

# Tethered cord syndrome in children: a review

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✓The treatment of a patient with symptoms of a tethered spinal cord and in whom a fatty infiltrated terminal filum is found is controversial. The authors review their experience and the literature regarding this aspect of occult spinal dysraphism. From experience, transection of a fatty terminal filum in patients with symptoms related to excessive caudal cord tension is a minor procedure that generally yields good results. The most problematic issue in the literature is what patients and symptoms are best suited to surgical treatment. (DOI: 10.3171/FOC-07/08/E2)

**KEY WORDS** • fatty terminal filum • pediatric neurosurgery • tethered cord syndrome

**P**ROGRESSIVE neurological, urological, and orthopedic dysfunction due to congenital fixation or tethering of the distal spinal cord by the terminal filum is known as TCS. This member of the OSD family is often seen by pediatric and general neurosurgeons, and both the diagnosis and treatment are controversial and can be difficult. Drawing from a detailed literature review and our own experience with this entity at the Children's Hospital, Birmingham, Alabama, we review facets of childhood tethered spinal cord syndrome, discuss many of the clinically relevant aspects of this disease, and share our views on the management of this problem.

## History of the Tethered Spinal Cord

The tethered spinal cord is a concept that has been discussed for about 150 years. Johnson<sup>18</sup> described a "fatty tumor from the sacrum of a child connected with the spinal membranes" in 1857. In 1875 Vichow<sup>46</sup> introduced the term "spina bifida occulta." In 1891 in England, Jones<sup>20</sup> described the first successful surgical intervention for a tethered spinal cord in which the "spine [was] trephined to relieve pressure on the cauda equine." In 1953, Garceau<sup>9</sup> described the "filum terminale syndrome" in three patients with progressive orthopedic spinal deformity and neurological dysfunction. He attributed the problem to tension on the conus medullaris from a thickened terminal filum, which was found on surgical exploration. He noted

good recovery in all three patients after the filum was surgically sectioned. Three years later, Jones and Love<sup>21</sup> described their experience at the Mayo Clinic, reporting a good recovery of neurological and urological function after sectioning of the filum terminale. They also observed a widened space between the cut ends, affirming that the filum was under tension. In 1976, Hoffman et al.<sup>13</sup> coined the term "tethered spinal cord" in patients with a low-lying conus medullaris with a thickened filum ( $\geq 2$  mm in diameter). These authors specifically excluded lipomyelomeningoceles, meningoceles, SCMs, and most other "dysraphic" conditions from their patient cohort.<sup>13</sup> We recognize that the term "tethered cord" has broadened to include cord tethering from many different etiological conditions in the literature, but we confine our discussion of TCS to a tethered cord due to a tight and/or fatty infiltrated terminal filum, holding to the original definition set forth by Hoffman and colleagues.<sup>13</sup>

## Incidence and Epidemiology

The true incidence of OSD and primary TCS is not known. Unlike open neural tube defects, closed defects such as the TCS are usually diagnosed with the onset of symptoms or found incidentally during workup of unrelated problems. The incidence of open neural tube defects has declined dramatically since the introduction of folate supplementation. On the contrary, the incidence of OSD and TCS has risen steadily, probably due to higher rates of incidental detection with the more commonplace use of MR imaging, greater clinical awareness, and a continued broadening characterization of the disease process. The true incidence, however, is not known, and little is known of its true prevalence. The literature does support a 2:1

*Abbreviations used in this paper:* CCM = caudal cell mass; CT = computed tomography; MR = magnetic resonance; OCA = other congenital abnormality; OSD = occult spinal dysraphism; SCM = split cord malformation; TCS = tethered cord syndrome; VB = vertebral body.

female predominance for OSD,<sup>3</sup> but whether this holds true for TCS is uncertain.

