Surgery for tethered cord syndrome: when and how?

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Abstract

Tethered cord syndrome (TCS) is a clinical condition presented with neurological and/or urological signs and findings. Increased hypoxic stress in the spinal cord secondary to traction or stretching is the pathophysiological mechanism of TCS. It is usually observed in childhood but adult cases were also reported. Its diagnosis and treatment are always challenging. Magnetic resonance imaging, electrophysiological and urodynamic studies are the main diagnostic tools for TCS. This syndrome may be primary or secondary to previous surgeries such as myelomeningocele. Asymptomatic patients with low conus medullaris and thick filum terminale are always questionable for surgical treatment. On the other hand, symptomatic patients with normal radiological imaging are critical for surgical decision. Surgical treatment involves correction of the spinal pathologies and release of the spinal cord by cutting the filum terminale. Time of surgery and technical nuances are still in debate among neurosurgeons. Neurological and urological outcomes of the patients usually depend on these factors. Timing of surgery and surgical technique need to be clarified based on the recent clinical studies. This review will focus on the time and technique of TCS surgery. Firstly, a brief description of TCS will be provided, then an extensive view on the surgical treatment of TCS will be performed.

Keywords: Tethered Cord Syndrome; Surgery; Timing; Technique; Filum Terminale.

The term “tethered cord syndrome” (TCS) represents both an individual diagnosis and a combination of neurological and urological signs and symptoms associated with various forms of spinal dysraphism. It is usually diagnosed in childhood. The natural history of spinal cord tethering remains unclear, but not all children experience clinical deterioration in this syndrome. In TCS, progressive neurological deterioration in the functions of the lower spinal cord results from traction on the conus medullaris (1). Current understanding of TCS began with the understanding and management of spina bifida; this later led to the gradual recognition of spina bifida occulta and the symptoms associated with tethering of the filum terminale. Occult tethered cord is first specifically described by Khoury et al. in a group of children who had a conus medullaris at normal level, but presented with incontinence. After the intradural section of filum terminale there was an improvement in 70% of cases (2). Publications by Harold Hoffman, Bruce Hendrick, and Robin Humphreys represent some of the most important modern series on TCS. They introduced the term “tethered spinal cord” to define a radiographic diagnosis of thickened filum terminale measuring 2 mm or more in diameter and a low-positioned conus medullaris (3). Yamada et al. (4) expanded the definition of TCS by linking it to symptoms of an impaired oxidative metabolism in the spinal cord. In experimental and clinical studies using dual wavelength reflection spectrophotometry, they demonstrated electrophysiological and metabolic deficiency in experimental model and human TCS (4). Today, the unifying concept of the conditions included within TCS is the pathophysiology of increased tension and aberrant stretching of the spinal cord. In this review, we will give brief information about primary and secondary TCS. Then surgical treatment of TCS will be explained in detail with the latest reports on this interesting disorder.

Primary (congenital) TCS

TCS may be primary or secondary (5,6). When the neural tube is closed, closure begins in the cranial side and proceeds towards the caudal side. Factors affecting the early stage are cranial, those affecting the later stage lead to neurilization defect on the caudal side and open spinal dysraphism (meningocele, myelomeningocele) (7). Programmed cell death in the tail bud causes the
formation of a filum terminale in the future. During this period, the neural tissue is completely covered with skin. In TCS, the problem usually arises at this stage, and a closed spinal dysraphism is developed. At the level of the conus medullaris, there is an ongoing change in intrauterine life and postpartum period. The conus medullaris, which usually extends to the sacrum during the intrauterine period, rises to L1-L2 level in adolescent period (7). The disruption in these embryological developmental stages causes primary (congenital) TCS (7,8). TCS may be associated with Chiari type I malformation and low lying cerebellar tonsils as described in Milhorat’s work in 2009 (9). In the cases associated with Chiari malformation, different surgical strategies have been described for surgical treatment of both pathologies and even Gluncic et al. proposed contemporary surgical approach. But he is the only author who adopts this strategy (10).

Secondary TCS

Causes of secondary TCS are those that increase the tension of the postpartum spinal cord. Acquired (infections, postoperative scars, tumors...) or developmental defects (split cord malformations, dermal sinus tracts), that cause adhesions or adhesion between the spinal cord and the surrounding tissues in a child with normal spinal cord during the postpartum period, may cause secondary TCS (7,8,11,12). The following structures are the responsible of this condition; fibrous bands, thick filum terminale, fatty filum terminale, intradural lipoma, lipomyelomeningocele, adhesions secondary to myelomeningocele repair, bony septum within the distal spinal cord, and tumors (dermoid, epidermoid tumors) (13,14) (Figure 1).

