

**RESEARCH ARTICLE**

# Orthopedic Lesions in Tethered Cord Syndrome: The Importance of Early Diagnosis and Treatment on Patient Outcome

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## Abstract

**Background:** Many of the patients with tethered cord syndrome (TCS) are admitted because of neurological symptoms, while some are admitted because of their orthopedic, urologic, anorectal, and dermatologic manifestations. Consequently, this study aimed to evaluate the importance of early diagnosis and treatment of tethered cord syndrome on patient outcome.

**Methods:** Forty-three patients who underwent surgery because of tethered cord syndrome from 2006 to 2010 were studied. Many of these cases were referred by orthopedic surgeons. All of the findings were recorded and follow up was done twice (1 and 3 years after surgery).

**Results:** Thirty-seven patients were less than 7 years old and 6 were between 17 to 33 years old. According to clinical and neurological exams, satisfactory results were achieved in both groups. Those with early surgical intervention, especially in their early follow up assessment, had the best results. Seventeen cases were referred by an orthopedic surgeon because of manifestations such as leg weakness and numbness, leg pain and spasticity, pes cavus, claw toes, and leg or foot length discrepancy. Cerebrospinal fluid leakage occurred in 3 cases after surgery and 1 showed pseudomeningocele formation.

**Conclusions:** After one year of follow up, initially the results of the treatment were better in early operated cases, but in later follow up assessment (after 3 years) the results were almost the same in both of the groups.

**Key words:** Orthopedic lesions, Tethered cord syndrome, Tight filum terminale

## Introduction

Tethered cord syndrome is defined as a neurological abnormality in which the spinal cord is not able to slide normally inside the spinal canal. This disorder is mechanically caused by the effect of a non-elastic structure on the caudal spinal cord (filum terminale), limiting the upward movement of the lumbosacral spinal cord (1).

According to the rapid growth of the spine, rather than the spinal cord during the neonatal period and childhood, a tethered cord can force the spinal cord to

be stretched more than normal, which is exacerbated in flexion and extension due to further stretching of the spinal cord (2).

Tethered cord syndrome is mostly observed in children and clinical symptoms may include pain, neurological disorders, orthopedic dysfunctions, and dermatologic and sphincter disorders. The syndrome is commonly reported accompanied with spina bifida (3). A series of metabolic disorders are also associated with this phenomenon, such as hypoxia, decrease of cytochrome a3, and the reduction of the interneuronal potential in

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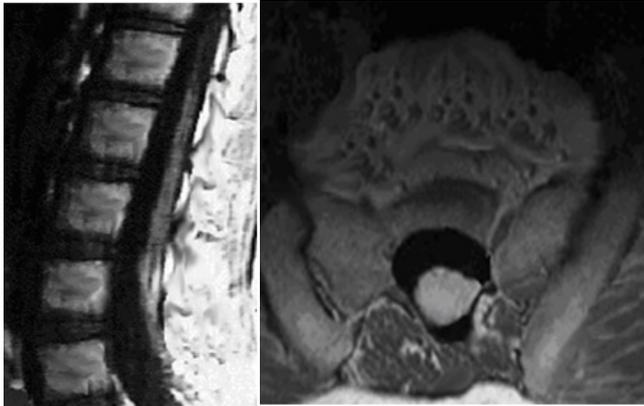


Figure 1. Tethered cord syndrome in a 5-year-old girl.

the spinal cord (4). It is an accepted fact that surgery can prevent the neurological deficits of the spinal cord and to some extent, may restore some of its functions (5).

This study has been performed to introduce clinical findings, evaluate surgical results, and identify orthopedic lesions of this syndrome, which can then assist clinicians in finding a definite diagnosis.

### Materials and Methods

The present study has been conducted in the departments of neurosurgery at the Mashhad University of Medical Sciences, Mashhad, Iran from 2006 to 2010. A total of 43 individuals with tethered cord syndrome, who

underwent surgery were enrolled into the study and 17 patients were referred to our department by orthopedic surgeons. The patients (27 females and 16 males) were mostly under 7 years old (37 cases), while 6 were adults and between 17 to 33 years old at the time of onset or manifestation of neurological symptoms of tethered cord syndrome. Also, the patients were at a different age at the time of diagnosis and surgery.

The surgery was performed using laminectomy or laminotomy, and detethering of the spinal cord by removing the lesion or dissecting the tight filum terminale (Figures 1, 2). Surgery indications included one of the following with a definite diagnosis by MRI:

1. A progressive or new neurological finding
2. Back pain or a progressive pain in distal extremities
3. A progressive musculoskeletal disorder

The whole clinical and neurological findings, radiographic and MRI manifestations, demographic characteristics, related disorders and abnormalities, and the cause of this syndrome of the study groups are listed in relevant tables. All of the patients were followed up in two intervals of 1 and 3 years, and the process of clinical and neurological improvements and radiographic and MRI findings were recorded in separate tables.

