

Tethered Cord Syndrome

Pankaj K. Agarwalla, BS, Ian F. Dunn, MD,
R. Michael Scott, MD, Edward R. Smith, MD*

Department of Neurosurgery, Children's Hospital of Boston, 300 Longwood Avenue, Boston, MA 02115, USA

Definition—what is a “tethered cord?”

A tethered spinal cord is best defined as an abnormal attachment of the spinal cord to the tissues that surround it. The term has acquired a number of different meanings over time. This label has been applied to descriptions of radiographic findings and to varied constellations of clinical signs and symptoms. For example, in 1976 Hoffman and colleagues [1] used the phrase “tethered spinal cord” to define a radiographic diagnosis—a spinal cord “with a low conus medullaris and a thickened filum terminale measuring 2 mm or more in diameter,” excluding other conditions such as “lipomyelomeningoceles, meningoceles, myelomeningoceles, diastematomyelia, and intraspinal space-occupying dysraphic conditions such as dermoid tumors, intraspinal meningoceles, neurenteric cysts, and teratomatous cysts” [1]—many of which today are considered typically representative of tethering of the spinal cord.

The radiographic diagnosis of tethered spinal cord is distinct from the clinical diagnosis of tethered cord syndrome, that is, the signs and symptoms believed to result from excessive tension on the spinal cord. The spinal cord most frequently is tethered in the lumbosacral region [2,3]. Ascribed clinical manifestations include pain (especially with flexion), bowel and bladder dysfunction, weakness, sensory changes, gait abnormalities, and musculoskeletal deformities of the feet and spine, such as scoliosis or clubfoot [1,2,4–11]. Cutaneous stigmata signifying an

underlying congenital defect of the spinal cord also are common [12].

Over time, the term “tethered cord” has been used interchangeably to include both the radiographic and clinical findings described here. Although the incidental radiographic finding of an asymptomatic tethered spinal cord is becoming increasingly common, more often some combination of clinical signs and symptoms results in a patient's coming to the attention of the neurosurgeon. Therefore this article focuses primarily on the clinical entity of tethered cord syndrome, with discussion of its history, pathophysiology, diagnosis, and treatment.

Brief history

Review of the early literature related to tethered cord syndrome reveals a gradual awareness that myriad causes can contribute to a similar presentation. Reports often had two common themes, a clinical scenario of progressive lower-extremity symptoms and recovery following surgical intervention. In one of the first recorded cases documenting the diagnosis and treatment of tethered cord syndrome, an 1857 report describes a young child who presented with worsening right-sided lower extremity weakness and twitching [13]. The child underwent surgical exploration of his spine, and a lesion consistent with a spinal lipoma was found. At surgery, the spinal cord was freed from its attachments to the dura, and the symptoms resolved [13].

In 1891, Jones [14] described what probably is the first true “untethering” operation in a 22-year-old patient who had developed talipes equinovarus deformities, weakness and atrophy of the lower limbs, difficulty with micturition, and pain in his feet. At operation, Jones [14] performed

* Corresponding author.

E-mail address: edward.smith@childrens.harvard.edu (E.R. Smith).

a successful division of a “dense adventitious fibrous band” within the spinal canal. Six months postoperatively, the patient was able to walk freely without pain, had improved micturition, and regained leg muscle size and strength.

Another important development in the history of tethered cord syndrome was the recognition that symptoms may be exacerbated by activity. In 1916, Spiller [15], a neurologist, described two adolescent patients who presented with symptoms of tethered cord syndrome that developed subsequent to strenuous activity. Two boys, 14 and 18 years of age, presented with leg weakness and enuresis after exercise, including flexion related to training for rowing. This new understanding—that there may be a dynamic component to the development and progression of tethered cord syndrome—resulted in an impetus for earlier identification of affected patients so they could avoid activities that could lead to spinal cord stretching, presumably the cause of the neurologic deficits [15].

In addition to a growing recognition of the distinct clinical entity of tethered cord syndrome, the literature also reflected a progressive evolution in the debate regarding timing of treatment of affected patients. As early as 1918, a correlation between prompt treatment and improved outcome was acknowledged [16]. The benefit of early treatment, coupled with increasing awareness of findings on physical examination associated with tethered cord syndrome, led to a proposal to treat asymptomatic patients prophylactically, “in the hope of obviating the development of symptoms during adolescence” [16]. Although the expeditious treatment of symptomatic patients has been accepted generally, the debate surrounding asymptomatic patients who have anatomically tethered spinal cords continues.

Tethered cord syndrome, attributed to a wide spectrum of causes and poorly understood mechanistically, remained vaguely defined for several decades [17]. In the 1950s, however, reports began to recognize the connection between disparate pathophysiologic entities, particularly diastematomyelia, spinal lipomas, and thickened fila terminalia, and a common clinical presentation [13,14,16,18–20]. In 1953, Garceau [21] conceived the terms “filum terminale syndrome,” and “cord-traction syndrome,” proposing a causal relationship between a thickened filum found on exploration of patients who presented clinically with spinal deformities and progressive neurologic deterioration. The distinction between the clinical “tethered

cord syndrome” and the radiographic “tethered spinal cord” continued with Hoffman’s [1] definition of a “tethered spinal cord” made in the 1970s.

Despite the creation of the distinct term “tethered cord syndrome” to encompass the signs and symptoms thought to be the clinical manifestations of a tethered spinal cord, the wide range of causes reported in association with this tethered cord syndrome, coupled with the continued lack of consensus regarding what constitutes the tethered cord syndrome, has resulted in the admission by one group that tethered cord syndrome constitutes, at best, “a loose diagnosis” [17,22]. In future efforts in this area, it will be important to make clear distinctions between clinical and radiographic findings.

Embryologic considerations

Although a lengthy discussion of embryology is not within the scope of this article (for a more thorough review, see Dias and McLone [23]), understanding the varied clinical manifestations of tethered spinal cord is enhanced by an appreciation of the relevant embryology. A considerable number of developmental errors can result in conditions that functionally tether the spinal cord. These congenital conditions, distinct from acquired causes of tethering (such as infection, tumor, or scar), can present in myriad ways and at different stages of a child’s maturation. A working knowledge of the embryologic processes underlying these conditions can aid the neurosurgeon in understanding and avoiding the potential hazards intrinsic to the treatment of these children.

Notochordal development

In the first few weeks of development, neurulation begins with the formation of the notochord arising from the primitive pit [23]. The primitive pit subsequently recedes caudally while the notochord elongates cranially [23]. The notochord then undergoes intercalation, fusing with the underlying endoderm to form the notochordal plate. This plate is continuous with the yolk sac and also is continuous with the amniotic sac [23].

Primary neurulation

The notochord induces formation of the neural tube dorsally from the overlying ectoderm by means of the neural groove from days 18 to 24 after ovulation [24]. This process gives rise to the cervical, thoracic, and lumbar neural tube [25].

