

# Tethered cord syndrome: 5 year clinical experiences and surgical results

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## Abstract

**Aim:** Occult spinal dysraphism, which manifests with motor and sensory disturbances in children and causes orthopedic deformities in the lower extremities and urological findings, is the result of developmental defects in the caudal part of the neural tube. One of the most common spinal dysraphism is tethered cord syndrome (TCS). In this study, patients who had been diagnosed with TCS in the past 5 years were retrospectively reviewed and clinical, radiological and surgical characteristics of these patients were tried to be revealed.

**Material and Methods:** We retrospectively reviewed the data about 26 patients with the diagnosis of TCS in our clinic between 2011-2015. The results of these surgeries were evaluated by age, gender, additional malformations, clinic symptoms, radiological and electrophysiological evaluation. The patient with urological symptoms was additionally performed in urodynamic tests.

**Results:** These 26 cases, 23% were children and 77% were adults. 81% of the patients were male and 19% female. Only 3.8% of the patients were primary TCS, 96.2% of the patients had other malformations. Urine incontinence was found in 19.2% of the patients and orthopedic deformities were found in 7.7% of the patients. Surgical treatment was done to 65% of the patients, and 35% of the patients were followed because they didn't want to be operated.

**Conclusion:** Tethered cord syndrome is usually not seem alone, it is associated with other developmental defects of the spine and spinal cord. If surgery done earlier, the greater the likelihood of regression of the neurological deficit or stablization the deficits.

**Keywords:** Tethered Cord Syndrome; Spinal Dysraphism; Surgery.

## INTRODUCTION

Many studies have shown that on the third month of ontogenesis, the spinal cord is located along the entire length of spinal canal. However, the spine develops faster than the spinal cord in the later period, at the midpoint of the L3 vertebra of the spinal canal at the time of birth. On 12-18th months it ends at the lower edge of the L1 vertebra or at the level of the upper edge of L2 vertebra. (1-3).

Developmental defects of the spinal cord lead to thickening of the filum terminale and sclerosis. The filum terminale fixes the final part of the spinal cord firmly, resulting in stretching of the spinal cord and leading to neurological or urological symptoms (4). This clinical condition is called "Tethered Cord Syndrome (TCS)". This condition may depend on whether the filum terminale alone tightening the spinal cord, or it may be seen with split cord malformation, lipoma, spina bifida, meningocele, myelomeningocele and other malformations of the spinal cord (1,5-9). Therefore,

these developmental defects are thought to play a role in the pathogenesis of this syndrome.

The incidence of TCS is 5-25/100,000 in the United States. TCS was seen in women twice more than men. Although it can be seen in children and adults, TCS is more frequently diagnosed in childhood. In children, this syndrome usually presents with skin findings such as hypertrichosis or dermal sinus tracts (8,10,11).

Therefore, the incidence of TCS and other malformations of the spinal cord is considered to be higher in children than the adults (8). The most common symptom of TCS is weakness (75-90%) in the lower limbs. Orthopedic findings of the lower limb (ankle deformity, neurogenic claudication and shortening of the limbs - 63%). Skin findings (hypertrichosis, lipoma, hemangiomas, dermal sinus - 54%), urological problems (40%), scoliosis and kyphosis (29%) are other symptoms (1,3,5,9,12,13). In the radiological examinations of these patients, spina bifida was found in the lumbar or sacral region in 98% of

**Received:** 08.10.2017 **Accepted:** 28.12.2017

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the cases. The most frequently used radiological studies are magnetic resonance imaging (MRI) and computed tomography (CT). In addition to radiological examinations, electrophysiological studies have been used both before and after surgery, as well as during the surgery (3,6,8,14).

In this study, the clinical, radiological and surgical characteristics of patients who were diagnosed TCS and treated in our clinic over the last 5 years were evaluated retrospectively. The results are discussed in the light of current literature.

## MATERIAL and METHODS

The data of 26 patients who had the diagnosis of TCS and who were hospitalized in our clinic between 2011 and 2015 were retrospectively evaluated. Spina bifida patients without any neurological or urological complaints not underwent surgical treatment and they were excluded from the study. Following the physical examination and radiological studies, the patients who were admitted and followed-up for TCS were included in this study. All the patients in our series were recommended surgical treatment for TCS, and surgical treatment was performed in patients who accepted the surgery. Others were followed-up with routine neurological and radiological examinations. Informed consent forms were signed by the patients who had undergone surgical treatment before the surgery. In case of pediatric patients, these forms were signed by their first degree relatives (mother and father).

Intraoperative neurophysiological monitoring has been performed in all cases.