![Figure 1](image1.png)

Figure 1. Magnetic resonance imaging of a patient with tethered cord syndrome secondary to type 1 split cord malformation. (A) Sagittal T1 MRI shows fatty filum (FF), (B) Axial T2 MRI shows the bony septum (BS) and 2 hemicords, (C) Sagittal T2 MRI shows bony septum (BS) emerging from the upper end-plate of L3 vertebra.

Symptoms and Physical Findings

Symptoms of TCS occur predominantly between 5 and 15 years of age (15). The most common symptom is motor weakness/loss on the extremities. Typically, weakness is progressive, and atrophy and thinning of the legs are observed on the same side. Loss or increase in deep tendon reflexes and positive Babinski’s sign can also be detected. Weakness on the extremities is often asymmetrical in children. TCS may also cause neurological deficits in adulthood after a normal childhood period (15). In adult TCS, weakness is typically symmetric and presents with multiple segment involvement (16). In adult TCS, unlike childhood age, the most common symptom is severe back and/or leg pain. Urological disorders are similar to those in childhood (15). Unlike adult TCS, there is usually no pain in childhood, including foot deformities and spinal deformities (17).

Clinical findings of TCS vary with age, but can be examined under four main headings. These include cutaneous findings, other co-morbid anomalies (orthopedic, urological, and, gastrointestinal), lower motor neuron findings associated with congenital spinal and nerve root abnormalities, upper motor neuron findings due to stretching of the spinal cord (18). Therefore, TCS may be suspected in children with subcutaneous lipomas, hypertrichosis, neurogenic bladder and deformity in feet.

Skin Findings

83% of spinal dysraphism cases have a sign on their skin. About 70% of TCS patients have a skin lesion. These include subcutaneous lipomas, hypertrichosis, telangiectasia, hemangioma, pigmentation, and atrophic skin (Figure 2). In the site and sacral area, midline, dimple or a small hole should be examined carefully before the radiological evaluation. Symptoms such as scoliosis, skin redness, dimples, holes, and stimuli such as back pain and bladder dysfunction should be considered in differential diagnosis (7).

![Figure 2](image2.png)

Figure 2. (A) Skin dimple and red colored skin are obvious in a child with TCS. (B) Lumbar hypertrichosis (Faun tail) is seen in an adult patient with TCS.

Skin findings, often in the lumbosacral region, can be seen along the entire spinal axis (19). Cutaneous signs such as atypical dimples, capillary hemangiomas, subcutaneous lipomas, hypertrichosis and tail (dermal appendices) are common findings (11,12). Faun tail and silky down are the types of hypertrichosis which are mainly observed in split cord malformations (19). It is important that they are on the back center line. The ones that are up to the top of the gluteal radius are even more important. It is stated that those who are below the gluteal radius are more related to the pilonidal sinus.

Orthopaedic Findings

Orthopaedic deformities have a significant place among the features of TCS (6). The neuromusculoskeletal syndrome used to describe the coexistence of neurological and orthopedic disorders is a common clinical picture in
functions are normal, these patients can be followed-up TCS (20,21,22,23). These studies are the most valuable urodynamic studies (cystometrogram) are of paramount patients with urodynamic studies (7,8,20). type malfunction (5). So, it is necessary to investigate all not imply that the patient does not have neurogenic bladder sensory disturbance and normal anal sphincter tonus do not confirm the absence of an underlying neurogenic cause (7). This may further increase neurological deterioration of the patient. Scoliosis can be seen especially in 90% of split cord malformation (Type 1) patients. The overall rate of scoliosis is around 25%. Other orthopedic deformities seen in TCS patients are; pit foot, turn of feet inside, etc. Pathological findings frequently seen in radiological studies are butterfly vertebra, block vertebra, and dysmorphic vertebra bodies (6,7,8).

Urological findings
In TCS, urological problems usually occur with pathologies that manifest in three parts of sympathetic, parasympathetic, and somatic pathways. Neurogenic bladder, incontinence, recurrent urinary tract infections are frequently observed (20,21). Among the urological problems, urinary incontinence has a very important place, and recurrent urinary tract infections are also important features to watch out for (8,18). Naturally, the age of the patient is very important for determining incontinence (4). It is not difficult to determine incontinence in patients over the age of the toilet habit should be settled. However, in younger children, the presence of sphincter dysfunction, urinary retention in the bladder after ultrasonography, frequent presence of infection in the urine microscope are the signs of bladder and sphincter function problems (5,6,20). Vesicoureteral reflux, enlarged ureter, enlarged bladder, post-void residual urine are problems in these patients when investigated by urodynamics studies (14,22). At least 30% of patients with TCS, sphincter functions are normal. It is important to note that the absence of perianal sensory disturbance and normal anal sphincter tonus do not imply that the patient does not have neurogenic bladder type malfunction (5). So, it is necessary to investigate all patients with urodynamic studies (7,8,20).