There are limited data regarding risk factors for OSD, but there is evidence to suggest that both open and closed neural tube defects may be genetically related. Therefore, siblings of patients with known neural tube defect are at higher risk for OSD and TCS. There are also data to support the possibility that OSD and TCS are linked to other congenital abnormalities such as cutaneous stigmata,<sup>50</sup> orthopedic/vertebral abnormalities, and anorectal/urogenital malformations.<sup>5,51</sup>

### Embryology and Development of TCS

To better understand developmental anomalies like TCS, it is important to review the normal development of the spinal cord. The development of the embryonic spinal cord begins around Day 18 postconception. The three major steps in spinal cord development are neurulation, canalization of the tail bud ("secondary neurulation"), and regression of the caudal cell mass. The cephalic portion of the spinal cord (cervical, thoracic, and upper lumbar spinal segments) is formed during neurulation, whereas its caudal portion (including the terminal filum) is formed via canalization and regression.<sup>8,16,31,48</sup>

Neural tube formation is the essential process of neurulation. During this process, the neural ectoderm along the primitive streak is induced to proliferate by the underlying notochord. Because of differential growth, the edges begin to fold inward toward one another, forming the neural groove. As the folding edges of the neural ectoderm join, they are covered by adjacent cutaneous ectoderm. Disjunction of the two ectoderm types occurs as the edges meet. With the closure of the neural tube around L-1 or L-2, neurulation is completed. Developmental errors during neurulation can lead to the formation of a myelomeningocele, meningocele, intraspinal lipoma, lipomyelomeningocele, dermal sinus tract, or SCM.<sup>8,16,48</sup>

The neural ectoderm cell mass caudal to the neural tube is called the CCM or tail bud. Canalization of the CCM begins around Day 28 with formation of vacuoles within the middle of the CCM. The vacuoles begin to coalesce, forming a central canal within the CCM. This canal eventually connects with the central canal of the cephalic neural tube formed during neurulation. The distal lumbar, sacral, and coccygeal segments are now formed. The terminal filum is formed through regression of the caudal portion of the CCM. The ventriculus terminalis is a central canal dilation within the distal CCM (the site of the future conus medullaris). Distal to this, at the tip of the coccyx, lies an epidermal cell rest (coccygeal medullary vestige). As regression occurs (between the ventriculus terminalis and coccygeal medullary vestige) and differential growth of the vertebral canal and the neural tube progresses, the terminal filum is formed as the spinal cord "ascends" or pulls away from its distal sacral attachments.<sup>8,16,48</sup> A tight or fatty filum is thought to result from errors that occur during the canalization and regression stages.

The "normal" position of the conus medullaris ranges from the midlevel of T-12 to the lower portion of L-3, but T-12 and L-3 represent the extremes of a bell-shaped curve distribution, with the majority of normal conus rest-

ing between L-1 and the L1-2 interspace. The authors of cadaveric studies of normal adults have shown that only 1.5% of patients have the conus as low as the L-3 VB. In MR imaging studies several authors have also confirmed these data.<sup>35,52</sup>

In 1970, Barson<sup>2</sup> performed postmortem examinations of 252 neurologically "normal" infants and children. Analysis of these data suggested that the conus of term infants lie at L2-3 but continue to ascend to the average adult level of L1-2 by 3 months of age. This is controversial because other cadaveric, MR imaging, and ultrasonography studies seem to suggest that the conus ascends to the L1-2 disc space earlier in life—by the 40th postmenstrual week.<sup>10,52,53</sup>

### Pathophysiology of the Tethered Cord

The proposed normal functions of the terminal filum are to fixate, stabilize, and buffer the distal cord from normal and abnormal cephalic and caudal traction. The filum is a viscoelastic band that usually allows the conus medullaris to move slightly during flexion and extension of the spine. It is believed that, if this viscoelasticity of the filum is lost or compromised by either fatty infiltration<sup>29</sup> or abnormal thickening, then caudal tension and traction may cause undue stress upon the conus, resulting in TCS. It is believed that this abnormal inelastic filum interferes with normal cord ascension and results in a low-lying conus medullaris (that is, a conus below the L1-2 interspace). This is classically believed to be the hallmark of the TCS, but we have published data that demonstrate that TCS can exist when the conus is positioned normally.<sup>44,49,50,54</sup>