Somites develop from the paraxial mesenchyme and represent the majority of the future vertebral column at these levels as well [25]. Most relevant to the tethered cord in primary neurulation is the closure of the neural groove. The level of final closure of the caudal neuropore corresponds to the second sacral vertebral level (S2) [24], suggesting that spinal malformations arising from S2 or above probably result from disordered primary neurulation [23].

Secondary neurulation

During the time of primary neurulation, the primitive streak regresses to form the axial mesenchyme of the caudal eminence (also known as end-bud [24]), which extends from the site of the neurenteric canal to the cloacal membrane [25]. The caudal eminence provides the cells for the formation of the neural tube caudal to somite 31, corresponding to the future S2 level. Once primary closure is complete, secondary neurulation from the caudal eminence begins but not in the form of a folding neural plate as in primary neurulation. Rather, a “neural cord” forms with a central canal continuous with the more rostrally formed primary neural tube; this distinct process of secondary neurulation helps explain the clinically relevant pathophysiologic entity of caudal agenesis [24,26,27].

Ascent of conus and relationship with meninges

Beginning at postovulatory day 43 to 48, the conus medullaris “ascends” relative to the vertebral bodies through two mechanisms: (1) differential growth of bony vertebrae compared with the neural tissue of the spinal cord and (2) retrogressive differentiation during which the caudal cord loses much of its thickness and character [23]. The conus does not ascend throughout childhood and remains at approximately its birth position of L1-2; a cord at L2-3 or above is considered within normal range [28]. Wolf and colleagues [29], using ultrasound, found that the conus is still “ascending” from L2-4 to L1-2 during postmenstrual week 30 to 40 and generally achieves its normal position of L1-2 after postmenstrual week 40. The clinical relevance of these data is that any patient who has a conus found at L3 or below should be considered for evaluation of tethered cord syndrome.

In addition to the formation of the neural tube, the spinal cord must be invested with membranes and a vasculature. These generally are considered

to be derived from the mesodermal layer, although there has been debate on their origins [30–32]. In both open and closed spinal dysraphisms, it is clear that the usual meningeal stratification often is abnormal, with the potential for improperly located tissue (eg, subdural extension of adipose tissue in lipomyelomeningocele). In abnormal development, therefore, the surgeon must be aware of unusual meningeal arrangements, both between the dura and the leptomeninges and between the dura and the conus.

This overview of the embryology helps explain the development of the abnormal anatomy that results in a tethered spinal cord. Although this information can be invaluable to understanding and interpreting physical findings and imaging studies, it is important to appreciate a distinction between the anatomic findings of a tethered spinal cord and the functional problems that produce the symptoms of tethered cord syndrome. Some of the symptoms that are part of the clinical presentation of these patients may be caused by intrinsic, congenital defects in the nerves and spinal cord, and, as such, cannot be remedied by surgical intervention. In contrast, some symptoms are secondary to reversible causes that are amenable to surgical treatment. It therefore is important for the treating physician to establish and document a baseline examination before undertaking any potential intervention to help distinguish between pre-existing and recurrent problems.

Causes

Any process that tethers the spinal cord can result in a patient who has tethered cord syndrome. Children can be born with normal anatomy and develop a tethered cord through an acquired process, such as infection, scarring, or tumor. Although these acquired (secondary) causes are important, this section focuses on congenital (primary) causes of tethered cord. The previous review of the embryology of the developing spinal cord provides a context for presenting the more commonly encountered congenital causes of tethered spinal cord discussed in this article.

Abnormal secondary neurulation and disorders of caudal eminence

Because the filum terminale and the caudal spinal cord are formed from the caudal eminence through secondary neurulation, disorders in this

process can lead to conditions in which the caudal cord might be tethered. The simplest form of such conditions is a filum terminale, which can be thickened, potentially with lipomatous tissue (the so-called "fatty filum"). Hoffman and colleagues [1] have suggested that a diameter of 2 mm or greater should be considered an abnormally thickened filum. Impaired canalization of the growing secondary neural tube (the neural cord) with cells capable of growth and differentiation, particularly preadipose tissue, is believed to be the cause of both the thickened and fatty filum terminale [33]. The fatty filum commonly is associated with cases of imperforate anus, suggesting a common timing of pathogenesis during development [33].

Terminal myelocystoceles, also thought to arise from disordered secondary neurulation, are found at the terminal end of the developing neural tube. These myelocystoceles usually contain two sacs, one a dilation of the embryologic terminal ventricle and another a dilated and ectatic dural and arachnoid sleeve [23]. They often are associated with a lipoma (lipomyelocystocele), and, because the mesenchyme of the caudal eminence also forms many of the structures of the hindgut, terminal myelocystoceles also are found commonly with abnormalities of other caudal systems, particularly in the complex of omphalocele, extrophy, imperforate anus, and spinal malformations (OEIS syndrome) [23]. The disorder is believed to be mesenchymal, and the surface ectoderm and skin usually are intact. The finding of dorsal bony dysraphism is common also.

Abnormal secondary neurulation can lead to a variety of other complex spinal dysraphisms in the caudal region. Termed "caudal agenesis" or "dysgenesis," these congenital malformations involve abnormal or incomplete formation of caudal elements of the embryo. They arise from problems with canalization of the caudal neural cord (the secondary neural tube) or in the process of retrogressive differentiation during the ascent of the conus [34]. Because the filum terminale forms as a gliopendymal strand during retrogressive differentiation, caudal agenesis (especially the simplest form, sacral agenesis, which affects coccygeal spinal levels) often leads to an elongated and tethered conus.

Caudal agenesis often is accompanied by other caudal hindgut and genitourinary malformations. This process can be viewed as a spectrum, ranging from a simple imperforate anus to complete caudal agenesis with sirenomelia (mermaid

syndrome), which shows malformation of limb buds, genitourinary apparatus, caudal neural tube, and anorectal system [23]. In between are a host of syndromes associated with caudal agenesis, congenital abnormalities, and tethering of the spinal cord. In particular, a tethered spinal cord is a common finding in patients who have OEIS, vertebral, anal, transesophageal, radial, and renal abnormalities (VATER syndrome), or the Currarino triad [34].

OEIS syndrome is defined by the presence of an omphalocele, extrophy of the cloaca, an imperforate anus, and spinal malformations, often including a tethered spinal cord. VATER syndrome refers to a presentation with the combination of vertebral anomalies, an imperforate anus, a tracheoesophageal fistula, and renal-radial anomalies. The Currarino triad, caused by a genetic defect in a homeobox gene at 7q36, includes three findings: an anorectal malformation, a presacral mass (usually an anterior myelomeningocele), and sacral bone abnormalities [35]. Patients who have Currarino triad also have distinct clinical features, including a narrow pelvis, flattened buttocks, a short intergluteal cleft, a prominent iliac crest, absent coccyx/sacral elements, and impaired lower extremity motor function [34]. These clinical findings are linked by a common error in embryologic development, a malformed caudal eminence resulting in abnormal canalization of secondary neural tube, subsequently leading to a dysfunctional ventral spinal cord [35].

