Tethered cord syndrome and occult spinal dysraphism

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Tethered cord syndrome is a progressive form of neurological deterioration that results from spinal cord tethering by various dysraphic spinal abnormalities. The syndrome, treatments, outcomes, and current controversies are reviewed.

KEY WORDS • tethered cord syndrome • occult spinal dysraphism

Progressive neurological deterioration localized to the lower spinal cord resulting from traction on the conus medullaris has been termed TCS and has been described in children[23,24] and adults.[57,61] This syndrome of neurological deterioration was described and treated in 1891 by Jones.[29] It was again described in 1918,[5] and the recommendation for surgical intervention was made. It was rediscovered in the mid-20th century.[19,27,44,45] In its classic form, TCS involves traction on a low-lying conus medullaris;[17,57] however, TCS has also been described in the setting of a conus medullaris in the "normal" position.[69,70] Features associated with the syndrome include the dysraphic spinal elements, cutaneous stigmata of OSD, vertebral anomalies, orthopedic abnormalities (scoliosis and extremity abnormalities), neurological deterioration at the level of the lower spinal cord including bowel and bladder dysfunction, and anorectal malformations.[12,71]

Mechanical traction on the spinal cord may be a cause of progressive symptoms.[48] Yamada, et al.,[73] have proposed that symptoms appear after hypoxic damage within the conus medullaris of patients with TCS. Blood flow improvement has been noted after the spinal cord has been surgically untethered.[63] Retethering of the spinal cord may occur in patients with OSD who have previously undergone an untethering procedure and may also occur in patients with previously repaired myelomeningoceles.[48]

Spinal dysraphism is a term that refers to all forms of developmental abnormalities occurring in the midline of the back—from the skin externally to the vertebral bodies internally.[26] Although the true incidence of spinal dysraphism is unknown, authors of some studies have estimated an incidence of 0.05 to 0.25 per 1000 births.[33,67] These abnormalities usually involve the lumbosacral spine, although lesions in the cervical and thoracic region do occur. Spinal dysraphism, which may result in a tethered spinal cord, exists in an open form, spina bifida aperta, and in a closed form, spina bifida occulta. The most common expression of spina bifida aperta is the myelomeningocele. Common forms of spina bifida occulta include the lipomyelomeningocele and diastematomyelia.

The same term, spina bifida occulta, when it refers to a benign bone cleft in the L-5 or S-1 spinous process that occurs in approximately 17% of the total population and 30% of normal individuals aged 1 to 10 years,[4,40] does not imply an association with a tethered spinal cord or TCS.

EMBRYOLOGICAL FEATURES OF SPINAL DYSRAPHISM

Human intrauterine development is divided into the embryonic and fetal periods.[16,43] The embryonic period includes the first 50 to 62 days postconception,[16] and has itself been divided into 23 stages.[68] Each stage encompasses approximately 2 to 3 days. The fetal period includes the subsequent 7 months of gestation.[16] Stages 8 through 14 (Days 18–32) are of particular importance with respect to the development of the central nervous system. During these stages the three major steps of the development of the central nervous system occur. These steps include neurulation, canalization of the tail bud, and regression. Neurulation, the process by which the neural tube is formed, occurs during stages 8 through 20 (Days 18–48). During this process, the flat neural plate folds on itself into a neural tube which is covered by a continuous layer of cutaneous ectoderm.[16] The closure of the neural tube begins in

Abbreviations used in this paper: CT = computerized tomography; MR = magnetic resonance; OSD = occult spinal dysraphism; SCM = split cord malformation; TCS = tethered cord syndrome.

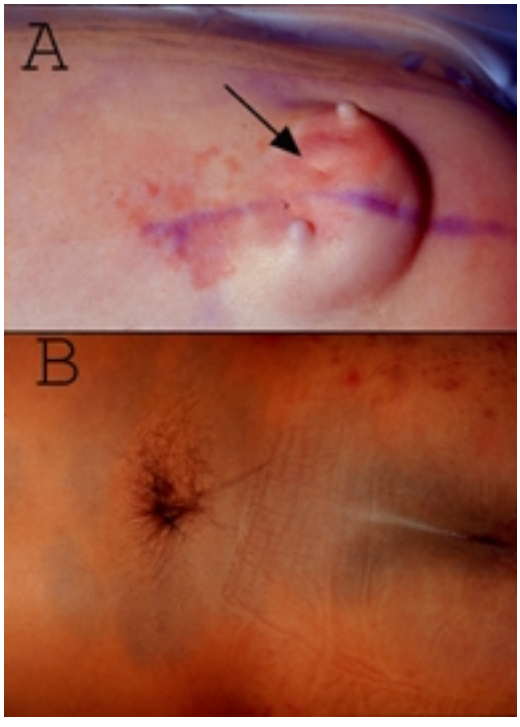

Fig. 1. Photographs showing cutaneous stigmata of OSD. A: One of three dermal sinuses *(arrow)*. Also present are a subcutaneous lipoma and cutaneous hemangioma. B: A hairy patch of the lumbosacral spine is displayed.