Yamada et al.<sup>52</sup> defined TCS as "a stretch-induced functional disorder of the lumbosacral spinal cord due to excessive tension" between the lowest pair of dentate ligaments and the caudal end of the spinal cord anchored to an inelastic terminal filum. In an anatomical study of the dentate ligament, however, we observed no evidence that this structure significantly interfered with either cranial or caudal traction on the spinal cord.<sup>45</sup> Nevertheless, Yamada et al. showed that caudal traction on the distal cord resulted in impairment of oxidative metabolism and that the degree of impairment correlated with the severity of the neurological deficits. Using animal models, they also showed that the dysfunction seen in TCS involves the gray matter of the distal cord (below L-1) and occurs because of derangement of oxidative metabolism. This metabolic dysfunction is at the mitochondria level with a significant reduction shift of cytochrome *a* and *a*<sub>3</sub> (similar to what is seen in hypoxia and ischemia brain/spinal cord models). This reduction shift is proportional to the amount of caudal traction applied to the distal cord. These authors also demonstrated a corresponding reduction in spinal cord blood flow in proportion to the traction force. They then postulated that such cord traction caused "traction induced hypoxia" and neuronal membrane stretch with "loss of transmembrane ion homeostasis and electrical activity depression."<sup>40,54-56</sup>

### Diagnosis: Clinical Signs/Symptoms and Neuroimaging Findings

Patients with symptomatic TCS can present with a wide

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TABLE 1  
Common presenting symptoms in patients with OSD and TCS

cutaneous manifestations
hypertrichosis
cutaneous capillary hemangioma
dermal sinus tract
midline subcutaneous lipoma
lumbosacral appendage
“cigarette burns”
orthopedic abnormalities
leg-length discrepancy
foot asymmetry
foot deformity (pes cavus, claw toes)
progressive scoliosis
urological problems
neurogenic bladder
incontinence
frequent urinary tract infections
neurological signs & symptoms
upper motor neuron signs
lower motor neuron signs
often mixed upper/lower neuron signs
asymmetrical weakness (decreased leg movement in infants)
gait difficulty or delayed ambulation
spasticity
sensory deficits (feet or perineum)
back/leg pain (young adults)

variety of signs and symptoms in combination with cutaneous, orthopedic, spinal, anorectal, and urological abnormalities, as well as pain (Table 1). In our experience, the common clinical presentations (Fig. 1 *left*) include the presence of cutaneous signatures associated with OSD (59%), neurogenic bladder with the development of primary or secondary incontinence or urinary tract infection (18%), leg or foot weakness, numbness and/or spasticity (12%), leg or foot length discrepancy (6%), foot deformity (for example, pes cavus, claw toes), spinal deformities, and nondermatomal back and leg pain (6%).<sup>32</sup> Although pain is a major presenting symptom in the adults with TCS, it is less common and more difficult to identify in the pediatric population because pain often manifests simply as irritability, especially in younger children.

Cutaneous signatures associated with OSD include lumbosacral hypertrichosis (Fig. 2), cutaneous capillary hemangiomas (nevus) (Fig. 3), dermal sinus tracts (Fig. 4), midline subcutaneous lipomas, lumbosacral skin appendages (Fig. 5), and so-called cigarette burns or atretic meningocele (Fig. 6). In our experience, cutaneous signatures were seen in 59% of patients with TCS, and the literature suggests that cutaneous anomalies are present in as many as 70% of patients with OSD. Only about 3% of healthy neonates will have such lesions.<sup>34</sup> Patients with TCS often exhibit multiple skin lesions when examined carefully.