The study was approved by the Research Committee of the Mashhad University of Medical Sciences and adult patients or a parent of children signed the consent form in order to be enrolled into the research study.

### Results

From a total of 37 patients, there were 5 cases who suffered from myelomeningocele, 3 cases had meningocele, 4 cases had diastematomyelia, 2 had epidermoid, and 23 were free of these lesions and only demonstrated tight filum terminale. In 6 patients who were operated recently, 2 had diastematomyelia, 1 had a dermoid tumor, and in three cases the tight filum terminale was identified.

In the group under 7 years old, low back pain (LBP) especially during activities, was the initial symptom in 21 cases, while 19 cases showed progressive motor disorders, 26 had sensory disorders such as hypoesthesia, 7 suffered from sensory loss below the level of the lesion, 13 had muscle stiffness in the spinal region, 11 had frequent contractions in the lower extremities, and 23 had sphincter disorders such as urinary or bowel incontinence. Also, there were 4 cases with dimples in the lumbosacral region, 11 cases with deformity of legs, and 11 cases with scoliosis.

Of the 17 patients who were referred to us by orthopedic surgeons, 3 had leg weakness, 7 suffered from numbness in their legs, 1 had spasticity in the distal extremities, and 5 had leg pain. Also, manifestations of spinal deformity (4 cases), pes cavus (6 cases), claw toes (2 cases), and leg or foot length discrepancy (5 cases) were clearly observed.

In the adult group, which was comprised of 6 patients, there were 5 LBP cases, 4 cases with progressive motor disorders, 4 cases with sensory disorders such as paresthesia, 2 cases with dysesthesia, 5 cases with muscle stiffness in the spinal region, 1 case with contractions of

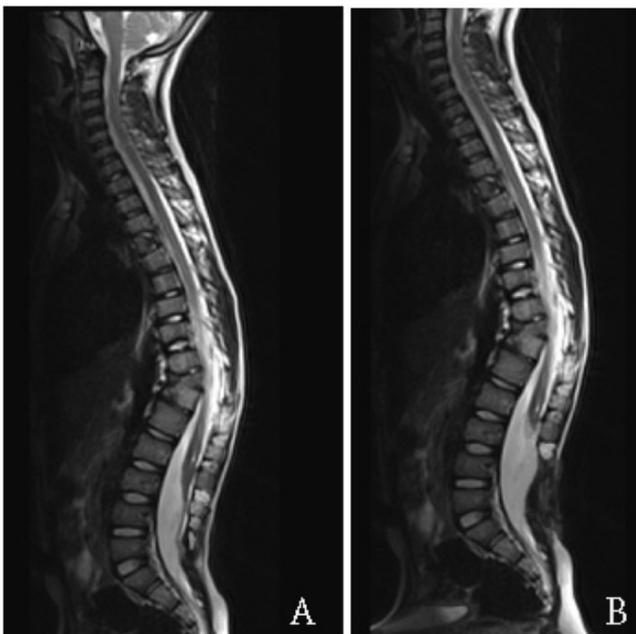


Figure 2. Tethered cord syndrome with hemivertebrae and kyphoscoliosis in a 16-years old boy before (A) and after (B) detethering.

**Table 1. Clinical & neurological findings in our patients with tethered cord syndrome before and after surgery**

S&S Time		LBP	Motor disorder	Sensory disorder	Stiffness In spine	Contraction In lower extremities	Sphincter disorder	Scoliosis
Before Surgery	<b>Child</b> (Number)	57% 21/37	51% 19/37	70% 26/37	35% 13/37	30% 11/37	62% 23/37	30% 11/37
	<b>Adult</b>	83% 5/6	67% 4/6	83% 5/6	83% 5/6	17% 1/6	50% 3/6	50% 3/6
After 1 Year of surgery	<b>Child</b>	27% 10/37	16% 6/37	81% 30/37	13% 5/37	19% 7/37	27% 10/37	5% 2/37
	<b>Adult</b>	0% 0/6	67% 4/6	33% 2/6	66% 4/6	0% 0/6	33% 2/6	17% 1/6
After 3 Years of surgery	<b>Child</b>	19% 7/37	11% 4/37	49% 18/37	8% 3/37	16% 6/37	27% 10/37	5% 2/37
	<b>Adult</b>	0% 0/6	0% 0/6	17% 1/6	50% 3/6	0% 0/6	33% 2/6	17% 1/6

distal extremities, 3 cases with sphincter disorders such as incontinence, and 3 cases with scoliosis.