the upper cervical region and then extends caudally and cephalically. During stage 12, the caudal portion of the neural tube closes at the level L-1 or L-2.[16] Neurulation gives rise to the spinal cord only down to the lumbar spine region. The spinal cord caudal to this is formed by the process of canalization. Errors during neurulation may lead to various congenital malformations such as myelomeningocele, meningocele, lipomyelomeningocele, SCMs, the dermal sinus, and intraspinal tumors such as dermoids and epidermoids. With the completion of neurulation, the neural tube is totally covered by cutaneous ectoderm.[16]

The tail bud forms after completion of neurulation. The formation of the neural tube caudal to that formed during neurulation occurs by canalization of the tail bud, which occurs during stages 13 through 20 (Days 28–48).[16] This process consists of the development of vacuoles within the tail bud, then coalescence of these vacuoles to form the canal, which then connects with the rostral neural tube formed during neurulation. Abnormalities that develop during canalization of the tail bud can give rise to the thick terminal filum, terminal myelocystocele, and lipomyelomeningocele.

The terminal filum and cauda equina are formed from the caudal portion of the neural tube by regression. The ventriculus terminalis marks the level of the future conus medullaris and is a dilation of the central canal that can be identified at stages 18 through 20 (Days 43–48) at which time it lies at the coccygeal level. The tip of the vertebral coccygeal segments contains an epidermal cell rest, the coccygeal medullary vestige. The terminal filum is formed when the caudal neural tube regresses between the ventriculus terminalis and the coccygeal medullary vestige and is first present at stage 23 (Day 52). During the fetal period, the vertebral canal grows faster than the neural tube, resulting in the "ascent" of the spinal cord. At the time of birth, the conus medullaris has reached the L2–3 space in the majority of individuals,[3] and it has reached the adult level by age 3 months.

ASSOCIATED CLINICAL FEATURES OF SPINAL DYSRAPHISM

Cutaneous Stigmata

The cutaneous changes associated with OSD and TCS include the midline lumbosacral cutaneous hemangiomas (Figs. 1A and 2B), lumbosacral hypertrichosis (Fig. 1B), the lumbosacral dermal sinus (Fig. 1A), the midline lumbosacral subcutaneous lipoma (Figs. 1A and 2), and the lumbosacral skin appendage (Fig. 2A).[27] The cutaneous stigmata of OSD are present in approximately 50%[70] of patients who present with TCS.

Neurological Orthopedic Changes

Progressive neurological changes including radicular pain, weakness, asymmetric hyporeflexia, spasticity, sensory changes, and bowel/bladder dysfunction occur in approximately 75% of patients presenting with TCS.[48,70] Some authors have divided the neurological symptoms into upper motor neuron and lower motor neuron symptoms[11] and consider that the lower motor neuron symp-

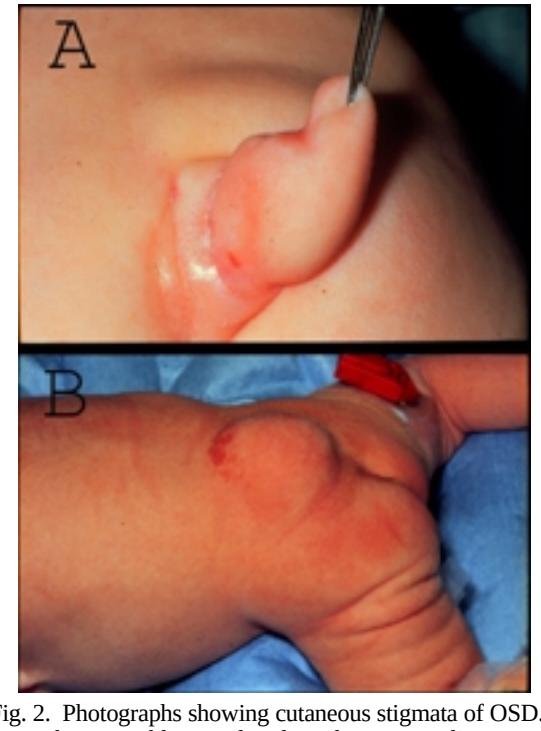

Fig. 2. Photographs showing cutaneous stigmata of OSD. A: A skin tag that resembles a tail and a subcutaneous lipoma are displayed. B: A subcutaneous lipoma and a cutaneous hemangioma are shown.