Lipomas

Lipomas can arise in numerous locations at the caudal end of the spinal cord. Presumably, lipomas arise after the completion of primary neurulation but before secondary neurulation and arise from embryonic mesodermal tissue that has infiltrated into an abnormal area [33]. Lipomas in the filum terminale were discussed earlier; the focus here is on lipomas affecting the conus medullaris. These lipomas most are commonly subpial, although a small number can be subdural. Subdural lipomas are infrequently associated with tethering and more commonly present like a mass lesion with cord compression [33].

More commonly, however, lipomas of the spinal cord occur in the lumbosacral region and have an associated dural defect. Chapman [36] classified such conus lipomas into three categories, those arising from (1) the terminal end of the conus, (2) the dorsal surface of the conus, or (3)

both the terminal end and dorsal surface of the conus. The most important clinical distinction is whether the lipoma involves neural tissue (ie, conus/cauda) or not (eg, filum) [33]. Approximately 75% occur in the conus, approximately 15% to 20% in the filum, with the remainder involving both the conus and filum [37]. In addition, lipomatous tissue can infiltrate almost any caudal spinal defect to give rise to malformations including the atypical forms lipomyelocele, lipomyelomeningocele, and lipomyelocystocele.

In a large series by Pierre-Kahn and colleagues [37], 63% of lipomas were classified as atypical. These embryologically distinct entities have direct relevance to surgical planning. The exact embryologic mechanism underlying this pathogenesis has been debated, with alternative theories proposed by several groups [30,38]. Several of these models propose that traction on the spinal cord caused by the lipoma may be asymmetric. This theory has been supported by findings at surgery, where eccentric lipomas have been observed to cause the affected side of the cord to be directed more posterolaterally, stretching the ipsilateral nerve roots [23,33].

Histologic analysis of fat supports the concept of primary and secondary neurulation contributing to distinct pathophysiologic processes, because lipomas rostral to S2 often contain typical fat cells, whereas lipomas caudal to S2 often contain other mesenchymal cell derivatives including a thick fibrous stroma as well as tissue with characteristics suggestive of muscle or bone [23,33]. Another study by Pierre-Kahn and colleagues [37] has shown that lipomas in their series contain nonadipose tissue apparently derived from all three primary germ layers.

Dermoid/sinus tract

Whereas lipomas are thought to occur from premature disjunction, delayed disjunction is the proposed cause for both dermal sinus tracts and dermoid/epidermoid tumors [23]. At a certain point in normal neural tube fusion, the neural tube separates from the cutaneous ectoderm, allowing mesenchymal cells to invade and separate the neural tube from the surface ectoderm. If the surface tissue does not separate successfully from the central nervous system, residual tissue or sinus tracts can develop in association with the central nervous system.

Another theory proposes that a more general disorder of gastrulation in which two paired

notochordal anlagen do not fuse properly results in the inappropriate deposition of ectodermal tissue between the notochords, engendering the development of dermoids, sinus tracts, and epidermoid tumors [39]. According to this theory, the separation of tissue layers is delayed, resulting in cutaneous ectodermal cells being carried in from the skin and subsequently residing at the site of neural tube closure. These cells can develop into a dermal sinus tract, an epidermoid, or a dermoid [23].

Complex spinal dysraphisms

Complex spinal dysraphisms are disorders affecting all three primary germ layers during embryogenesis, and they share a common embryologic basis. The common complex spinal dysraphisms that can lead to tethering are spina bifida, split cord malformations (ie, diastematomyelia and diplomyelia), and neurenteric cysts. They can occur as open neural tube defects but more often are closed defects [23]. (For a more detailed review, see Dias and McLone [23].)

One theory has been proposed by Dias and Walker [39], who believe that these malformations arise when the anlagen of all three germ layers are established in gastrulation. During the formation of the notochordal process, paired bilateral notochordal anlagen come together to form a single notochord with a narrow primitive streak. Should these bilateral anlagen become separated, two distinct spinal cords would develop. The space between the hemicords also could give rise to tissues from each of the three germ layers: endoderm (neurenteric cysts), mesoderm (bony spurs, muscle, fat), and ectoderm (dermoid/epidermoid tumors) [39]. This proposed mechanism of complex spinal dysraphisms also supports the unified theory of split cord malformation proposed by Pang [40,41], who posits a common embryologic basis for type I split cord malformations (diastematomyelia, two spinal cords with two dural sleeves) and type II malformations (diplomyelia, two spinal cords sharing one dural sleeve).

Split cord malformations (diastematomyelia and diplomyelia) often are associated with tethered cords, with the tethering often attributed to bony or fibrous spurs and/or thickened filum. In a series of 31 children, all split cord malformations below T7 were associated with a low-lying conus and a spinal lipoma or fatty filum [40,41].

Neurenteric cysts often are found on the ventral side of the spinal canal and consist of a fluid-filled cyst that may communicate with the

gastrointestinal tract through a vertebral defect such as a hemivertebra or butterfly vertebra [34]. The neurenteric cyst itself can cause compression, but its adherent fibrous bands also can result in tethering [34]. They usually are intradural and extramedullary, and their origin is debated, although positive immunoreactivity for carcinoembryonic antigen suggests endodermal origins [42].

In all these cases, tethering can occur when improperly placed mesenchymal tissue creates various abnormal structures such as bony spurs, fat, and fibrous bands that impede the normal ascent of the conus or attach to tissue at inappropriate locations, causing a tethering effect.

Myelomeningocele, meningocele, meningocele manqué

Myelomeningocele, meningocele, and meningocele manqué reflect abnormal development during primary neurulation or immediately after during the formation of the meninges. In myelomeningocele, the spinal cord does not fuse dorsally, leaving neural tissue known as the “neural placode.” The groove in the neural placode is the remnant of the central canal [43]. In meningocele, the neural tube fuses properly, but the dura does not fuse correctly, creating a cystic lesion that is often skin-covered. In both cases, tethering can occur as functional cord attaches itself dorsally either to dura or to surface ectoderm. An interesting case of meningocele known as the “meningocele manqué” (the “missing” meningocele) occurs when a meningocele has formed during embryogenesis but has healed spontaneously or scarred creating a dorsal band. These dorsal bands can extend from intrathecal structure into the dura or outside structures creating a significant tethering effect [34]. The dorsal band of meningocele manqué may reflect a fibroneurovascular stalk derived from the same endomesenchymal tract that is the basis for split cord malformations [41].

Clinical presentation and evaluation

In the original description of the tethered cord syndrome, Hoffman and colleagues [1] chose in their subtitle to refer to the syndrome’s “protean manifestations,” a term that comes from the Greek god Proteus who would change shape. “Protean” is an apt term for the tethered cord syndrome because its presentations are as varied as its causes. This section describes some common presentations and highlights some unique findings.