Urodynamic studies (cystometrogram) are of paramount importance in diagnosis and follow-up of patients with TCS (20,21,22,23). These studies are the most valuable method about bladder dysfunction. If the urinary system functions are normal, these patients can be followed-up by these studies. Therefore, urodynamic tests should be repeated annually. Cystometrograms to be performed preoperatively and postoperatively are recommended to make diagnosis and follow-up of patients more objectively (20). In older patients where this examination can be performed, the presence of a hypertrophic, hyperreflexible bladder is a more valuable finding for spinal cord tension in TCS (21). Hyperreflex contractions (especially in the filling phase) are also valuable findings indicating that hypertonicity is not due to bladder muscle fibrosis. Hypertonicity in the bladder is another valuable finding indicating a decrease in capacity (20).

Radiology
Magnetic resonance imaging (MRI) is the gold standard imaging method for diagnosis (6). But, the diagnosis of occult TCS is often difficult. For the diagnosis of this occult syndrome, it is important to mention about the work of the Japanese colleagues with lumbar MRI in the prone position (24). In this method, lumbar MRI in prone position shows that the filum terminale is located significantly posterior and the cauda equina is located anterior in patients with occult TCS. This suggested a difference in elasticity between the filum terminale and cauda equina (24). Conus medullaris terminating below the L3 level in MRI and presence of 2 mm thick filum terminale are common radiological findings of TCS. In some cases, it should not be forgotten that the patient may be TCS even if the conus medullaris is at normal level and the filum terminale is at normal thickness and patients with skin signs, bladder dysfunction, neurogenic symptoms, or scoliosis should be evaluated for TCS (4,7). On T1-weighted images, normal anatomy, location of the conus, presence of lipoma in the spinal cord, filum or canal and filum diameter can be clearly observed. Fatty filum terminale is a common radiological finding in patients with the symptoms of TCS and can be easily detected by T1-weighted MRI scan (6). T2-weighted sections are useful for showing and differentiation of tumors such as dermoids or epidermoids. These tumors are located in or around the conus medullaris and cause traction of spinal cord. MRI is also useful on the determination of attachments or fibrous bands which are secondary to previous intradural surgeries or arachnoiditis (8). Computed tomography (CT) is the radiological tool for the detection of osseous lesions and malformations (Figure 3). Type 1 split cord malformation, bone hypertrophies, butterfly vertebræ, posterior fusion anomalies, spinal calcifications can be detected by CT scans. CT-myelography can be performed to plan the surgical strategy when MRI can not be obtained due to some technical problems (11). Plain x-rays can be used for scoliosis. Ultrasonography is used during the pregnancy and in newborns to diagnose spinal malformations, dermal sinus tracts that may cause TCS (12,13).
Figure 3. Axial computed tomography shows the bony septum, which is splitting the spinal cord.

Electrophysiological Studies

Electrophysiological studies are not only important tools for the diagnosis of TCS but also a valuable parameter for the follow-up of patients who underwent surgery for TCS (6). An adjunct to clarify the findings of TCS is SSEP (somatosensory evoked potential). If the conduction velocity slows down in the SSEP or if a conduction block is detected, this finding can be interpreted in favor of spinal cord tension. As is known, SSEP is transmitted to the brain via the posterior spinal cord (fasciculus gracilis and cuneatus), which is generated by the electrical stimulation of the brain, what should be known at this point is that the cerebral cortex has a corrective effect, and if it does not have a very serious conduction time span or conduction block, it makes the SSEP waves appeared normal in time and appearance. For this reason, the warning records should be made on the sciatic nerve, lumbar, thoracic, cervical, and finally in the center to determine whether there is not a peripheral problem. The result we have seen in this test is that normal waves are obtained in brain recordings, even if the block is in the lumbar or thoracic region. Delays in transmission can be better noticed by alerting from different locations (8). SSEP and MEP (motor evoked potential) can be used intraoperatively for a safe and effective surgery in TCS. Changes in SSEP and MEP records of patients during surgery may alert surgeon for a possible neural damage. Moreover, continuous free-running electromyography is useful for monitoring the rootlets of the cauda equina. Electrophysiological stimulation of the rootlets is used for the differentiation of filum terminale before cutting this fibrous band for the release of the spinal cord.