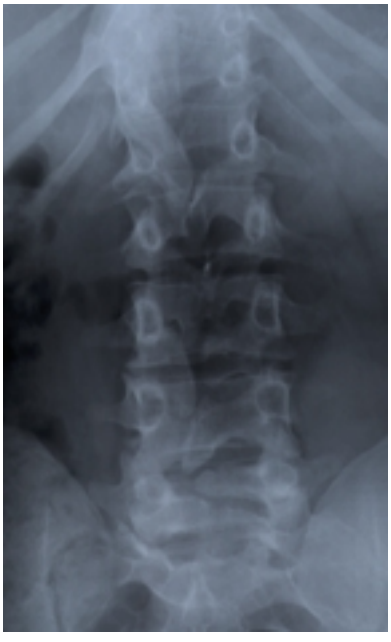

Fig. 3. A plain anteroposterior x-ray film of the lumbosacral spine revealing typical, widespread posterior fusion defects of the laminae.

toms may be due to local compression of the cord, nerve root dysgenesis, or local nerve root damage but may not be related to tethering. The upper motor neuron symptoms, however, are believed to result from the ischemic damage to the neural tissue caused by tethering.[11]

The neuroorthopedic syndrome[21,26,41] includes foot deformities, limb-length abnormalities, muscular atrophy of the legs, gait disturbance, limb pain, and scoliosis. Approximately 75% of patients with TCS present with orthopedic anomalies.[48]

Vertebral Anomalies

Bone anomalies found in patients with TCS include bifid vertebrae, laminar defects (Fig. 3), hemivertebrae, sacral aplasia, sacral agenesis and multiple segmentation errors[70] and may be found in approximately 95% of patients with TCS.[70] The bifid spinous process at L-5 or S-1, occurring in 30% of the normal population of children aged 1 to 10 years, itself is not a pathological problem.[4]

Anorectal Anomalies

Occult spinal dysraphism has been found to be associated with various anorectal and urogenital malformations including cloacal exstrophy, imperforate anus; vertebral defects, anal atresia, tracheoesophageal fistula, radial limb, and renal dysplasia; and bladder exstrophy.[12,71] Approximately 10 to 50% of the patients with anorectal anomalies will also have an element of OSD.[12,71]

OCCULT DYSRAPHIC ELEMENTS

Intraspinal anomalies common to OSD include the lipomyelomeningocele, the dermal sinus, diastematomyelia, the tight terminal filum, the neurenteric cyst, the terminal myelocystocele, and meningocele manqué.

SPECIFIC DYSRAPHIC ELEMENTS

Lipomyelomeningocele/Spinal Lipoma/Fatty Filum

Fatty accumulations within the spinal cord represent 70% of the lesions associated with tethering[55] and take three different forms. The lipomyelomeningocele is a subcutaneous lipoma within the spinal cord that extends through a defect of the lumbosacral fascia, lamina, dura, and pia into a low-lying spinal cord.[25] It is the most common form of spinal lipoma. Patients with these lesions usually come to clinical attention within the first few months to years of life.[52] A subset of the lipomyelomeningocele is the lipoma of the conus medullaris.

The intradural lipoma (spinal cord lipoma) is a rare intramedullary lesion that is usually found within the thoracic spinal cord.[1] It is not associated with cutaneous or bone anomalies and often presents with symptoms of spinal cord compression.[1]

The fatty filum involves fatty infiltration the whole length or part of the terminal filum. The fat within the short, thick filum is discernible by unenhanced CT or MR imaging.[46] The occurrence of incidental fat within the terminal filum in the normal adult population has been estimated to be 3.7% in cadaveric studies[22] and 1.5 to 5% in MR imaging studies.[6,72]

Lipomyelomeningoceles may be diagnosed by the associated subcutaneous lumbosacral mass that is found in approximately 90% of patients[48] or by sagittal MR imaging (Fig. 4) or ultrasonography studies.[48] For those who come to medical attention before the age of 1 year, 59% of patients with the lipomyelomeningocele or fatty filum are asymptomatic whereas 41% are symptomatic. Of those patients with symptoms in one study, 60% presented with urological symptoms and 58% with neurological symptoms, whereas 58% presented with orthopedic abnormalities (extremity abnormalities and scoliosis).[7] Of patients presenting with motor deficits, 50 to 70% do not experience improvement after surgery.[32,60] Surgical intervention in the symptomatic patient has been advocated by many

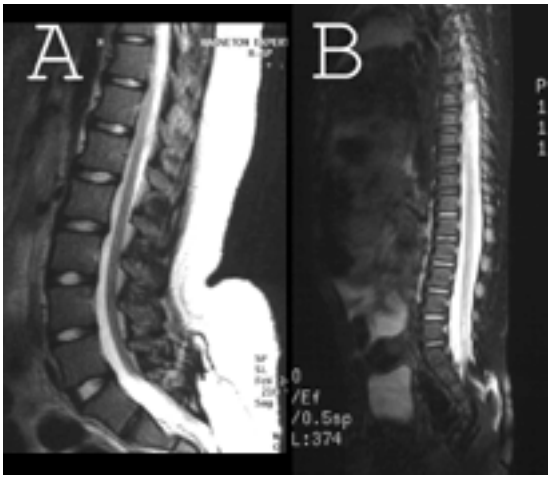

Fig. 4. Sagittal MR images demonstrating a lipomyelomeningocele. A: The spinal cord extends to the level of the sacrum. An associated superficial subcutaneous lipoma is discernible. B: The spinal cord extends to the level of the sacrum. A syrinx is present.