Cutaneous findings

Cutaneous findings are commonplace in closed spinal dysraphism. A retrospective study by Guggisberg and colleagues [44] examined the diagnostic value of midline cutaneous lesions in the lumbosacral region for closed spinal dysraphism. A large number of cutaneous lesions were reviewed, including cutaneous lipoma, tail, dermal sinus, atypical dimple, deviation of gluteal crease, hamartoma, hemangioma, port-wine stain, hypertrichosis, and pigmentary nevus, among others, with the recommendation of MRI testing if patients had two or more of the listed cutaneous lesions or one high-risk lesion such as a lipoma, tail, or dermal sinus [44]. Other lower-risk lesions such as an atypical dimple, a deviation of the gluteal crease, or an unclassified hamartoma suggest the need for ultrasound evaluation before 6 months of age or an MRI after 6 months. The rationale for the age difference is that the acoustic window to the spine closes at approximately 3 to 6 months. Other isolated findings such as hypertrichosis or vascular abnormalities have a lower likelihood of being associated with spinal cord tethering in the absence of any other signs or symptoms [44].

A common finding in closed spinal dysraphism is a palpable subcutaneous lipoma, often associated with a cutaneous hemangioma [33]. Dimples often are cited as a common finding, but it is important to distinguish a sacrococcygeal dimple as a marker for a dermal sinus tract or more dangerous abnormality from a more benign coccygeal dimple [23]. Sacrococcygeal dimples are almost always cranial to the intergluteal cleft, and the intergluteal cleft is often abnormal or deviated. Its distance from the anus is more than 2.5 cm, its diameter is larger (>5 mm), and cutaneous stigmata often are present [45]. Simple coccygeal dimples usually are intergluteal and smaller than sacrococcygeal dimples with no significant cutaneous abnormalities and are thought to be a remnant of the primitive pit with some cells from the caudal eminence [23,45]. There are rare reports associating low, coccygeal dimples and presacral masses that might warrant a conservative approach including ultrasound examination and digital rectal examination [33].

As a rule of thumb, a lesion rostral to the gluteal cleft often is associated with neurosurgical disease and should be considered for detailed imaging evaluation, whereas lesions within or caudal to the gluteal cleft are less likely to require neurosurgical attention.

Neurologic findings

A variety of neurologic findings can be present in patients who have tethered cord syndrome. Common findings change with age and depend on the underlying cause of the tethered cord [34]. A full neurologic examination is vital for initial diagnosis and for establishing a baseline for follow-up. In infants, one may find decreased spontaneous leg movement, abnormal reflexes, foot asymmetry, and leg atrophy (occasionally hidden by baby fat). Toddlers often show developmental delay in acquiring gait or have an abnormal gait. Older children have asymmetric motor and sensory dysfunction, painless foot burns (trophic ulcerations), hyperreflexia, and back and leg pain that often is worsened with flexion or vigorous physical activity. Young adults have similar pain and reflex changes but may present with predominantly sensory dysfunction. Generally, however, pain and motor dysfunction are more prominent, perhaps because the ventral aspect of the conus medullaris derives primarily from the secondary neural tube, whereas the primary neural tube extends slightly dorsally into the conus during development [34].

Orthopedic findings

Common deformities include clubfeet (often equinovarus), asymmetry in leg length, trophic ulcerations of the foot in advanced cases, atrophy of lower leg muscle occasionally masked by baby fat in an infant, hip subluxation, and scoliosis [33]. These conditions call for orthopedic consultation and mandate treatment of the underlying tethering by the neurosurgeon. In the older patient, severe scoliosis, gait change, leg weakness and atrophy, and pain can occur either as an exacerbation of a previously undiagnosed tethering or as a retethering of the cord [33]. Spinal radiographs, as discussed later, are useful in cases of scoliosis and are recommended when vertebral deformities are present.

Urologic function and assessment

Urologic decline is one of the most important indicators for early and definitive treatment. As a child grows older or as urologic function deteriorates, it often becomes more difficult to restore urologic function after untethering [33]. Common symptoms include frequent urinary tract infections, abnormal voiding, urinary incontinence, and fecal soiling. Incontinence and

infections are more common in older children and young adults [34].

A careful history and physical examination are important screening tools for evaluating urologic function, especially in infants and young children. Particular attention should be given to histories that include a loss of previously attained milestones in continence that is progressive in nature. Treatment for any urinary tract infection and a full work-up for any hematuria should be performed [46].

Urodynamic assessment provides quantifiable evidence of neurologic dysfunction in the setting of tethered cord. Evaluation includes urodynamic measurements by simultaneous cystourethrography/cystometry and sphincter electromyography. Sacral innervation can also be tested by examination of perianal sensation, anal sphincter tone, the bulbocavernosus reflex, and voluntary sphincter control [46].

Electromyographic measurements include a bulbocavernosus reflex latency time and an electromyographic examination of the perineal floor muscles. If the sympathetic pathways are damaged, incontinence results from lack of internal sphincter control. If parasympathetic pathways are damaged, an areflexic and either hypotonic or hypertonic bladder will result. Patients who have hypertonic bladders may be treated with anticholinergics and self-catheterization in addition to treatment of any underlying neurologic proximal cause, such as spinal cord tethering [46]. Detrusor dyssynergia, another common finding with hypotonic and hypertonic bladder, is caused by a lesion between the brainstem and the sacral spinal cord [33]. Both dyssynergia and hypertonic bladder with high intravesical filling pressures require treatment to prevent further upper urinary tract disease. High intravesical pressures have been shown to be predictive of future urinary tract problems in patients who have tethered cord syndrome [47].

As discussed later, urologic examination and assessment are vital in the follow-up after surgical treatment of a tethered cord, because new dysfunction or a postoperative progression of dysfunction may herald retethering of the cord. Successful treatment of a tethered cord may lead to stabilization or even reversal of urologic dysfunction. In addition, preoperative urodynamics can document pre-existing problems that may not be clinically evident immediately but may present later in life (for example, in younger children who later may have difficulty in toilet

training). These preoperative tests may provide evidence that problems discovered later in life are not a result of the surgical intervention or necessitate additional treatment. Nevertheless, the consensus view is that early, aggressive treatment, particularly in the infant and young child, can help significantly with urologic function in the context of a tethered cord [48,49].

Imaging

Ultrasonography

Ultrasound imaging, although not very useful for surgical planning or proper spinal anomaly evaluation, can have a role as a relatively quick and easy screening tool in young children. The acoustic window into the lumbar spine in the infant closes in the first months of life. Ultrasound is best able to detect the position of the conus, the presence of any fat, and decreased spinal cord motion, any of which might indicate tethering [34]. Should ultrasonography be performed, it can be useful to image with the patient's head elevated to distend any potential meningocele or closed spinal dysraphism [50].