Surgical Treatment

The natural history of occult spinal dysraphism remains largely unknown. Making a recommendation for prophylactic release of the spinal cord in asymptomatic patients is questionable. But in symptomatic patients, it is inevitable to perform surgery. Meanwhile most of the authors suggest the prophylactic release of the spinal cord as soon as possible in cases of secondary TCS due to split cord malformation even if the patient is asymptomatic. The main goal of treatment in TCS is to remove the pathology leading to stretching the spinal cord by blocking its movement in the cranial direction. The basic principle in surgical technique is to remove all the connections that stretch the spine or the height of the child without damaging the spinal cord and roots (6,7,8,25).

The time and technique of surgery are mostly depended on the pathophysiology of the syndrome. The degree of traction on the conus medullaris is postulated to determine the age of onset of symptoms, with lesser degrees of tethering remaining subclinical. Timing is very important for TCS surgery. The time of the appearance of first motor neuron signs may be the time of surgery (26). In addition, recognition of detrusor hyperactivity and detrusor-external sphincter dyssynergia that may cause vesicoureteral reflux and hydronephrosis is another indication of surgery for TCS (6,20,21). It seems that early surgical repair may reduce the risk of neurological deterioration of the lower urinary tract, and allows a more physiological development of urinary function (27). It should also not be forgotten that deteriorated neurological/urological functions could not be reversed if the surgical treatment is delayed (5). Of course, surgical treatment depends on the nature of tethering pathology (6).

The classical surgical technique is the open surgical release of the spinal cord using microsurgical techniques (6,8,11,12). While the patient is in the prone position, laminectomy is performed with an incision that fits the lesion and the filum is then reached and cut (28,29,30). During this approach, not only the filum terminale but also adhesions, fibrous bands and lesions that caused stretching of the spinal cord are also removed (6,11). After the identification of filum terminale, it should be absolutely cauterized before cutting because it has a feeding artery and this maneuver may prevent postoperative hemorrhage within the dural sac and consequent neurological complications (6). During the cut of filum terminale, a small sample of this structure may be obtained for histological examination (Figure 4). This may provide information about the tethering pathology of the patient, such as fatty filum, filar lipoma or ependymoma.
Intraoperative neuromonitoring (IONM)

IONM techniques are useful to prevent additional neurological deficits after surgery (31,32,39,40,41). During the surgery, the filum terminale can be identified by the presence of characteristic non-uniform vessels on its surface. But this is not always reliable technique to find the filum because some thick rootlets that are forming the cauda equina may also have vessels on their surfaces (11). These rootlets may mimic the filum terminale. Therefore, the best method to identify the filament is direct stimulation of the rootlets and the filament by IONM techniques (11,12). IONM, one of the methods developed to protect nerve tissue in surgical procedures, has been widely used in spinal surgery (40). Spinal tumors, spinal stenosis, trauma and congenital spinal malformations are the most common spinal diseases that are treated using IONM (40). Although the IONM basically records SSEP and MEP, it is suggested that sphincter MEP, free-running EMG, and bulbocavernous potentials are also recorded (Figure 5).

IONM is frequently used in meningocele, myeloschisis, myelomeningocele, lipomyelomeningocele, split cord malformations and TCS surgery (13). There is a significant contribution to the understanding of the functionalities of the roots emerging from the primitive neural placod (13), especially in the neural tube closure defects, and of the evoked potentials in the full recognition of the filum terminale in the TCS. In addition, free-run EMG follow-up and periodic MEP control during dislocation of bone or fibrous septum in split cord malformation are important for preserving neural structures (hemicords and roots) and neurological functions (40).

Electrodes are attached to the median nerve and ulnar nerve for upper extremity monitoring, and it is attached to the tibialis posterior and peroneal region of the lower extremity. MEP is the potency measuring motor function. It is an effective method to evaluate the structural and functional integrity. It should be preferred because it allows rapid response to surgery (40,42,43). Abdurator polices brevis for upper extremity records, tibialis anterior and abductor hallucis for lower extremity records, and external sphincter for sphincter tonus, electrode should be inserted. On the other hand, when the MEP and SSEP records are evaluated together during the IONM procedure, the specificity of the procedure is 92% and sensitivity is 99% (42,43).