Urological abnormalities range from obvious incontinence to subtle, subclinical findings seen on urodynamic testing. In the pediatric population, urological abnormalities usually do not become obvious until the child grows out of his/her infant years. They also tend to be more subtle than other clinical findings.<sup>15</sup> The urological presentation can include incontinence, urgency, increased/abnormal frequency, and recurrent urinary tract infections. Urodynamic testing usually confirms a neurogenic bladder in the symptomatic child and can often verify subtle neurogenic bladder dysfunction in young infants suspected of having TCS without any obvious urological dysfunction. We routinely obtain urodynamics studies in all patients with TCS. We find this a very valuable diagnostic tool that can detect subtle or subclinical dysfunction and allows more objective assessment of operative and follow-up risk. Although the test is invasive, young infants generally tolerate it well.

The neurological dysfunction in TCS is unusual, frequently having elements of both upper and lower motor dysfunction. Motor weakness is more prevalent than sensory deficits. Such motor dysfunction is usually asymmetrical. Children can present with delayed gait development, spasticity, hyperreflexia, hyporeflexia, and muscular atrophy. In infants, muscular atrophy can be hidden by subcutaneous fat. Sensory deficits, when present, are usually in the feet or perineum. Children can sometimes present with painless ulcerations of the foot or leg.<sup>30</sup> In our series of 73 patients with TCS, changes in strength, tone, or reflexes were seen in 47 patients (64%).<sup>50</sup>

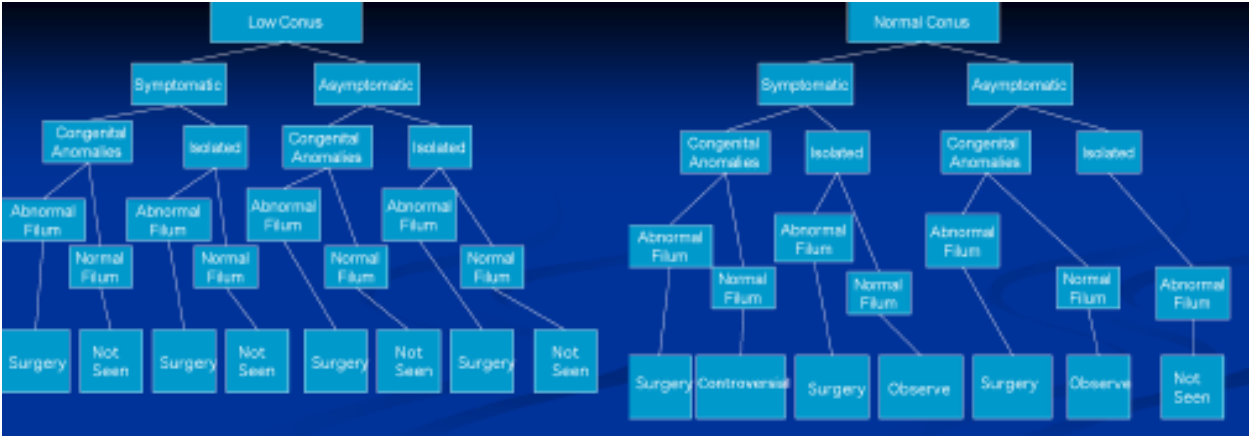


FIG. 1. Flow charts. *Left and Right:* treatment paradigms for patients with a suspected tethered cord.



FIG. 2. Photograph demonstrating hypertrichosis in a 14-year-old girl presenting with leg-length discrepancy.

Orthopedic manifestations include foot deformities (most common), limb-length discrepancies, leg malformations, gluteal asymmetry, and vertebral abnormalities (for example, laminar defects, bifid vertebrae, hemivertebrae, SCM, sacral agenesis, segmentation errors, and scoliosis).<sup>48</sup> Orthopedic deformities of one form or another are found in more than 90% of patients with TCS,<sup>43,48,50</sup> and scoliosis is seen in up to 25%.<sup>15</sup>

It is now well recognized that TCS is often seen with other congenital syndromes. The two most common associations are caudal agenesis (a spectrum of caudal regression abnormalities)<sup>7,42</sup> and anorectal atresia syndromes (OEIS [omphalocele, exstrophy, imperforate anus, spinal defect] syndrome, VATER [vertebrae, anus, trachea, esophagus, and renal] syndrome, and Currarino triad).<sup>4,5,8,24,47</sup> Patients with these syndromes should be screened for OSD and TCS.