Plain-film radiography

When a congenital defect with a vertebral or bony component (such as midline bony spurs in split cord malformation type I) is suspected, plain-film radiographs may be useful but in most cases have been largely supplanted by MRI and CT. In complex spinal dysraphisms, one should look for anomalies in the laminae, vertebral bodies, disc spaces, or pedicles [34]. Widening of the spinal canal, as evidenced by an increased interpedicular distance or scalloping of the posterior of the vertebrae, is particularly evident on plain-film radiographs [34]. More global assessments, including any change in the number of vertebra or obvious malformation of an individual vertebra, should be made also. In cases of caudal agenesis, radiographic evidence of absence or splitting of the sacrum should be noted.

The one particularly helpful role of plain-film radiography is in assessment of spinal curvature. Radiographs can be measured to evaluate the degree of kyphosis, lordosis, or scoliosis.

CT and MRI

Previously, the standard of diagnosis for tethered cord was lumbar myelography. CT

myelography (CTM) later became accepted, and criteria were established for the diagnosis of a tethered cord: a low-lying conus (below L2-L3), a thickened filum (>2 mm), or fat in the filum. CTM has proven useful when examining the axial plane to see the relationship between lipomas, subarachnoid space, nerve roots, and neural placode, if there is one. Pang [40] also has suggested that CTM with iohexol is superior to MRI in the diagnosis of split cord malformations. Through varying window settings, CTM can elaborate on bony spurs and other midline structures more clearly, and in the case of a bony spur without a marrow cavity, it is more sensitive than T1-weighted MRI, which loses the signal void of the bone marrow. MRI, however, better delineates intramedullary syrinxes and is comparable to CTM for tethering lesions such as thickened fila, lipomas, and dural adhesions [40].

More recently, however, MRI has replaced CTM myelography as the reference standard for the diagnosis of various causes of the tethered cord syndrome. MRI is particularly good at highlighting fat on T1-weighted imaging (Fig. 1). Nevertheless, a small number of reports have described patients who have tethered cord syndrome without a low-lying conus or other clues from imaging [4,6]. MRI is well suited to identifying the level of the conus relative to vertebral bodies, the presence of a syrinx, or visualization of other pathologic processes. MRI also is able to define the anatomy of other causes of tethered cord, such as the anatomy of terminal or multiple lipomas (Fig. 2), the presence of congenital lesions (such as dermoids), and the presence of myelomeningocele.

Pathophysiology

The mechanism by which tethering produces its effect on the spinal cord has long been the subject of debate [17]. Yamada and colleagues [51] performed the first scientific experiments to investigate the pathophysiologic basis of tethered cord. Using spectrophotometry on human and animal spinal cords to measure reduction/oxidation changes in cytochrome c, they demonstrated decreased mitochondrial oxidative metabolism with constant or intermittent cord stretching, particularly at higher forces of traction. Furthermore, they proposed that local hypoxia might contribute to the pathogenesis of symptoms in patients who have tethered cords. The hypothesis that the spinal cord may undergo ischemic changes in

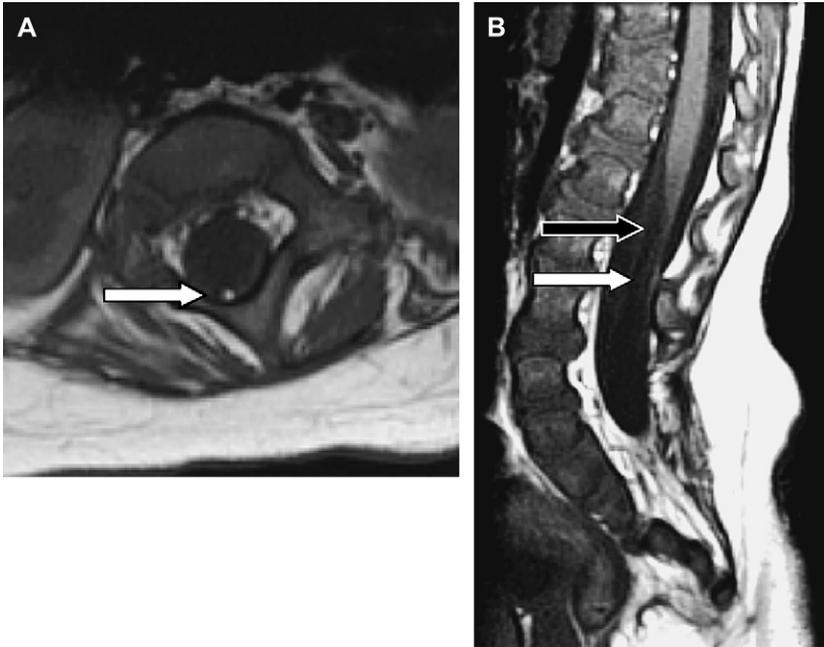


Fig. 1. MRI showing a fatty filum (*white arrow*), bright on T1 image (*A*), with low-lying conus (*black arrow*) (*B*).

response to tension was bolstered by the findings of experiments using laser Doppler to monitor the microcirculation of human spinal cords during untethering operations [52]. Using this approach, the authors demonstrated increased blood flow after surgical release of the spinal cord by untethering.

Sarwar and colleagues [3] and Tani and colleagues [53] provided evidence that helps explain why symptoms of tethered cord are referable predominantly to the caudal spinal cord and roots. Their work revealed that caudal cord traction produces primarily local elongation in the lumbar cord and that the filum acts as a distensible buffer to prevent cord stretching [3,53]. Thus, any process that reduces the filum's distensibility and role as a buffer leaves the cord more susceptible to stretching forces. A shortened, thickened, or fatty filum may exert this effect and produce injury to the distal spinal cord.

It also has been demonstrated that flexion of the torso increases longitudinal tension if the cord is tethered and increases local compression of the spinal cord if a mass is present [54]. This finding suggests a dual mechanism of injury in the setting of a large spinal lipoma, because a terminal lipoma can tether the cord and, when the lipoma is large, can compress it [55]. Taken together,

these findings indicating that forces on the spinal cord are exacerbated by flexion may help explain the development of symptoms during or after repetitive activity involving flexion (eg, Spiller's adolescent patients who were rowing and cycling [15]).

Traction on the spinal cord can occur from a variety of directions depending on the underlying cause of the tethered cord. Thickened filum terminale and some lipomyelomeningoceles produce caudal traction, as can dorsal bands, meningocele manqué, dorsal lipomas, and dermal sinus tracts. Split cord malformations and neurenteric cysts may cause ventral traction [34]. The tethered cord syndrome can occur without any obvious lowering of the conus [4,6,8–11]. In support of tethered cord syndrome with a normally positioned conus, Selcuki and colleagues [9] have postulated that an increase in abnormal dense connective tissue around the ependymal canal of the conus can reduce the elasticity of the normal-lying conus and its ability to act as a buffer against tethering.

