed adipose tissue consistent with lipoma encasing a small portion of the filum terminale. The patient had an uneventful postoperative recovery. Postoperative MR imaging showed untethering of the spinal cord, resection of the fatty filum, and subtotal resection of the lipoma (Fig. 3).

Three months postoperatively, the patient reported an increase in sensation over the posterior thighs and perineum. Nocturnal incontinence had resolved completely. Postoperative motor examination was noteworthy for 5/5 power in bilateral proximal lower extremities and in the left distal lower extremity (dorsiflexion and plantar flexion). Right lower extremity power and extensor hallucis longus strength remained 0/5. Sensory examination improved throughout the left lower extremity and perineum but was not normal as compared with the opposite side.

Additionally, the patient reported marked improvement in sacral sensation, menstrual awareness, and sexual feeling.

Discussion

Tethered cord syndrome is a congenital syndrome resulting in mechanical rostrocaudal traction on the spinal cord and variable neurological deficits. It is a part of the constellation of disorders related to a defective retrogressive differentiation of the caudal neural tube.

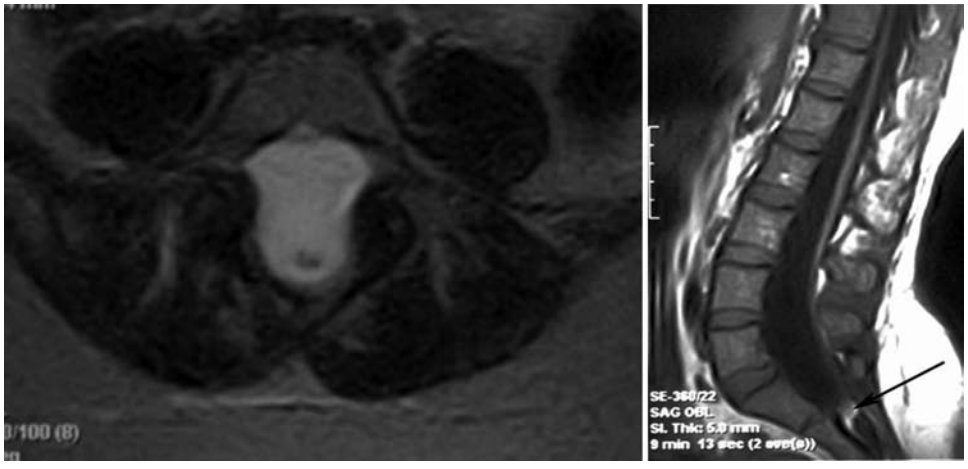

Fig. 1. Left: Preoperative axial T2-weighted MR image at the L-5 pedicle level demonstrating a bony defect dorsally with enlargement of the thecal sac and dorsal location of the spinal cord. **Right:** Preoperative sagittal T1-weighted MR image showing a low-lying spinal cord terminating in a lipoma at the S1–2 level *(arrow)*.

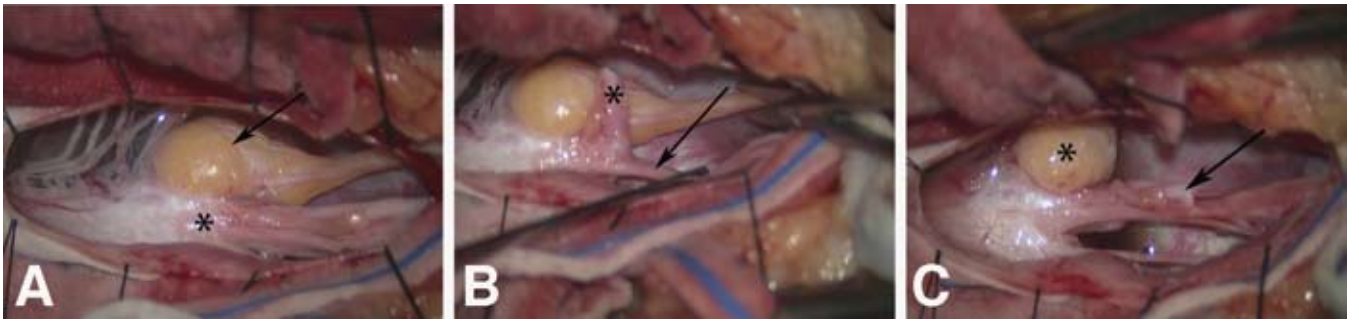

Fig. 2. **A:** Intraoperative photograph showing the lipoma tethering the cord *(single arrow)* and fatty infiltration of the cord/thickened filum terminale *(asterisk)*. **B:** Intraoperative photograph showing sectioning of the lipoma causing tethering of the cord (note scissors on right) and the stump of the previously sectioned fatty filum terminale *(asterisk)*. Normal sacral nerve roots were preserved (beneath retractor, *arrow*). **C:** Intraoperative photograph showing the residual lipoma *(asterisk)* and stump of the previously sectioned filum terminale after removal of all resectable fatty infiltration *(arrow)*. Note that the cord terminates at the level of the lipoma. Preserved sacral nerve roots are visible emerging from distal end of the cord. The extent of lipoma resection was dictated by intraoperative EMG findings.