#### Neuroimaging Modalities

Neuroimaging is used to confirm the clinical suspicion of OSD and/or TCS (Table 2).

**Ultrasonography.** Ultrasonography can be a useful tool in young infants. The advantages are the ability to obtain a dynamic view without having to submit a young child to irradiation or sedation. The disadvantages are that images can be difficult to interpret and the quality is often operator dependent. Identifying the level of the conus medullaris is not difficult in the very young child, but searching for fat or the thickness of the terminal filum can be challenging. The acoustic window in the lumbar spine is usually lost by 4 to 5 months of age.

**Plain Radiography.** Plain x-ray films of the spine help identify occult spinal abnormalities that may aid in the diagnosis of OCAs. The modality can be a useful tool in observing an associated scoliosis over time. Many of the associated vertebral abnormalities can be seen on routine spine radiographs (Fig. 1, right). This modality is also often used as a preoperative tool in planning surgery.

**Magnetic Resonance Imaging.** Magnetic resonance im-

TABLE 2  
Common neuroimaging abnormalities in OSD and TCS

Abnormality
laminar defects and malformations
fused lamina
VB narrowing
butterfly vertebrae
hemivertebra
midline vertebral cleft
midline osseous spur
pedicle flattening
widened interpedicular distance
widened spinal canal
posterior VB scalloping
missing or reduced VB
scoliotic deformity
lordotic deformity
sacral agenesis/dysgenesis

aging is the modality of choice in visualizing the level of the conus medullaris and for identifying a thickened and/or fatty filum. Sagittal T1- and T2-weighted images are best for localizing the level of the conus, whereas T1-weighted axial MR images are better for identifying fat within the terminal filum and for measuring the diameter of the filum (Fig. 7). We routinely image the entire spinal axis to search for concomitant lesions. Dynamic MR imaging may have some use in understanding pathological conditions of the distal spinal cord,<sup>19,28</sup> although we have found it to be of limited value. This modality can be difficult to undertake in young children, who may require sedation or general anesthesia to obtain quality images.

**Computed Tomography or CT Myelography.** Computed tomography scanning or CT myelography can be used if MR imaging is not available. These modalities permit excellent visualization of the osseous anatomy, and the position of the conus can also be clearly seen. However, CT myelography is invasive and can be difficult to perform in young infants.

**Urodynamic Testing.** Routine urodynamic tests are performed in our patients suspected of having TCS. In the pediatric patient, being able to determine bladder dysfunction simply based on history or symptoms can be problematic. Palmer and colleagues<sup>33</sup> have used preoperative



FIG. 3. Photograph of a neonate with a flat capillary hemangioma of the midline lumbar spine.

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FIG. 4. Photograph showing a dermal sinus tract in a neonate with underlying tethered spinal cord.



FIG. 5. Photograph demonstrating a lumbosacral skin appendage in a neonate with imaging findings of a tethered spinal cord.

urodynamic tests to unmask subclinical bladder dysfunction in children presenting with nonurological symptoms indicative of TCS. They reported that such urological dysfunction improved postoperatively in 75% of their patients. Our experience supports this view, and we have found urodynamics to be a very good objective data point to follow pre- and postoperatively.

### Surgical Decision Making

The fundamental goals of surgical intervention in TCS are as follows: 1) to improve or stabilize deficits in the symptomatic patient and 2) to prevent future deficits in the asymptomatic patient. These two goals are predicated on the fact that sectioning of the terminal filum can be conducted safely with minimal risk and a very low rate of morbidity. This relative safety has been studied extensively and is commonly held to be true. The reported complications of surgery are cerebrospinal fluid leakage (most common), wound infection, meningitis, bladder dysfunction, and neurological injury. The incidence of neurological injury due to sectioning is less than 1%.<sup>1,17,22,23,32,39</sup> The general consensus from the literature and from our professional experience is that surgical filum sectioning can be completed with little morbidity, and the results, as discussed later, can be quite good.