In aggregate, this evidence supports the premise that entities that produce excessive tethering or compression of the spinal cord can produce neurologic dysfunction, presumably through a combination of local ischemia and direct

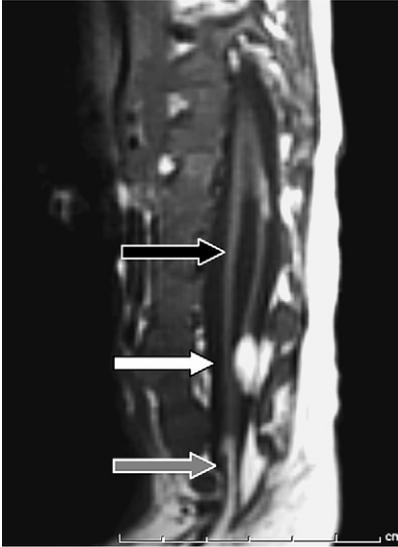


Fig. 2. MRI showing dorsal (white arrow) and terminal (gray arrow) lipomas with an associated syrinx (black arrow).

mechanical dysfunction. Limitation of injurious activity (such as excessive flexion) may minimize cord injury. Furthermore, relief of this tension and/or compression (as seen with surgical untethering) may result in improved circulation and abrogation of further damage.

Indications and rationale for treatment

Children who present with tethered cord syndrome fall into one of two groups, symptomatic or asymptomatic. In general, treatment of symptomatic children found to have tethered cord by imaging is indicated. It is important, however, to note two potential confounding conditions related to specific disease entities.

Confounding conditions

One group that merits special mention is the population of patients who have large spinal lipomas that exhibit mass effect on the spinal cord. A limited subpopulation of this group can develop worsening in symptoms in association with rapid weight gain. It has been hypothesized that the spinal lipoma increases in size, and thus in mass effect, as the patient gains weight. Some have proposed that weight loss may be a helpful adjunct in the treatment of this patient population. Although it may be difficult to justify

delaying treatment of patients who have neurologic deficits, those presenting only with pain may be considered for a trial of weight loss before committing to surgical intervention.

A second population that is important to discuss is the group of myelomeningocele patients who have symptoms suggestive of tethered cord syndrome and who also have ventricular shunts. A malfunctioning shunt sometimes can cause signs and symptoms that may mimic a tethered cord. In these patients, even with radiographic evidence of findings suggestive of a tethered cord (eg, progressive scoliosis or a syrinx), it is important to confirm that the shunt is working before committing to an untethering operation.

General principles

Outside these two specific scenarios, the development or progression of symptoms in patients who have a tethered cord often calls for an untethering operation [56]. Symptoms may develop early in life but can occur later, as exemplified by one case report of the development of symptomatic diastematomyelia in a 78-year-old woman [57]. In general, these more complicated cases should be referred to neurosurgeons who have extensive experience in the treatment of this condition and only after a discussion with the child's family and caregivers outlining the risks and benefits of the procedure.

Chronicling a decline in neurologic, orthopedic, or urologic status may be difficult across multiple health care providers. As discussed previously, neurologic symptoms include lower extremity weakness, gait impairment, and pain. A detailed neurologic examination can provide clues to the causes of sensory/motor disturbances and progression of lower back and leg pains. Obvious deficits on initial examination may be fixed, but progression of any of these symptoms as documented by serial neurologic examinations further supports the indication for surgery. Surgery can arrest the progression of symptoms in the majority of patients; a smaller percentage of patients show improvement after untethering [34]. Improvement is more likely to be seen in patients whose primary symptom is pain, although these patients tend to be an older population including young adults and adults [34].

Orthopedic symptoms may be present also and should be considered in deciding on indications for treatment. Patients sometimes are seen first by an orthopedic surgeon for progressive scoliosis,

gait problems, or for consideration of heel cord release because of toe walking. These symptoms in the setting of a tethered cord should prompt consideration of surgical release of the cord. It also is crucial to be aware of the population of children who have the concomitant presentation of severe scoliosis and a tethered cord. Untethering often arrests symptoms, but orthopedic intervention still may be required in cases of more severe scoliosis. If these children are scheduled for correction of the curvature of the spine, the cord should be untethered first to avoid excessive traction on the cord when subsequent bony realignment and lengthening occurs.

Pang's [40] study of split cord malformation in patients who had scoliosis reveals that in the majority of patients the scoliosis was stabilized after untethering, although subsequent correction of the deformity was limited. Similarly, Pierz and colleagues [58] studied 19 children who had myelomeningocele, scoliosis, and a tethered cord. They found that for curves of less than 40° there was subsequent improvement in the correction of the deformity, but there was no improvement for curves greater than 40°. These data suggest that the neurosurgeon should recommend untethering as treatment of the root cause of scoliosis in selected cases, but that correction of the deformity may be limited, and orthopedic involvement may be necessary.

Urologic symptoms can be stabilized or improved in many cases following successful untethering operations. Metcalfe and colleagues [59] demonstrated marked improvement of medically managed neuropathic bladders after sectioning of the filum terminale in 36 pediatric patients (age range, 1.2–15 years). Other studies also have shown marked improvement in urologic function after untethering, even in patients who have a normally positioned conus and a normal thickness filum [6,10,60–62]. When retethering occurs in patients who have myelomeningocele, Tarcán and colleagues [63] have shown that a second untethering surgery can improve urologic outcome markedly.

Controversy, however, still surrounds treatment options for the asymptomatic patient who has signs of a spinal anomaly, particularly a milder anomaly such as a thickened filum or an asymptomatic lipoma. (For a thorough review, see McLone and Thompson [33].) The risks must be weighed, because lipomas of the filum (or a thickened filum) have much better surgical outcomes than those of the conus, which can be

significantly more difficult to remove. Some patients can lead fully active lives with a fatty filum and remain symptom-free throughout their lifetimes. In this setting, surgery may not be necessary and may not justify the risk of complications such as cerebrospinal fluid (CSF) leak.

Conversely, some believe that evidence supports a role for prophylactic surgery, noting that surgery does not always provide a reversal of dysfunction or abnormality in symptomatic patients. Studies of patients who had delayed diagnosis and treatment of occult spinal dysraphisms reveal that they are more likely to present with irreversible urologic and neurologic deficits that might have been prevented with an earlier diagnosis and surgical treatment [64]. Thus, proponents of early surgery argue that there is value in attempted prevention of irreversible defects, particularly in avoiding urologic dysfunction. Improvements in surgical technique with reductions in perioperative complications have added to the enthusiasm for early treatment and treatment of asymptomatic patients. Amid this controversy, however, many agree about the need for well-designed, randomized, controlled trials involving patients who have asymptomatic closed spinal defects [5,56,65].

Treatment options

General principles

Latex precautions

Full latex allergy precautions should be considered. The incidence of latex allergy in children who have spinal dysraphism is high because of the likelihood of exposure to latex antigens from repeated bladder catheterization [66].