authors to prevent further decline in the neurological status.[7,33] A few authors believe that some of the lower motor neuron symptoms and orthopedic symptoms cannot be prevented by surgery.[11]

The operation for lipomyelomeningoceles has been significantly advanced by the use of the carbon dioxide laser and the ultrasonic aspirator.[7,52] Intraoperative monitoring of nerve roots may be useful to distinguish those that are functional from those that are not.[74] Once the spinal cord has been untethered, closure and enlargement of the dura with a graft material has been recommended.[52,74] In one study[7] of those patients younger than 1 year of age who underwent an untethering procedure for lipomyelomeningocele or fatty filum and who presented with motor, urological, or orthopedic symptoms, 39% improved, 58% stabilized, and 3% worsened as a result of surgical intervention.[7]

Surgical intervention for the lipoma of the filum in the symptomatic or asymptomatic patient has been well described and is well tolerated.[48,74] In the symptomatic group, long-term follow-up review has demonstrated improvement or resolution of the presenting neurological or orthopedic abnormalities in 42.5% of patients, as well as stabilization of symptoms in 57.5%.[48] In the asymptomatic group, the great majority of patients remain asymptomatic in the long term and suffer no neurological worsening as a result of the operation.[48,74]

Surgical intervention for the asymptomatic lipoma of the conus medullaris has been an area of controversy,[11,48,74] primarily because of the paucity of studies in which the natural history of this disorder is detailed. Many authors have advocated the use of early prophylactic untethering to prevent deterioration,[7,48] noting that asymptomatic patients rarely became symptomatic after the surgical procedure[32] and that a minority of symptomatic patients experienced reversal of their preoperative deficits.[7,48] Other authors have maintained that, in their series of patients, prophylactic untethering may not prevent some deterioration, and, because the natural history of the asymptomatic lipoma of the conus medullaris is not clearly known, prophylactic untethering may not be warranted.[74] A recent prospective study in which the authors examined the influence of surgery on the appearance of upper motor neuron signs in newborns with a tethered spinal cord (of various causes)[11] included 22 consecutive patients, 17 of whom had a lipomyelomeningocele. In approximately 29% of the patients with a lipomyelomeningocele, upper motor neuron signs were present at birth, and 60% of the asymptomatic patients with a lipomyelomeningocele developed upper motor neuron signs over a short period of 2 to 24 months postbirth. Approximately 70% of the patients with a lipomyelomeningocele in this prospective study underwent untethering of the spinal cord because of either the presence of upper motor neuron signs at birth or the development of those signs during follow up.[10,11]

The majority of authors[7,32,48,52] propose early prophylactic untethering in patients with asymptomatic lipoma of the conus medullaris because of the low rate of neurological worsening (3–4%) resulting from the operation and because of the better neurological outcome at follow up of the asymptomatic patients as compared with that of symptomatic patients.

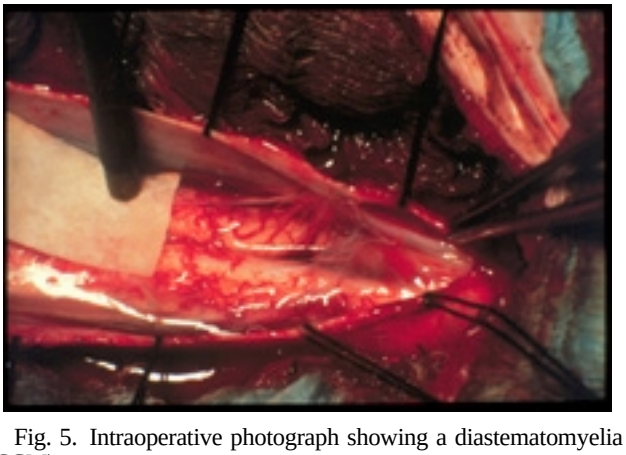

Fig. 5. Intraoperative photograph showing a diastematomyelia (SCM).