Although surgical intervention has been established as safe, surgical decision making for patients with TCS can be quite complex. Furthermore, surgery in these patients can be controversial because of the broad spectrum of pathological involvement.<sup>25,32</sup> Figure 1 provides a summary of our institution's experience and our general approach to surgical decision making in the pediatric TCS population.

There is a clear consensus in the neurosurgical community that symptomatic patients with a low-lying conus medullaris (one positioned below the L1–2 interspace) and a fatty filum should be surgically treated<sup>26</sup> whether they

have concomitant OCAs or not. From this end of the spectrum on, the data become less clear and more controversial. We will discuss our institution's approach to and experience within this wide spectrum. Based on our work on determination of TCS in patients with a normally positioned conus, symptomatic patients with a normally positioned conus and an abnormal or fatty filum (Fig. 7) are offered surgery. We have found the results in this group to be comparable to those in the symptomatic, low-lying conus group. We believe that surgery is indicated for the asymptomatic patients with a low-lying conus and a fatty filum (with or without OCAs) to prevent future deterioration. A less common scenario for which we would occasionally offer surgery would be the asymptomatic patient with a normally positioned conus in the setting of multiple OCAs and a fatty filum. We recommend not being dogmatic about this patient group and making the decision to perform surgery on a case-by-case basis because the data we have gathered on the natural history of the disorder are not encouraging. We typically will observe symptomatic patients with normally positioned conus, no OCA, and normal filum and search aggressively for other causes for the symptoms. Observation is also advocated for asymptomatic patients with a normally positioned conus and normal filum but who have multiple OCAs. We have not seen and therefore do not have any experience to bring to bear on the following groups: 1) symptomatic cases involving a low-lying conus and with or without OCAs but normal filum; 2) asymptomatic cases involving a low-lying conus and with or without OCAs but normal filum; and 3) asymptomatic cases involving a normally positioned conus without OCA but with an abnormal or fatty filum. The one area of growing controversy is in the group of patients with symptoms but a normally positioned conus, without OCAs, and with a normal filum, known as "occult" TCS. More and more surgeons are beginning to undertake surgery in this group, but there are published data supporting both operative and nonoperative strategies.<sup>6,36</sup> Many of the gray areas in this decision tree cannot be clarified until we are able to obtain a better understanding of the natural history of the many variations in this disease process.

Our philosophy for surgical intervention is based on three important observations: the major cause of neurolo-





FIG. 6. Photograph of an atretic meningocele cigarette burn-type lesion over the lumbosacral spine of a newborn child. The conus medullaris in this patient was inferiorly displaced.

gical deterioration in these patients is due to abnormal fixation and traction on the conus and distal cord; the neurological deterioration is progressive if left untreated; and once deficits develop, they are difficult to reverse.<sup>16</sup> Offering intervention in adolescent patients with normal imaging findings, back pain, and urological symptoms not supported by abnormal results on urodynamic tests is problematic in our view.

#### Surgical Planning, Techniques, and Postoperative Care

Once TCS has been identified and the decision for surgical intervention has been made, we electively schedule the patient for surgery within a few weeks. In young children with multiple congenital abnormalities, we allow surgical repairs of more serious defects to be conducted before proceeding with the untethering, unless neurological deficits are present. In patients with scoliosis and TCS, we prefer to perform the untethering at the time of the scoliosis repair. This saves the child from two sessions of general anesthesia. The untethering is done before the fusion, and an effort is made to keep the two incisions separate, which is not usually a problem because very few of these patients require corrections involving the sacrum.

Preoperatively, we obtain routine laboratory testing and anesthesia evaluation. The majority of our patients will also undergo preoperative plain radiography, MR imaging, and urodynamic testing. Although not routinely used at our institution, intraoperative neuroelectrophysiological monitoring can be used to help distinguish functioning neural elements from the terminal filum, if confusion exists intraoperatively. At our institution, with inferior transection of the filum performed by our senior surgeon, we do not believe that such monitoring contributes to the safety of this relatively benign procedure.