As a surgical technique, in the primary TCS, release of the spinal cord was performed by cutting the fibrous bands and filum terminale and if there was a secondary TCS or accompanying malformation, surgical treatment was firstly performed for this malformation, following filum terminale was cut in the same session. All adhesions and fibrous bands around the rootlets and the surrounding neural tissues had been removed. In the study, statistical analysis of patients' age, gender, complaints, clinical findings, somatosensory evoked potentials (SEP) was performed. Results of MRI and CT scans were also evaluated.

## RESULTS

First, the overall results of the entire series were given, followed by clinical results of children and then adult patients. Among 26 patients, 23% were children and 77% were adults. In totally, 81% of these patients were male and 19% were females.

Only 3.8% of the patients had the diagnosis of primary TCS without any accompanying malformation. In 96.2% of patients, TCS was accompanied with others spinal cord or spinal column malformations.

Among these malformations, spina bifida was in 96.1%, diastometamyelia was in 50% (Figure 1), fusion of the thoracic and lumbar vertebrae was in 34.5%, meningocele

or myelomeningocele was in 30.8%, syringomyelia in 26.9% of the cases. Remaining 15.4% of patients (3 cases) had dermoid tumor, sacrum and coccygeal agenesis, Tarlov's cyst and hemivertebrae. (Figures 2, 3).



Figure 1. T2 sagittal image of split cord malformation

The main complaint of the 73.1% of patients was back or leg pain, followed by lumbar hypertrichosis in 30.8% of the patients, swelling in the lumbar region in 26.9% of the patients, urinary incontinence in 19.2% of the patients. Moreover, 11.5% of patients had paraparesis, 7.7% had orthopedic deformities of the lower limbs, and 3.8% had impotence.

The preoperative SEPs were prolonged in 34.6% of the patients, but in remaining 65.4% of patients, SEP values were in normal range. 65% of the patients were operated on, and 35% were only followed because of their refusal of surgery. Postoperatively 47% of the patients had back pain, in 17.8% had cerebrospinal fluid (CSF) fistula, in 5.9% CSF collection, numbness of the leg, impotence or urinary incontinence. But subsequently these complications were resolved. Only in 17.6% of the patients were asymptomatic after surgery.

Pre-existing foot deformities, urinary incontinence and impotence remained unchanged. Half of the pediatric patients were under 1 year of age and the other half was over 1 year old. 83.3% of the children were girls and 16.7% were boys.

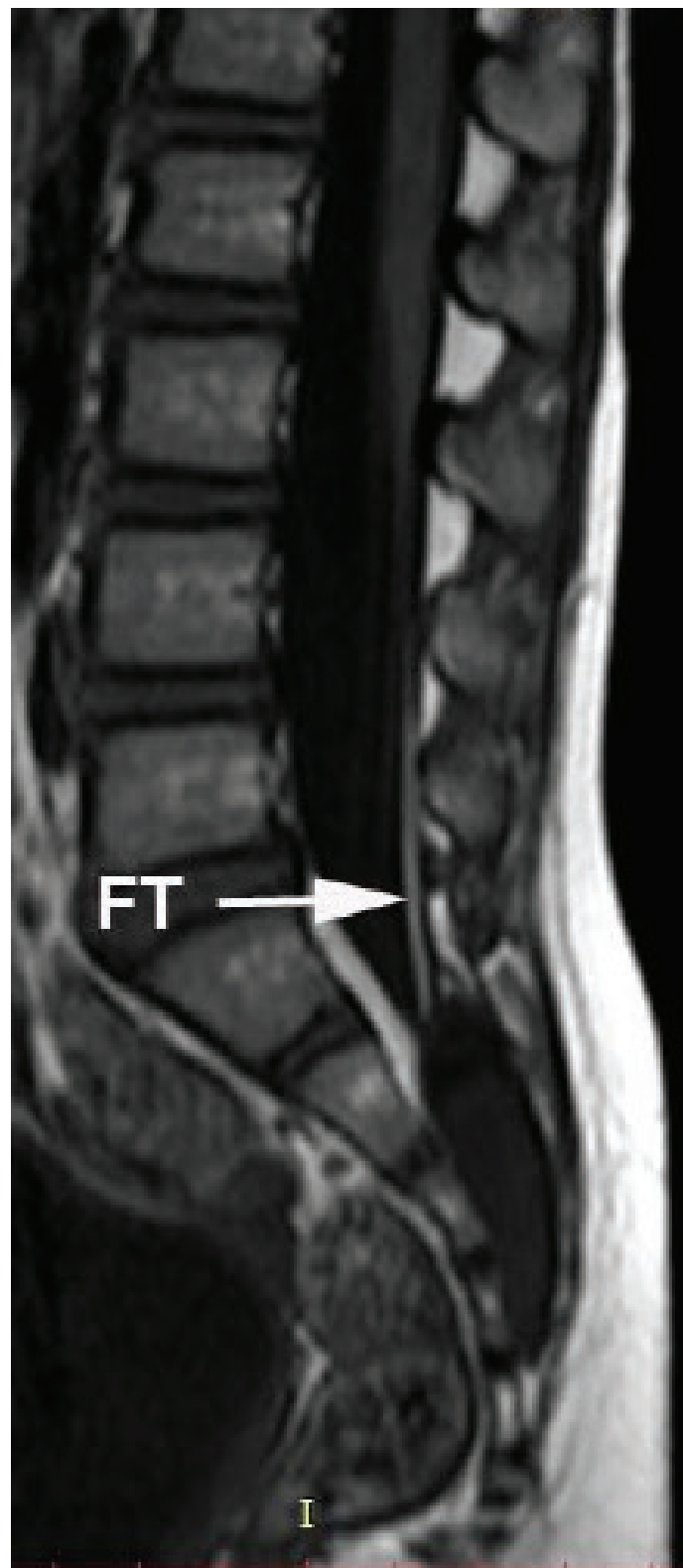
In all pediatric patients was detected an additional developmental and 83.3% of these cases consisted of diastematomyelia and spina bifida, 50% of these cases consisted of thoracolumbar vertebra fusions and dermal sinus tracts and 16.7% of these cases consisted of others congenital malformations (sacrum and coccyx agenesis, Tarlov's cyst, hemivertebra, syringo- and hydromyelia).