After following the patients for one year, 11 cases recovered from LBP in the group under 7 years old. While progressive motor disorders were impeded in 13 cases and there was no trace of this disorder in 10 cases, sensory disorders recurred in 4 patients. There was improvement in 8 cases with muscle stiffness in the back, 4 cases with contractions of distal extremities, and 11 cases with urinary incontinence and 2 cases with fecal incontinence. Moreover, the severity of scoliosis did not progress in 9 cases.

In the adult group, 5 cases no longer had back pain and 2 recovered from progressive motor disorders, but it recurred in 2 other patients. Three cases were free from sensory disorders while dysesthesia in 2 cases and in one case each with muscle stiffness in the back, contractions of the lower extremities, and sphincter dysfunction was reversed. Albeit scoliosis did not develop further in 2 cases; one case required surgery in order to correct the scoliosis after one year and the operation was performed successfully.

After 3 years of follow up, LBP and motor disorders improved significantly in 14 and 15 children respectively. Sensory disorders in 8 cases, muscle stiffness of the spinal region in 10 cases, contractions on the lower extremities in 5 cases, sphincter dysfunctions in 13 cases, and intestinal disorders in 3 disappeared. The scoliosis angle decreased in 4 cases, and 9 cases showed signs of a halt in the progression of scoliosis.

In the adult group, LBP in 5 cases, motor disorders in 4 cases, sensory disorders in 4 cases, dysesthesia in 2 cases, muscle stiffness of the spinal region in 1 case, contractions of the lower extremities in 1 case, and sphincter disorders in 1 case improved. Two patients showed progressive scoliosis without surgical intervention and 1 patient underwent surgery to correct the scoliosis (Table 1).

Subsequent complications of surgery were observed in 4 patients; 3 of which was leakage and one pseudomeningocele. While infection and neurological deterioration was not present in the observed patients, one person showed retethering of the spinal cord, and so underwent surgery again. From the 17 cases that were referred to us, 14 underwent orthopedic surgery consequently after the detethering surgery.

### Discussion

The comprehensive recognition of tethered cord syndrome as a clinical syndrome was done 30 years ago. The syndrome is also referred to as "tight filum terminale", "tethered conus (trapped conus medullaris)" and "filum terminale syndrome". The clinical findings of the syndrome were first noticed and documented by Fuchs and Linchtenstai. At that time, this disorder could only be diagnosed by surgery, but in recent years MRI has become a successful non invasive tool for an accurate diagnosis of the disease (1, 6). Tethered cord syndrome is more likely diagnosed in patients who have been referred with concomitant spina bifida followed by either myelopathy, progressive radiculopathy, or other disorders such as sphincter complications, motor disorders, back pain, scoliosis or deformity of distal extremities (4,7).

Most cases with tethered cord syndrome in the lumbosacral region are identified in patients with spinal dysraphism. Spina bifida may be seen as aperta, which is a state in which congenital anomalies such as myelomeningocele, meningocele, and myeloschisis may lead to an incidence of tethered cord syndrome. In some cases, spina bifida occurs as occulta, in which lesions such as lipomyelomeningocele, diastematomyelia, neurenteric cyst, and dermal sinus tract are accompanied by tethered cord syndrome. Other types of lesions in the lumbosacral region such as tumors may induce the syndrome and spinal trauma and a previous surgery

are risk factors of the occurrence of this disorder, as well. Moreover, since the syndrome is not limited to the lumbosacral region, rare cases have been reported in the cervical and dorsal regions (4,8).

Clinical findings may be observed as hairy patches in the lumbosacral region, deep dimples, or lipid tumors on the end of the caudal spine in younger ages. These patients usually present with orthopedic complications such as leg deformity, leg weakness, leg numbness, spasticity of the lower extremities, leg pain, spinal deformity, pes cavus, claw toes and leg, or foot length discrepancy, urologic disorders, scoliosis, back pain and spinal tenderness.

Tethered cord syndrome may remain unrecognized for years (9, 10). Involved cases of any age may present at clinic due to sensory and motor disorders, intestinal or urinary bladder sphincter disorders, and in some cases incapability of motion; all of which could be indicated after imaging evaluations. In a study conducted by Selcuki and Inan, 73% of children with tethered cord syndrome had back pain, 80% had motor dysfunction, and 57% had sensory dysfunction. In addition, urinary and intestinal disorders were reported as 73% and 29%, respectively. Scoliosis was present in 47% of patients as well. In adults, back pain was reported in all cases, while 67% of cases had motor disorders and 62% had sensory disorders. Urinary and intestinal disorders were reported as 67% and 17%, respectively (2). Delay in diagnosis may be due to the lower tension on the spinal cord which exacerbates with the growth of the spine. In some cases, these people become symptomatic following spinal cord cysts and syringomyelia (11,12).