Intraoperative monitoring

The differentiation of nerve roots from other structures such as a lipoma, dorsal band, or filum terminale is a critical element of any detethering procedure and is facilitated by the use of intraoperative neurophysiologic monitoring with combinations of motor-evoked potentials and sensory-evoked potentials. Monopolar nerve stimulators can be used to stimulate nerves so that they can be identified and preserved. Stimulation of S2, S3, and S4 can be monitored through anal manometry or electromyographic recordings [67]. The external anal sphincter is innervated by the anterior roots of S2 and S3 and by both roots of S4 through the proximal branch of the pudendal

nerve. Because the distal branch of the pudendal nerve, the perineal nerve, supplies the external urethral sphincter, rectal manometry usually reflects activity of the external urethral sphincter as well [67]. The integrity of the conus medullaris and the cauda equina can be monitored by motor root mapping, motor-evoked potentials, sensory-evoked potentials, and electromyography.

Avoiding cerebrospinal fluid leak

Measures should be taken to decrease the risk of CSF leak. These measures include careful attention to a watertight dural closure, with graft if necessary. The integrity of the closure can be tested by a Valsalva maneuver under direct observation. Adjuvant sealants, such as fibrin glue or other commercially available products, may be used to enhance closure of the dura. In addition to the dural closure, careful attention also is paid to the superficial soft tissue closure as a further means of minimizing CSF leak. Consultation with plastic surgery colleagues may be helpful in complex cases to assist in possible planning of alternative closure strategies, such as rotational flaps. Postoperatively, the child often is kept flat for 1 to 5 days (depending on the complexity of the repair) and gradually is elevated in bed to minimize pressure from a standing column of fluid on the repair site.

Avoiding retethering

Many spinal anomalies with tethered cord have a tendency to retether postoperatively. To avoid retethering, meticulous attention is needed in hemostasis and closure. Dural closure with 4-0 Nurodon is adequate for many straightforward detethering operations. With more complex spinal dysraphisms, a running monofilament suture can be used with good results [68]. In complex lesions, such as extensive lipomas, resection of the maximal amount of pathologic tissue should be performed, followed by imbrication of the pial surface to create a smooth surface apposed to the dura [69,70].

Sometimes the dural sac is developmentally deficient or becomes compromised during the operation so that a significant portion of the dural sac must be reconstituted. Various materials have been used as grafts with varying degrees of success including autologous fascia, Gore-Tex (W. L. Gore & Associates, Baltimore, Maryland), biological collagen, and cadaveric tissue such as Alloderm (Lifecell, Branchburg, New Jersey) [71,72]. Silastic sheeting (Dow Corning, Midland,

Missouri) has been used in the past, but it can cause the formation of a fibrous envelope to which neural structures may attach [68]. Finally, in severe cases of retethering in patients who have myelomeningocele and compromised neural function, transaction of the spinal cord above the neural placode can be performed to prevent the placode from scarring and forming adhesions [73].

In the immediate postoperative period, some have reported keeping patients prone to minimize adhesions of the cord to the dural suture line. The patients are turned supine and slowly elevated in bed over several days.

General strategies

Many untethering operations are associated with markedly abnormal anatomy resulting from primary development and also secondary postoperative scarring. A general strategy of starting the dissection from normal tissue (usually rostrally) is often helpful. The finding of a normal bony lamina may allow identification of the dura and subsequent improved understanding of abnormal tissue planes. This principle holds true intradurally as well, where rostral exposure of normal spinal cord may facilitate safer dissection of more caudal abnormalities.

For many cases involving lipomatous tissue or scar, the use of the laser can greatly enhance the ease and safety of surgery. Excellent results have been reported with both the carbon dioxide laser and the yttrium-aluminum-garnet contact laser. Lipomatous masses in the conus and spinal cord can cause significant tethering and can be difficult to remove completely, but marked debulking often can be achieved, despite the occasional need to leave a rim of residual lipoma. At closure, a pial imbricating sutures to reconstitute more normal spinal anatomy may help reduce retethering [69].

Specific entities

Filum terminale

In filum terminale, only the distal filum terminale needs to be exposed. The filum is recognizable by its fatty appearance, by its straight midline location, and by its vasculature (Fig. 3). It is important to visualize the underside of the filum before sectioning, because nerve roots can travel along with the filum (Fig. 4). The intraoperative microscope can be invaluable in this exercise. Intraoperative nerve monitoring can be helpful in improving discrimination of nerve root from

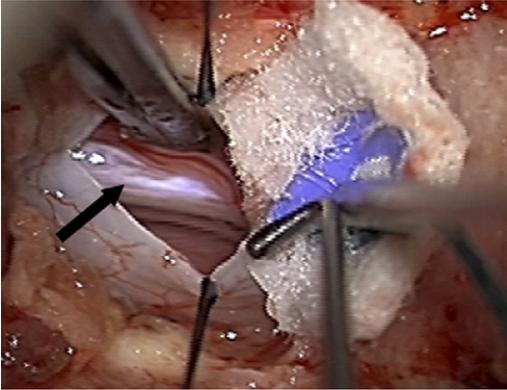


Fig. 3. A fatty filum (*arrow*).

filum. Once the filum is sectioned, care should be taken that there is no bleeding at the site of section before the proximal stump is released, because it may retract out of reach (Fig. 5). A watertight dural closure should be performed (Fig. 6).

Split cord malformations including meningocele manqué

Hemicords tether at the median septum, and therefore it is imperative to remove the septum. During the approach, care must be taken to avoid damaging to the spinal cord through inadvertent traction, because the cord often is tethered strongly to the bony septum or bony structures with dorsal bands. For a type I split cord malformation, the bony spur is removed subperiosteally from the dura and resected with either rongeurs or a drill. Then the dural sleeves of both hemicords are opened, and the median dura is resected along with the ventral dura with no ventral repair. A similar and easier approach to

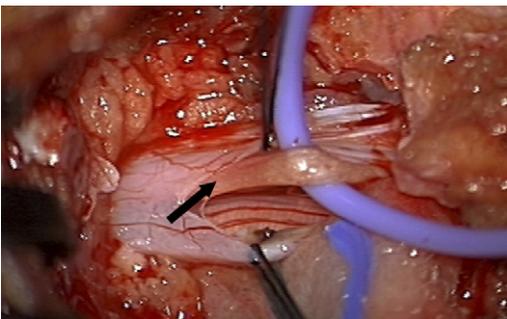


Fig. 4. Isolation of the fatty filum (*arrow*) with normal nerves below.