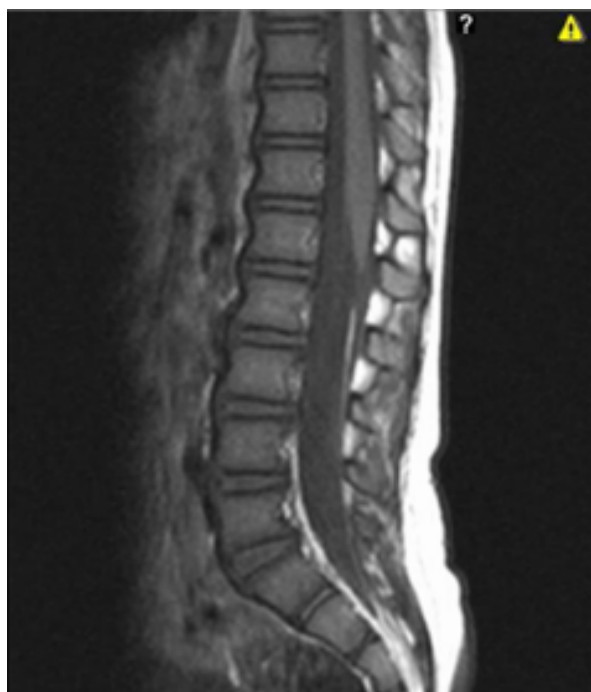


FIG. 7. Sagittal (*left*) and axial (*right*) T1-weighted MR images obtained in a 7-year-old child who presented with leg pain and urinary symptoms. Imaging demonstrated a normally positioned spinal cord with fatty terminal filum.

After induction of general anesthesia, we routinely administer a broad-spectrum prophylactic antibiotic agent, which is continued for 24 hours postoperatively. We try to avoid using any paralytic agents for induction of anesthesia. If necessary, only very short-acting agents are used. We do not give preoperative steroids. The patient is positioned prone on two bolsters placed under the chest and iliac crests to prevent abdominal compression and minimize epidural venous engorgement. The L5–S1 space is localized with either manual palpation of external landmarks or with fluoroscopy. Although the filum can often be isolated at higher levels, we believe that the lumbosacral junction is optimal because the filum typically fuses with the dura mater at the S-2 level.<sup>12</sup> This permits the filum to be sectioned distal to the conus and thus minimizes the injury of any low-lying sacral cell bodies. A small midline incision is made and dissection is extended down in a standard fashion to expose the inferior aspect of L-5, part or all of S-1, and the L5–S1 interlaminar space. From this exposure, one can either 1) perform an S-1 laminectomy, 2) perform a partial inferior L-5 laminectomy and superior S-1 laminectomy, or 3) place an interlaminar spreader to obtain enough exposure to be able to safely open and close the dura. With good magnification and illumination in place, a midline dorsal durotomy is made. Usually a 2-cm opening should be adequate. The dural edges are held up in tension with retention stitches. After opening the arachnoid, we advocate clipping it to the dural edge (the clips should be removed before closure). The

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FIG. 8. Intraoperative image revealing a fatty impregnated terminal filum (coursing over the suture).

filum can be identified by its typical dorsal midline location, its slightly bluish discoloration, its anteriorly located vessels, and the fat that often infiltrates it (Fig. 8). If the filum is not found dorsally, one may need to look laterally and/or rostrally; we reported a cadaveric study in which we found that approximately 10% of the terminal fila fused off the midline, and 15% may fuse above S-1.<sup>12</sup> Care must be taken in bluntly dissecting all nerve roots away from the filum and visualizing the undersurface of the filum free of nerve roots before sectioning it. Distal sacral roots, particularly the S-5 nerve root, can be much smaller than the lumbar or proximal sacral nerve roots, and inadvertent sectioning of these may result in a significant fixed deficit. If neuromonitoring is used, then the filum should be stimulated before cutting. When used, monitoring must include that of appropriate sacral function. A suture is placed through the filum to aid in retaining a piece for pathological examination. The filum is then coagulated and sectioned, first proximally, then distally. A specimen is sent for histological evaluation. To better interpret the pathology report, one should recognize that there are normally scattered neural elements with the filum.<sup>32,41</sup> The cut ends typically retract, and the ends rarely retether.<sup>37</sup> With a good dural closure, we typically do not use any additional dural sealant. A layered closure is done with the skin typically closed with an absorbable stitch. Some institutions have used skin sealants like Dermabond instead of a skin stitch with good success.