**Figure 2.** Sagittal MR imaging of a patient with tethered cord syndrome accompanied by dermoid tumor and intramedullary lipoma.

Among clinical signs, rotoscoliosis and orthopedic deformity in right foot were found in 33% of children, urinary incontinence, paraparesis, lumbar and leg pain, swelling and hypertrichosis in the lumbar region in 16.7% of children.

The preoperative SEPs were prolonged in 50% of the patients, but in remaining in 50%, SEPs were in normal range. All children underwent surgical treatment. 16.7% of children had CSF collection in postoperative period.



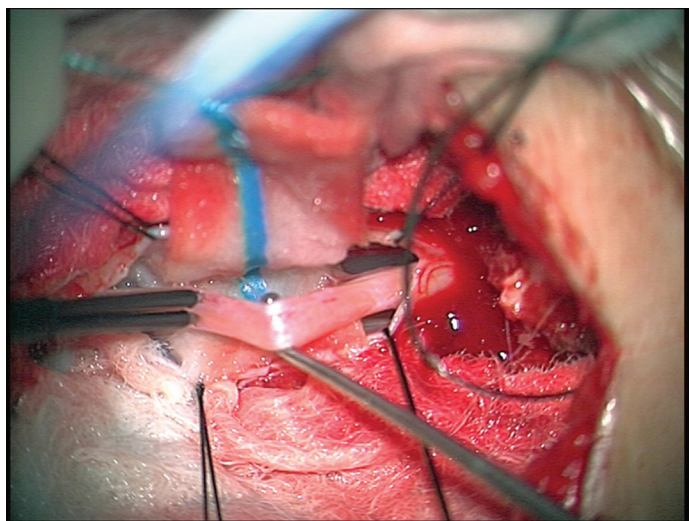
**Figure 3.** T1 sagittal MR image of a patient with tethered cord syndrome attached to fatty filum terminale. (FT: Filum terminale)



All adults (100%) were male. Sixty five percent of them was between 18 and 20 years old and 35% of them was over 20 year-old. In this group, others developmental defects

In addition, 33% of them had back pain. Only 33% of children were neurologically intact, 16.7% of them with pre-existing orthopedic deformities and paraparesis which were not resolved after surgery.

were also found during radiological studies. Among them, diastematomyelia was in 40% of cases, meningocele or myelomeningocele was in 40% of cases, dermal sinus tract was in 35%, syringomyelia and thoracolumbar vertebra fusions was in 30%, lipoma in 15%, scoliosis in 10%, and other malformations were found in 5% (hemivertebra and dermoid tumor) of cases. The main complaints in this group were leg pain in 90%, back pain in 30%, swelling in lumbar region in 25%, urinary incontinence in 25%, impotence in 5% and gait disturbance in 5% of cases. The preoperative SEPs were prolonged in 30% of the patients, but they were in normal range in 70% of cases. 55% of the adults were operated on, and 45% were only followed because they did not accept surgical treatment. In postoperative period, 30% of patients had lumbar pain, 10% of them had CSF fistula, 5% of them had numbness in foot, in 5% of the patients pre-existing urinary incontinence and impotence did not resolved after surgery. Intraoperative stimulation was used in all cases as a component of intraoperative neuromonitoring (Figure 4).