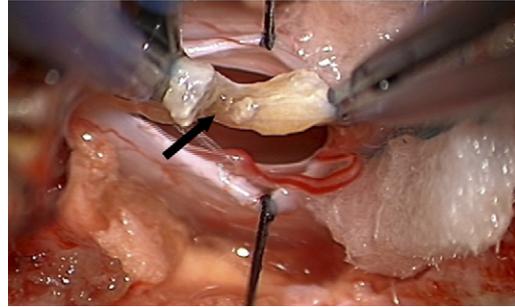


Fig. 5. Sectioning of the fatty filum (*arrow*). Note the fatty appearance and the absence of bleeding after coagulation.

the median fibrous band is applicable to type II split cord malformations.

Meningocele manqué should be treated similarly to a split cord malformation or a lipomyelomeningocele. The primary finding is dorsal bands tethering the cord. These bands can extend extradurally to attach to the laminae, and adherent nerve roots may be found also.

Other entities

Dermal sinus tracts identified radiographically as being in continuity with the central nervous system also may lead to tethering and should be explored intradurally. Simple excision of the extradural component of the tract may not alleviate intradural tethering. The child who presents with a patent sinus tract (as evidenced by obvious leaking of CSF or recurrent bouts of meningitis) should be treated in an expeditious fashion to minimize the risk of further infection. Preoperative imaging should be reviewed carefully for evidence of an intradural or intramedullary dermoid, which must be completely excised to avoid recurrence.

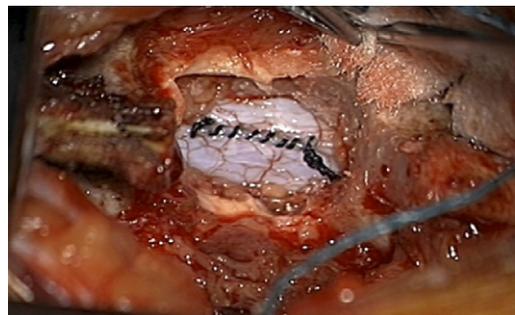


Fig. 6. Watertight dural closure using a running stitch.

Neurenteric cysts often are extremely adherent to the spinal cord and, because of the risk of recurrence, should be resected completely if possible. The surgeon must weigh the risks and benefits of total resection against the possibility of creating unacceptable neurologic deficits, paying particular attention to vital ventral spinal cord vasculature to avoid inadvertent cord ischemia. Finally, in some neurenteric cysts it may be useful to consider a ventral approach for improved exposure [42].

Surgical complications

Complications

Cerebrospinal fluid leak

CSF leak is a worrisome complication of surgeries in children who have a tethered cord because the dural anatomy may be abnormal before surgery and may be compromised further by the operative procedure. A meticulous, watertight dural closure is critical to avoid this potential complication, with dural substitutes and sealants used when necessary [68]. Several centers, including Children's Hospital of Boston, routinely keep patients who have complex untethering operations (but not an uncomplicated fatty filum) prone for several days following durotomy to facilitate dural apposition, with slow incremental elevation of the patient's head over subsequent days. Re-exploration may be warranted should CSF leak be observed, and the inclusion of plastic surgery staff for patients who have had extensive surgery or multiple operations may be helpful in planning alternative methods of closure, such as transposition flaps.

Retethering

Retethering of the cord is common, particularly in complex cases when not all of the adherent, tethering tissue can be removed, such as in a deep-seated transitional lipoma. At Children's Hospital of Boston children who have complex lesions may be kept prone to minimize adhesions to the dural suture line in the immediate postoperative period. In addition, this positioning helps minimize potential contamination of the wound by urine and feces.

Patients who have transitional lipomas have a significantly higher frequency of symptomatic retethering than patients who have either caudal or dorsal lipomas [36,74]. In a retrospective study by Colak and colleagues [74] with a median follow-up of 58 months after lipomyelomeningocele repair, 20.2% of patients had symptomatic retethering.

No dural graft material is completely free from the complication of retethering, including Gore-Tex, pericardial grafts, Silastic, and allograft dura.

In general, the diagnosis of retethering often is based on clinical examination and history. Any new or significantly progressive orthopedic, urologic, or neurologic symptom should be evaluated for the possibility of retethering. Unfortunately, although a large number of diagnostic tools are available to the clinician, few have reliably predicted retethering.

Worsening scoliosis may be part of the clinical picture of retethering and may be identified objectively by serial plain-film radiographs. Urodynamics has been shown to be a useful tool in the evaluation of the patient suspected of having symptomatic retethering. Creating a urodynamic score (based on bladder volume, compliance, detrusor activity, and vesico-sphincteric synergy) both preoperatively and postoperatively has been shown to be a reliable method for detecting retethering [75]. MRI has not proven particularly effective in evaluating retethering because the postoperative conus position often is similar to preoperative images, although evidence of an enlarging syrinx often is considered worrisome for retethering. Serial somatosensory-evoked potential testing for retethering has high false-positive (71%) and false-negative (43%) rates and has not proven to be particularly useful in supporting the clinical diagnosis of retethering [74,76].

Follow-up

Strategies for following patients who have been diagnosed as having a tethered cord often vary based on the underlying cause, treatment, and outcome. Patients who have been treated for a fatty filum with a straightforward sectioning operation and who are neurologically well may not need much in the way of long-term follow-up. In contrast, children who have more anatomically complex lesions, such as large lipomas or myelomeningocele, may need long-term, regular follow-up, involving multidisciplinary medical and social services, to monitor for potential retethering [66]. Interdisciplinary care, often including neurosurgery, orthopedics, urology, psychiatry, medicine, and social work, may be required to minimize potential further medical complications while maintaining the child's educational and social developmental trajectory as much as possible. In addition, some patients may suffer from complex pain syndromes, and consultation with a pain service team may be helpful.

From practical standpoint, the role of the neurosurgeon in the follow-up of patients who have complex tethered cords often involves annual neurologic examinations, periodic imaging studies (to evaluate for scoliosis, syrinx development or—in shunted patients—shunt failure), and review of periodic urodynamic studies.

Although retethering can occur at any time, the risk often decreases once adult stature is reached and growth has stopped. Nonetheless, in children who have complex tethering lesions delayed retethering can occur, sometimes decades later. A current problem faced by many pediatric neurosurgeons is managing the transition of patients treated initially as children into adulthood. Although debate regarding the best strategy to manage this transition is ongoing, it is accepted that a continued relationship with neurosurgery is mandatory for the treatment of this challenging population.

Summary

Tethered cord syndrome is a clinical phenomenon resulting from anatomic restriction of the normal movement of the spinal cord or vascular compromise leading to hypoxia of its distal structures. Causes of tethering can be acquired (secondary) or congenital (primary). A detailed understanding of the embryologic causes of primary tethered cord can aid in the diagnosis and treatment of patients who have these conditions. Surgical intervention, when indicated, is directed at releasing the tethered cord; intraoperative neurophysiologic monitoring in certain patients and meticulous dural closure whenever possible are important adjuncts to the operative procedure, regardless of the mechanism of cord restriction. Retethering of the released spinal cord may occur over time in certain subgroups of patients who should be regularly followed over time to monitor their neurologic, orthopedic, and urologic stability.

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