Postoperatively, patients are kept flat with bedrest for 48 to 72 hours before gradually raising the head of the bed and increasing the activity level. Pseudomeningoceles are rare but do occur. If persistent cerebrospinal fluid leakage does occur, reexploration can be conducted with minimal added morbidity. Patients are routinely discharged from the hospital the following day. After discharge, the patient is seen a week later to assess the wound. In the young patient with scoliosis who requires a body cast after the combined untethering and fusion, a good wound evaluation can be a challenge. If a wound “window” cannot be made on the inferior dorsal aspect of the cast, then the cast change should be done within 2 weeks to allow adequate wound follow-up. Afterward, patients undergo follow-up at 3, 6, and 12 months and then yearly. In symptomatic

patients with preoperative orthopedic, urological, or neurological problems, the extent of improvement 6 months postoperatively is all the improvement that can be anticipated. Coordinated follow-up evaluation with the orthopedic and urology services should be done to maximize patient follow-up compliance.

### Results of Untethering Surgery

The results of surgical untethering in symptomatic TCS patients are generally favorable, but the extent of improvement varies depending on the preoperative symptoms and deficits. In 1975, Anderson<sup>1</sup> reported in his series of 73 pediatric patients with OSD and TCS that the rate of improved pain was 100%, whereas the rates of improved sensorimotor and sphincter function were 42 and 43%, respectively, and those of symptom stabilization were 45 and 48%, respectively. In their patients, Lee et al.<sup>27</sup> reported that pain improved in approximately 80% of patients, neurological improvement or stabilization occurred in 90% (with quicker recovery of motor rather than sensory function), and bladder function improved in 50%. Guerra et al.<sup>11</sup> reported similar results, with 48% improvement seen in pediatric patients with abnormal urodynamics. Other studies have yielded higher improvement rates for urological dysfunction, with an average of 87% seen in seven studies having a total of 161 patients.<sup>36</sup> It must be noted that urological improvement is not as favorable in older children and in the adult population.<sup>26</sup> Huttman et al.<sup>14</sup> compared the durations of symptoms with outcomes and concluded that pain and spasticity responded favorably to surgery regardless of the duration of symptoms, whereas improvements in sensorimotor and bladder function were inversely related to the duration of symptoms. It must also be noted that even though many authors have noted significant improvement in bladder function, complete urological recovery is rare.

### Conclusions

The challenge that faces neurosurgeons does not lie in the technical aspects of sectioning the terminal filum but in correctly identifying which patients have TCS, which patients are at risk for TCS and future neurological deterioration, and which of these patients would benefit from surgical intervention. If the correct diagnosis is made and timely treatment implemented, one is rewarded with a neurologically normal child with a potential for a healthy life. We hope that the present review and the experience of our institution shed more light on this topic, but the reader must be aware that there is still much controversy and uncertainty regarding this topic. A recent questionnaire-based survey of the practice pattern of 105 neurosurgeons showed that there is still much uncertainty and disagreement in the neurosurgical community with regard to this complex disease process.<sup>38</sup> Even though there has been significant insight into this disease gained over the last 150 years, there is still a considerable lack of Class I data. Therefore, there is a need to continue critically looking at this disease process to obtain better data through randomized prospective studies. The relative safety of the procedure should not be a reason to arbitrarily advise surgery

in patients with normal studies and symptoms of uncertain origin.

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