**Figure 4.** Intraoperative stimulation of fatty filum terminale with bipolar stimulator.

## DISCUSSION

In this article, our results of surgical treatment of TCS are presented. The pathophysiology, clinical and radiological findings related to TCS and spinal malformations accompanying this syndrome are also discussed. In the pathophysiology of the syndrome, detailed information was given about the development of malformations as a result of the neural tube closing at the time of neurulation. In our clinical experience, urinary incontinence, impotence, motor and sensory disturbances, orthopedic deformities were firstly seen in these patients. However, these findings

are not solely attributable to TCS, but also to premorbid dysplastic disorders of the spinal cord.

TCS is usually the result of a process beginning in embryonic life. As known, on the 17th day of gestation (triploblastic period) the embryo is a disc formation. On the 19th day, the notochordial plaque begins to develop from the remaining cells of the dorsal side of the notochord. This plaque thickens and matures to form notochord (5, 9, 15). The layer formed by thickening these new neuroectodermal cells is called neural plaque. The development of neural tube from neural plaque begins in the cervical region and continues in the caudal direction. Interestingly, the caudal parts of the spinal cord, conus medullaris and filum terminale develop from different structures in the terminal. This event allows the discoordination of the caudal part of the spinal cord to be isolated from other regions of the cord, and thus the developmental defects of the cord are detected in this region (almost all). So, TCS may develop secondary to the discoordination of the caudal part of the spinal cord. Disturbances in the separation of the maternal neural tissue from the ectoderm may cause congenital dermal sinuses, hypertrichosis and hyperpigmentation of the lumbar region, disconnection disorders of the neural tube (discrete spinal cord, spina bifida, meningocele and myelomeningocele), as well as other consequences such as syringomyelia and hydromyelia. The open neural plate in the myelocele is named "neural placode" (15). Neural roots begin to develop from the ventral surface of the placodes and extend in the ventral direction. The presence of ectodermal elements in the spinal canal leads to lipoma, dermoid, and epidermoid tumors (16).

Discussions on the triggering factors in the pathogenesis of these anomalies are still underway and risk factors include the use of ischemia, folic acid deficiency, hypoglycemia, hyperthermia and antiepileptic drugs (valproate) (4,15). The other cause of the syndrome is not congenital, but usually the postoperative scar tissue, adhesions, and reactive tissues fix the spinal cord after spinal surgery, usually performed in childhood or early childhood (6,13,16,17). This is also called the "secondary TCS". Tensioning of the spinal cord in primary TCS leads to organic deformation of neuronal membranes and to the release and not to renew adenosine triphosphate (ATP) stores. In our series, most of the patients had the diagnosis of secondary TCS at admission and only 3.8% of the patients had primary TCS.

Circulatory impairment in the caudal parts of the spinal cord, ischemia and metabolic disorders result in deterioration of integration and functions of the spinal cord (4,8,9). This leads to loss of progressive motor and sensory functions in the patient, pain and musculoskeletal deformities. Deterioration of blood flow in the affected areas has been proven by the laser doppler examinations. In addition, neurological findings that may be the result of stretching can be seen not only in tension, but also in myelodysplasia and masses that cause the spinal cord to develop. Stretching of the spine causes neurological,

urological and orthopedic findings in patients. However, these symptoms may occur in the early days of life or may remain hidden until adulthood. Pain is the most pronounced symptom in adults (3,4,8,9,10) (Table I), but in children it presents with skin findings (hypertrichosis, hemangiomas, nevus and swelling caused by meningocele or subcutaneous lipoma). In our series, as most of the patients had secondary TCS, symptoms related to the stretching of the spinal cord were mostly seen in adults. So, our results are compatible with the current literature.