Diastematomyelia (SCM)

Diastematomyelia, also referred to as SCM, refers to the splitting of the spinal cord, conus medullaris, or terminal filum in the sagittal plane into two not necessarily equal halves.[26,56] A united theory of SCMs describes a Type I malformation as a split cord residing in a common dural tube and a Type II malformation as a split cord divided by a bone spur with each hemicord residing in a separate dural tube.[56] A thick cutaneous hairy patch usually overlies the region of the diastematomyelia.[56,59] Diastematomyelia may account for up to 25% of OSD cases.[2] Bone anomalies are present in 85% of cases, and scoliosis is present in 50% of cases of diastematomyelia.[48] In 91% of cases, the two hemicords reform a single cord below the site of division.[48] In 50% of cases, the hemicords are contained within a single dural tube (Type I malformation; Fig. 5) without a midline bone spur.[48,56] Tethering may occur as a result of dorsal tethering bands between the hemicords and dura or from a thickened terminal filum.[54] In the other 50% of cases, a median septum divides the hemicords, which are contained within separate dural tubes (Type II malformation).[48,56] The median septum lies external to both dural tubes.[48,56] Axial CT or CT myelography, or MR imaging (Fig. 6) is useful in demonstrating the hemicords and median septum.[48] Surgical intervention may involve resecting the median septum and dividing a thickened filum and dorsal tethering bands.[48,56] Andar, et al.,[2] have suggested that, although surgical intervention in patients with diastematomyelia may stabilize progressive neurological and urological symptoms and may prevent the onset of neurological and urological deterioration, it may not affect the emergence of the neuroorthopedic syndrome[21] of limb-length asymmetry and foot deformities.

The Dermal Sinus

Dermal sinus tracts appear as midline dimples in the lumbosacral region[26] and may extend from the skin surface to the dura, subarachnoid space or the spinal cord,[48] thereby causing tethering. The sinus tracts may serve as a pathway for infection and may present as meningitis.[48] The extraspinal course of the dermal sinus may be visualized by CT or MR imaging whereas the intraspinal course

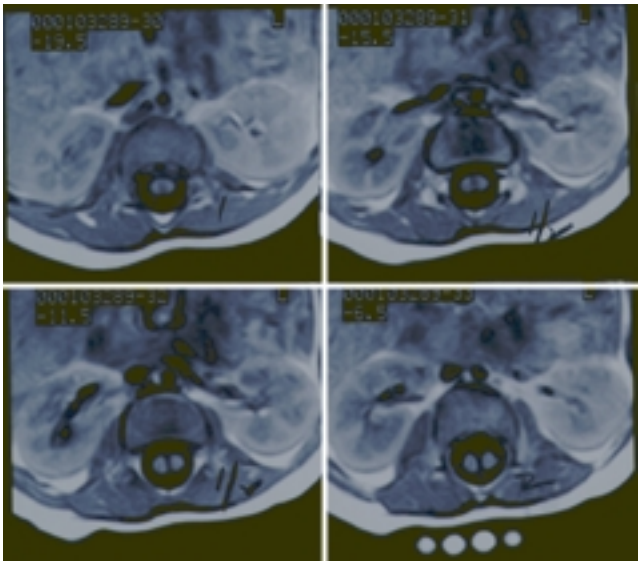

Fig. 6. Axial MR images revealing a split spinal cord (diastematomyelia). The images are displayed in a cranial-to-caudal direction, left to right and top to bottom.

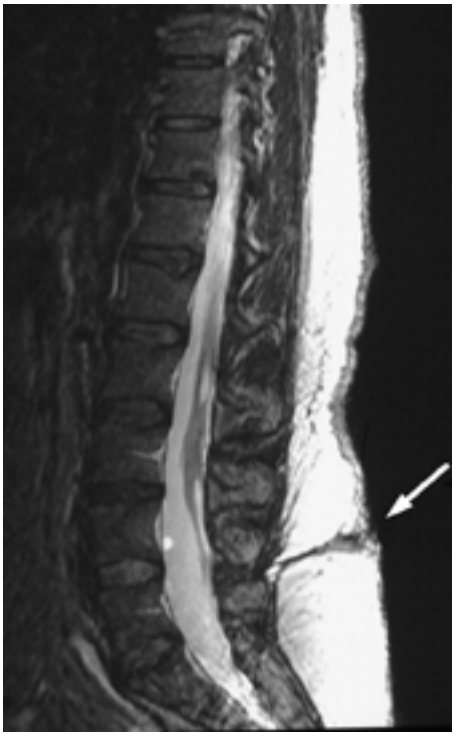

Fig. 7. Sagittal MR image revealing a dermal sinus tract *(arrow)* that extends from the skin and attaches to the spinal cord.

of the dermal sinus may be visualized with CT myelography or MR imaging (Fig. 7).[48]

Tight Filum Terminale Syndrome

The tight filum terminale syndrome refers to the clinical presence of the TCS in a patient with a low-lying conus medullaris, a terminal filum greater than 2 mm in diameter and no other tethering agents.[19,48] In 86% of the patients, the tip of the conus medullaris lies inferior to L-2.[13] The terminal filum is the tethering agent and these patients respond to sectioning of the terminal filum.[69,70] The low-lying conus and thickened terminal filum may be visualized on CT or MR imaging.[48]