Approximately 75% of pediatric patients with TCS have progressive neurological deficits. These may include radicular pain, weakness, asymmetrical hyporeflexia, spasticity, sensory changes, and sphincter dysfunction.[16] The upper motor neuron symptoms are believed to result from mechanical rostrocaudal traction and ischemic damage to the cord.[2,3] The goal of a detethering surgical procedure is to release the conus from the abnormal filum terminale, which anchors it distally.[16] If the procedure is performed early, excellent neurological recovery is expected in sphincter function, pain, motor deficits, and sensation. Preferably, surgery is performed before the onset of fixed or irreversible neurological deficits.[18] Objective improvements in blood flow and nerve conduction velocities have been reported after early surgical untethering of the spinal cord.[10,22] The surgery itself is an effective procedure associated with low morbidity and prevents long-term morbidity resulting from the natural progression of the disease process.[7,8]

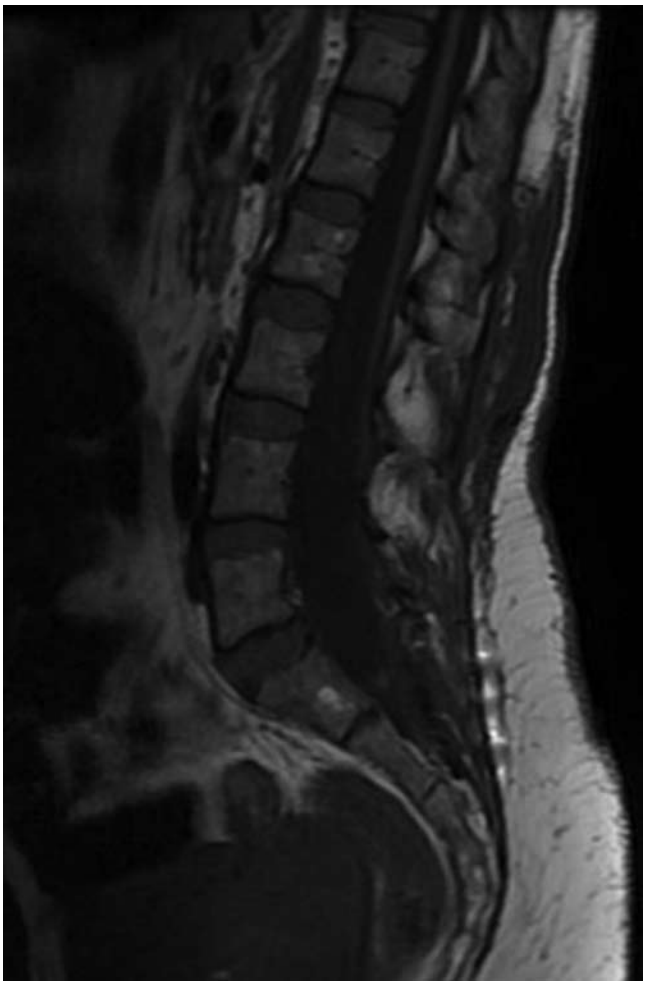

Fig. 3. Postoperative sagittal T1-weighted MR image demonstrating a subtotal resection of the lipoma and release of the tethered cord.

In adult TCS, the late onset of symptoms is attributed to repeated microtrauma during flexion and extension of a mechanically tethered spinal cord.[5,11,25] A traumatic event that acutely stretches the conus may cause acute deterioration or may even precipitate initial presentation in previously undiagnosed adults.[11] Studies have shown that detethering in an adult population may relieve pain and improve sphincter function.[19]

Several features in our patient's history and findings on physical examination were not compatible with her diagnosis of an axonal form of CMT,[20] including sparing of the hands, bulky calf muscles, saddle hypesthesia, neurogenic bladder, and asymmetrical deficit in the lower extremities.

Several studies of adult TCS have shown that the potential for reversal of upper motor neuron dysfunction can be poor once neurological symptoms are detected.[3,4,9] Lee et al.[15] reviewed several large studies looking at the natural history and long-term outcome. They found that detethering was clearly beneficial when symptoms were of recent onset. Similarly, Aufschnaiter et al.[1] published a case report of an excellent outcome in a patient with symptoms and objective MR imaging diagnosis of tethered cord 1 year prior to surgery. After a literature review of more than 450 patients, these authors concluded that excellent results are generally achieved in adult patients with symptoms 1 year or less in duration.

The neurological deficits that reversed in our patient

were present for more than 20 years, and there was MR imaging evidence of a tethered cord 5 years prior to surgery. This report challenges the usual presumption that late definitive surgery may not be as beneficial in such cases. The fact that the symptoms remained progressive may be the clue to reversibility. Given this experience and that described in other anecdotal reports,[1,6,12,23] we propose that in chronic cases, detethering more than 5 years after symptom onset may result in significant improvement, particularly when progression has continued.

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: Heary, Michaels. Drafting the article: all authors. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: Heary, Michaels.

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