**Table I. The table comparing clinical findings in children and adults is presented**

Findings	Children	Adult
Motor weakness	Often; delayed walking, neurogenic claudication	Commonly; in lower extremity
Motor weakness	Often; delayed walking, neurogenic claudication	Commonly; in lower extremity
Pain	Rarely, in back and lower extremity, not in perianal region and perineum	Often, most prominent symptom, severe, most frequently in perianal region and perineum
Ankle deformity	Often, early and progressively	Not
Skin findings	Incidence %80-100; hipertrichosis, angioma, nevus	Incidence <%50
Urinary incontinence	Often; mostly urinary incontinence, delaying in bladder control training, enuresis, recurrent urinary tract infections	Often, frequent urination, Feeling of incomplete bladder emptying, Urinary incontinence after bladder filling
Progressive deformities of spinal column	Often, commonly progressive	Rarely (<%5)
Trophic changes	Often, commonly in lower extremity	Rarely

In general, this syndrome progresses progressively (70-75%), which means that patients complain progress slowly over time and neurological and urological deficits progress gradually. (8,9). It presents with permanent back and leg pain, but these pains are often interpreted as myofascial or juvenile spondylopathy until the necessary radiological examinations are made. To have such as local or radiculopathic pains indicates that, problem may be in the lumbosacral part of the spinal cord and to have the anal or urinary sphincters failure indicates that, problems are related to the conus medullaris and cauda equina parts of spinal cord (1). Urinary incontinence refers to the neurogenic dysfunction of the bladder sphincter (5). Children with this complaint are usually examined in pediatrics, urology and nephrology clinics, treated, and followed up for a long time. Children who do not benefit from these treatments are referred to neurology or neurosurgery specialists (8). In our series, all patients were referred to our clinic from pediatrics or neurology

clinics. So, urinary incontinence, and other cardinal findings of this syndrome includes bone-joint deformities, muscle atrophy, gait disturbance, scoliosis, congenital hip dislocation and other orthopedic disorders were detected by the other clinics.

The development of scoliosis and hyperlordosis in these patients may be due to an infarction of the axial musculature of the spinal cord (3,8,10,16). The closure defect of the caudal part of neural tube causes deformities in the lower extremities and result from the weakness of the leg muscles and the deterioration of the reciprocal activity of the agonist-antagonist muscles. This causes gait disturbances, contractures in the joints of the lower extremities, and causes the patient to remain in the long-term bed. To live bedridden long-term reveal the secondary muscle atrophy of the leg muscles (2,9,11). In our series, leg pain was the most common symptoms in adults due to the deformities in the lower extremities as well as weakness of the legs. But scoliosis was present in only 10% of cases among adult patients.

A few radiological methods can be used in TCS. Among them, direct lumbar radiography, computerized tomography, magnetic resonance imaging can be counted in children and adults (8). Ultrasonography can be used in neonates. In addition, electromyography, SEPs, and urodynamic and uroflowmetric studies can be helpful in the objective evaluation of neurological and urological deficits. In our series, we used these methods both in preoperative diagnosis and postoperative follow-up of all the cases in our clinic. Particularly, preoperative and postoperative SEPs recordings are an important electrophysiological method in the objective evaluation of early and late postoperative patients (12,14). Intraoperative neurophysiological monitoring is also very useful for a safe and effective surgery (Figure 4). MRI is the main preoperative radiological method. The prolongation and fibrotic thickening of the filamentous fascia (2 mm and thicker) is seen except at the lower end of the spinal cord or at L2 level in MRI. In addition, other malformations or dysmorphological tumors (dermoid, epidermoid) accompanying this syndrome can also be seen on MRI. Direct radiographs and CT examinations may show bone pathologies (spina bifida, vertebral fusions) and calcification of the filum (6,8,9,15,18). Although the first symptoms of TCS include bloating, excessive hair growth, hyperpigmentation on the lumbar region, the exact topographical status of the conus medullaris and filum terminale is revealed by MRI exam, which supports the diagnosis of this syndrome. In our series, the most common clinical symptoms were back and leg pain, hypertrichosis, swelling in the lumbar region, urinary incontinence, gait disorder, orthopedic deformities and impotence. Meningocele or myelomeningocele, syringomyelia, rotoscoliosis and lipoma and other malformations (dermoid tumor, sacrum and coccygeal agenesis, Tarlov cyst, hemivertebra) have been the most common spinal bifida, split cord malformation, and thoracolumbar vertebrae fusions from the spinal

malformations accompanying the syndrome. Only one third of our patients have SEPs anomalies. We used MRI and CT scans in all patients in pre- and postoperative periods.

## CONCLUSION

Tethered Cord Syndrome is not usually solitary, but is associated with spinal cord and other developmental defects of the spinal canal. Risk factors for the syndrome include not only spinal dysraphism but also past spinal surgical history in childhood. This syndrome usually presents with urinary and fecal incontinence, and motor and sensory deficits. The basic useful radiological method for the evaluation and follow-up of TCS is MRI. The main treatment step of this syndrome is to release the spinal cord by cutting the filum terminale with the adhesions in the spinal cord. The earlier the surgical treatment is done, the greater the chance of improvement or stabilization of the neurological deficits is achieved.

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