Terminal Myelocystoceles

Terminal myelocystoceles represent a rare form of OSD in which elements include an expansion of the central canal of the caudal spinal cord by a cerebrospinal fluid–containing terminal cyst, which itself is surrounded by an expanded dural sheath.[47,58] Associated with this is a lipoma.[58] The inner terminal cyst communicates with the central canal of the spinal cord, whereas the outer dural sac communicates with subarachnoid space.[58] The outer and inner fluid spaces usually do not communicate.[58] Tethering results from the attachment of the myelocystocele to the inferior aspect of the spinal cord.[49,58] Terminal myelocystoceles are associated with multiple congenital defects including cloacal exstrophy, imperforate anus, omphalocele, pelvic deformity, talipes equinovarus, renal abnormalities, and ambiguous genitalia.[9] Neurosurgical intervention involves separating the spinal cord from the fluid-filled terminal myelocystocele, reconstructing the neural tube, and then incising and reapproximating the dura to recreate the subarachnoid space.[9] Patients with terminal myelocystoceles typically have no bowel or bladder control and possess poor lower-extremity function.[2] The diagnosis of terminal myelocystocele can be made by performing MR imaging or ultrasonography.[9,50]

Neurenteric Cysts

Spinal neurenteric cysts are rare congenital malformations lined by alimentary tract mucosa[18] and are formed by entrapment of endodermal tissue between a split notochord.[53] The cyst may be extraspinal with mediastinal or abdominal extension, or it may be intramedullary.[30] Whereas the cyst is frequently associated with anterior or posterior spina bifida, it may be found without any associated dysraphic anomalies.[53] Patients harboring these lesions may present with progressive signs of spinal cord compression that may be acute.[30,53] Neurosurgical intervention involves gross-total resection or partial excision of the cyst. In one study, the cyst recurrence rate in patients in whom partial excision was performed was 11%; no recurrence of the lesion occurred in those in whom gross-total resection was performed.[53] The diagnosis of neurenteric cyst can be made with MR imaging or ultrasonography, and intraoperative ultrasonography may be of value for the intraoperative localization of the lesion.[30,53]

Meningocele Manqué

Meningocele manqué refers to a dysraphic element of dorsal tethering bands composed of fibrotic or atretic neural tissue connecting the spinal cord to dura or surrounding structures (Fig. 8).[42] They are usually found incidentally during surgical exploration for other elements of OSD [31] and may exist distant from the site of obvious tethering. These tethering bands are usually found at the site of diastematomyelia; surgical treatment involves the lysis

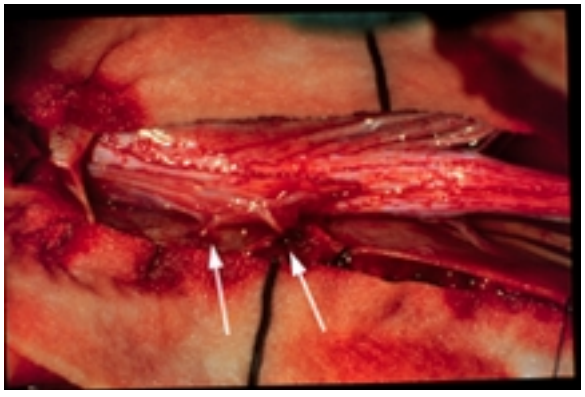

Fig. 8. Intraoperative photograph showing a meningocele manqué. Dorsal tethering bands *(arrows)* attaching the spinal cord to the dura are displayed.

of the bands. Meningocele manqué may be visualized by MR imaging or CT myelography.[31]

SYNDROMES OF THE TETHERED CORD

Tethered Cord Syndrome and the Conus in a Normal Position

In its classic form, TCS implies a low-lying conus medullaris.[23,24] There is some variation in the definition of an "abnormally" low conus. To some, below the L1–2 disc space is abnormally low,[51] whereas to others, below the inferior aspect of the L-2 vertebral body is abnormally low.[57] Both definitions refer to Barson's[3] anatomical measurements. Reimann and Anson[62] demonstrated in an autopsy study in the normal adult population that the spinal cord terminates at or above the inferior aspect of the L-2 vertebral body in 95% of the population and that it terminates at or above the L1–2 disc space in 57% of the population (Fig. 9). The conus medullaris reaches its mature adult level approximately 3 months after full-term gestation.[3] Tethered cord syndrome may occur in the presence of a conus in the normal position.[69,70] In patients who present with TCS in the setting of a conus in a normal position, the same frequency of the following is seen: cutaneous stigmata of OSD (46% compared with 52%), extremity abnormalities (39% compared with 32%), bone abnormalities (100% compared with 95%), dysraphic anomalies (62% compared with 78%), and neurological abnormalities (77% compared with 87%) as patients with TCS in the setting of a low-lying conus, respectively.[69,70] None of the patients studied with TCS in the setting of a conus in a normal position presented with urological dysfunction as the only abnormality,[70] and this patient population appeared to be different from that in whom a tight filum terminale is expressed solely in the presence of a hyperreflexic neurogenic bladder and a conus at a normal level.[34,64,65]