It was thought that is to say the effect of anesthetic agents and deepening of muscle relaxants in the case of bipolar cautery used during surgery when that there was no change in IONM values despite neurological damage, to be the reason of the false negative result in the literature (38,44). Beyazova et al. (43), 10 patients with TCS were selected using the IONM and no change in MEP and SSEP values was found according to the results. It has been emphasized that the stimulation obtained by touching the probe with direct nerve tissue is effective in recognizing and protecting the normal nerve tissue during the surgical procedure.
Intraoperative neuromonitoring of a child who underwent surgical treatment for TCS. MEP values are effective in providing immediate information, especially with direct probe, facilitating the operation of nerve stimulation potentials. The evaluation of SSEP and MEP values together increases the precision and authenticity of IONM (40). SSEP values are also high sensitive for early detection of neurological deterioration. At the same time, it is also recommended to use sphincter MEP, free-run EMG values and potential values obtained by direct nerve stimulation to minimize the possibility of neurological damage during surgery (42). Bipolar or unipolar stimulating probes can be used during the surgery. However, due to the incomplete myelinations of child patients and the difficulties of placing electrodes, the IONM may not always give accurate results in children. Therefore, the stimulation treshold of filum terminale and rootlets should be higher in children than the adults to receive better responses from the sphincters and lower extremities.

Postoperative Results
Patients are usually benefiting from the surgery after relieving tension by cutting the filum terminale. Pain complaints are the most beneficial symptom of surgical intervention, and approximately 90% of the patients recover from pain after surgery (28). A complaint of incontinence can be defined as a bladder dysfunction. If the patient is admitted with this incontinence, the surgical benefit is more limited (21). Only those who applied with a thick filum terminale will be able to recover about 70% of incontinence after surgery. In the event of another development defect (such as myelomeningocele, lipomyelomeningocele) that accompanies the thick filum, the improvement of incontinence is reduced by up to 25% (20). If a preventive surgical procedure is applied without incontinence, the problem does not progress as naturally tethering is relieved in such cases (5). Selçuki et al. in their study, 38 (95%) patients improved and 2 (%5) remained the same within the group of 40 patients with back-leg pain. Of the 16 patients with urological complaints, 10 (62.5%) had improved, 5 (31%) were unchanged (30).

Postoperative Complications
Following TCS surgery, re-tethering may occur in the spinal cord in some of the cases because of arachnoid adhesions and dural bands between the internal surface of the dura mater and conus medullaris (30). The main complications after surgery are CSF fistula and wound infections (1,6,13,30,31). Surgical intervention is intended to prevent further progression of neurological function losses through recycling. The possibility of early postoperative complications should not be forgotten. These complications can be classified in two groups. The first group included superficial complications such as inadequate wound healing, infection, CSF fistula, and subcutaneous CSF collection (6,30). The rate of these complications varies between 10-25% (6). In this group of complications, patients are followed for a while by wound dressing and antibiotherapy. If wound healing is not achieved during taping, wound revision is needed. Or, before that, lumbar drainage is installed from the upper level. However, this causes prolonged use of antibiotics and prolonged stay in hospital (8). The second group is the emergence of new neurological deficits or the worsening of existing deficits. This group is a complication that may lead to more serious consequences. Although there is a 5% possibility of developing permanent neurological deficit in the postoperative period in TCS surgery (17), this rate increases to 10% if transient deficits are added.
The risk of symptomatic re-tethering after intradural filum terminale cutting as a result of a scar formation in large pediatric populations varies from a minimum of 2.7% to a maximum of 8.6% (45,46,47,48,49,50). The risk of re-tethering after a simple intradural filum terminale section is lower than that after primary repair of more complex spinal dysraphism. The percentage of symptomatic re-tethering increases significantly, from 15% to 45%, in patients undergoing myelomeningocele or lipomyelomeningocele repair (51,52).

Extradural section of the filum terminale, proposed by Veronesi et al.(33), has no neurological complications, CSF-related complications, or re-tethering, which may be seen after intradural section of the filum terminale.

TCS is an important clinical condition in childhood and adulthood. The possibility of TCS must be kept in mind especially in case of normal MRI studies in a patient with neurological/urological symptoms, low back pain and/or sciatalgia. Even in the presence of degenerative disease findings on radiological studies, the MRI should be carefully evaluated for fatty or thick filum terminale, other tethering lesions for the possible association of TCS, as a tethered spinal cord is more frequent than expected.

In order to exclude or diagnose the TCS, additional investigations such as SSEP and/or urodynamic studies should be done. In the direction of established studies and clinical experience associated TCS, it can be said that the surgical treatment is the most appropriate treatment method either without deficits or with the slightest. But timing of surgery and surgical method are more important than the diagnosis of TCS and decision of surgery.

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