Tethered Cord Syndrome In Adults

The new onset of TCS in the adult population is an unusual but well-described entity.[20,57,61] The late onset of presentation may be related to the cumulative effects

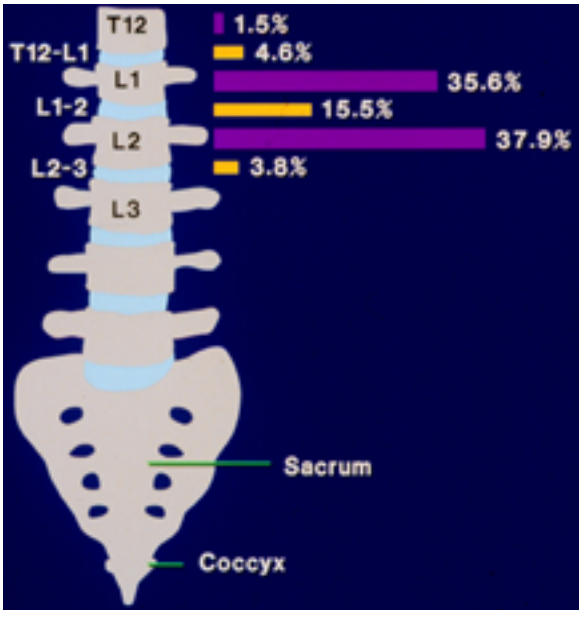

Fig. 9. Representative photograph distribution of the position of the conus medullaris in the normal adult population. The level of termination of the spinal cord in the normal adult population, as determined by Reimann and Anson, in 1944, is displayed.

of repeated microtrauma during flexion and extension.[20] Some authors have noted distinct precipitating events preceding symptoms in approximately 60% of the adult patients.[57] Such precipitating factors include heavy lifting, traumatic injury, and the lithotomy position.[57] Tethered cord syndrome in the adult population is similar to that in the pediatric population with respect to the incidence of cutaneous stigmata of OSD, neurological abnormalities at presentation, vertebral and orthopedic anomalies, and specific dysraphic elements.[20,57] In the adult population, however, TCS is accompanied by nondermatomal low-back and leg pain in 50 to 78% of patients.[20,57] Surgical untethering in the adult population may relieve pain in 80% of patients,[57] and it may improve bowel and bladder dysfunction in 38% of patients.[57] Opinions vary regarding the surgical treatment of the asymptomatic adult, particularly the elderly or infirm adult with a tethered cord. Some authors believe that surgical intervention should be offered to all individuals[20,47] because recovery of lost bowel and bladder function is low. Other authors, however, maintain that only asymptomatic adults who lead physically active lifestyles should be offered surgery.[20,57]

The Normal Conus and the Hyperreflexic Neurogenic Bladder

Urinary incontinence from bladder instability is a common problem for which patients present to a urologist.[35] Urodynamic studies in the setting of incontinence often reveal detrusor hyperreflexia.[38] The occurrence of urinary incontinence has two age-related peaks: it occurs in children less than 10 years of age and in adults between 60 and 80 years of age.[38] There is a wide variety of medical and urological treatments for bladder instability–related urinary incontinence. In the pediatric and young adult

populations, there is an annual spontaneous cure rate for urinary incontinence of 14 to 16% in each age group up to 19 years of age.[15]

Tethered cord syndrome has been postulated as a cause of the neurogenic hyperreflexic bladder even in the presence of a conus medullaris in a normal position, a normal terminal filum, an otherwise normal neurological status, no elements of OSD, and no orthopedic or vertebral anomalies.[34,37,64,65] This diagnosis of urinary incontinence in patients with a tethered cord in a normal position is one of exclusion because, by definition, the only abnormality, clinically or radiologically, is urinary incontinence. Another diagnosis of exclusion for urinary incontinence in the pediatric population includes the nonneurogenic neurogenic bladder.[28]

Several studies have included patients with a neurogenic bladder in whom the authors documented detrusor hyperreflexia, a conus in a normal position, but no elements of OSD, and no orthopedic, vertebral, or neurological anomalies.[34,36,37,65] These patients underwent division of the terminal filum. Table 1 summarizes data regarding the presentation and outcome of the patients in these studies. Kondo, et al.,[36] presented 27 patients treated over a 5-year period, who had a tight filum and a conus in a normal position. In five patients there were also cutaneous stigmata of OSD. All 27 patients underwent sectioning of the filum. Only 27% of the patients experienced improved bladder function as determined by urodynamic testing at the 6-month follow-up visit, whereas 44% experienced improvement at 26 months. The increase in improvement over time was believed to have resulted from a spontaneous cure rather than from the untethering of the spinal cord. The authors concluded that, although a tight filum might be the cause of intractable urinary incontinence, resection of the filum was not recommended because of the low success rate.[36]

In a more recent study, Selçuki, et al.,[65] described a group of 17 patients treated over 6 years who had urinary incontinence with documented detrusor hyperreflexia and a conus in a normal position. In one patient, there were independent anomalies associated with OSD. All patients underwent sectioning of the filum. Immediately following surgery, 96% of the patients noted improved bladder function, whereas at 1 month postoperatively 58% had improved function, as determined by urodynamic testing.[65] The authors concluded that surgical untethering of the conus medullaris in a normal position should not be the sole treatment for incontinence from bladder hyperreflexia, although it was an effective treatment.

TABLE 1

Summary of studies proposing a neurogenic hyperreflexic bladder resulting from a tethered spinal cord with a conus medullaris in a normal position

Articles	No. of Patients (yrs to accumulate patients)	Percentage of Patients W/ Other Independent Elements of OSD	Urodynamic Evidence of Improvement*
Kondo, et al., 1986	15 (14)	87	improvement in 67% at 20 mos
Kondo, et al., 1988	27 (5)	19	improvement in 27% at 6 mos; improvement in 44% at 26 mos
Khoury, et al., 1990	31 (3)	52	improvement in 59% at 13 mos
Selçuki, et al., 2000	17 (6)	6	improvement in 96% immediately; improvement in 58% at 1 mo

* Demonstrated after sectioning of the terminal filum.

SUMMARY OF ISSUES

The diagnosis of TCS is most easily made in the setting of clinical findings that are supplemented with neuroimaging studies. Clinical evidence of OSD including skin changes, orthopedic anomalies, vertebral anomalies, and associated anorectal malformations may suggest a tethered spinal cord. Progressive neurological/urological dysfunction localized to the conus medullaris may also suggest a tethered cord. Ultrasonography, CT scanning, MR imaging, and plain radiography assist with the localization of the level of the conus medullaris and the identification of the specific tethering agent. A patient's preoperative neurological and urological status may be further investigated by using electromyography, cystometrography, and evoked potential monitoring.

Surgical intervention for the tethered spinal cord involves the intraoperative identification of the tethering lesions, release of the spinal cord, and reconstruction to as normal anatomy as possible. The carbon dioxide laser has been found to be useful for debulking and dissecting tethering lesions and reducing blood loss. Intraoperative electromyography or evoked potentials may also be used. Surgery-related complications include the standard anesthesia-related risks, neurological worsening, cerebrospinal fluid leakage, and meningitis.[48,61]

The goal of untethering of the spinal cord is to stabilize neurological function. The presenting symptoms of motor weakness, sensory dysfunction, bladder dysfunction, and pain improve to different degrees following treatment depending on the different dysraphic lesions. Improvement of motor weakness occurs in 12 to 60%,[8,39] sensory dysfunction improvement in 40 to 60%,[8,70] pain symptom improvement in 50 to 88%,[20,70] and bladder dysfunction improvement in 19 to 67%[14,37,39] of patients undergoing spinal cord untethering. In the majority of patients who undergo untethering of the spinal cord either improvement or no deterioration of neurological status is demonstrated at long-term follow up.[2,39,48,66,70]

In patients who have previously undergone untethering procedures, a progressive neurological deterioration may signal retethering, which may occur in up to 15% of patients depending on the specific tethering lesion.[33] Long-term neurological and urological follow-up study can aid with the identification of patients with retethering of the spinal cord who do respond to reoperation for untethering.[48]

Current controversies with respect to the TCS include: 1) the untethering of the spinal cord in asymptomatic patients; and 2) the proposed neurogenic hyperreflexic bladder resulting from a tethered spinal cord with a conus medullaris in a normal position.

The many different tethering lesions of the TCS and the

unknown natural history of many of these lesions have led a few authors to propose the conservative management of some asymptomatic patients.[10,11,74] However, because early intervention is an effective prophylaxis against progressive neurological decline, which may occur precipitously, and because full recovery of neurological function after a decline is not certain, the majority of authors recommend an untethering procedure for the asymptomatic patient with a tethered spinal cord.[20,33,39,48,61,67]

The development of a neurogenic hyperreflexic bladder from a tethered spinal cord with a conus in a normal position and an otherwise normal status is a diagnosis of exclusion and has been postulated;[34,64,65] sectioning of the filum for this has been performed with varying results (Table 1).[34,36,37,65] Different results have led some authors not to recommend sectioning of the filum in this setting because of the poor success rate,[36] whereas others concluded that this was an effective treatment.[65] The interpretation of the response rate of the neurogenic hyperreflexic bladder to the sectioning of the filum is somewhat obscured by the spontaneous rate of cure for this condition. Because of the high prevalence of the neurogenic bladder of the hyperreflexic type, and the high spontaneous cure rate for this disorder, better controlled studies may provide important data regarding the efficacy of this proposed treatment.

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Manuscript received December 4, 2000.
Accepted in final form December 21, 2000.
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