Tethered cord syndrome is related to neural tube impairment in the embryonic period and its physiopathology has remained unknown. In investigations on the mitochondria, it has come to light that as the arterial pressure of oxygen decreases in humans and animals, cytochrome a3 declines as well. These alterations are reversible until the hypoxia has not induced infarction. (13). Yamada et al. noted that metabolic disorders, particularly in interneurons of the spinal cord may be emerged followed by this impairment in oxidative metabolism. The same event occurs in tethered cord syndrome due to the stretching of the spinal cord and may have irreversible drawbacks if not treated in advance (14,15).

If the clinical symptoms of tethered cord syndrome are suspected, it should be proven by imaging diagnostic measures such as MRI, myelography, CT scan, and ultrasonography. MRI is the preferred tool (11, 16, 17). In children, early surgery is recommended to prevent further neurological deterioration. In adults, surgery to free (detether) the spinal cord can reduce the size and further development of cysts in the spinal cord (if they exist) and may impede developing clinical and neurological signs and in some cases alleviate other symptoms as well. With surgery, individuals with tethered cord syndrome return to normal life expectancy. However, some neurological impairments may not be fully correctable, which are mostly reported in patients who undergo surgery in older ages (18).

Treatment of symptomatic cases of the syndrome is reliable nowadays; nonetheless, some researchers also advice repairing the disorder using prophylactic treatments. They also consider imaging techniques to be adequate presurgical examinations. The recommended surgical measures include laminectomy or laminotomy, and spinal cord detethering. Recovery has a close relation with the degree of neurological impairments prior to surgery. Also, prompt diagnosis and treatment has been associated with satisfactory outcomes (12,19). Postoperative complications include CSF leakage, pseudomeningocele, neurological deficits, rethetering, and mortality; particularly in the neonatal period. Bulsar and Zomorodi's study reported infections at 6.6%, CSF leakage at 13.2%, pseudomeningocele at 4.4%, rethetering at 4.4%, and neurological sign intensification at 2% (19). In the present study, CSF leakage occurred in 11.1% of the cases and pseudomeningocele in 3.7%; however, the other above-mentioned complications were not present in our patients. Since these patients are usually suffering from a series of orthopedic, anorectal, urologic, cutaneous, and neurological issues concomitantly as well, there is no doubt that an early diagnosis is beneficial to relieve the patients' health issues. According to our clinical and neurological evaluations, there are satisfactory results with surgery and imaging diagnostic measures, particularly when the treatment is done early on in younger patients.

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## References

1. Pinto FC, Fontes RB, Leonhardt Mde C, Amodio DT, PorroFF, Machado J. Anatomic study of the filum terminale and its correlations with the tethered cord syndrome. *Neurosurgery*. 2002; 51(3):725 -9.
2. Tu A, Steinbok P. Occult tethered cord syndrome: a review. *Childs Nerv Syst*. 2013;29(9):1635-40.
3. Akay KM, Ersahin Y, Cakir Y. Tethered cord syndrome in adults. *Acta Neurochir*. 2000; 142(10):1111-5.
4. Kural C, Guresci S, Simsek GG, Arslan E, Tehli O, Solmaz I, et al. Histological structure of filum terminale in human fetuses. *J Neurosurg Pediatr*. 2014; 13(4): 362-7.
5. Chern JJ, Dauser RC, Whitehead WE, Curry DJ, Luerssen TG, Jea A. The Effect of Tethered Cord Release on Coronal Spinal Balance in Tight Filum Terminale. *Spine*. 2011;36(14):944-9.
6. Krassioukov AV, Sarjeant R, Arkia H, Fehlings MG. Multimodality intraoperative monitoring during complex lumbosacral procedures: indications, techniques, and long-term follow-up review of 61 consecutive cases. *J Spine Neurosurg*. 2004; 1(3): 243-53.
7. Yamada S, Siddiqi J, Won DJ, Kido DK, Hadden A, Spitalieri J, et al. Symptomatic protocols for adult tethered cord syndrome. *Neurol Res*. 2004; 26(7): 741-4.
8. Cornips EM, Vereijken IM, Beuls EA, Weber JW, Soudant DL, van Rhijn LW, et al. Clinical characteristics and surgical outcome in 25 cases of childhood tight filum syndrome. *Eur J Paediatr Neurol*. 2012; 16(2):103-17.
9. Swaroop VT, Dias L. Orthopaedic management of spina bifida-part II: foot and ankle deformities. *J Child Orthop*. 2011; 5(6):403-14.
10. Bowman RM, Mohan A, Ito J, Seibly JM, McLone DG. Tethered cord release: a long-term study in 114 patients. *J Neurosurg Pediatr*. 2009; 3(3):181-7.
11. Sanchez T, John RM. Early Identification of Tethered Cord Syndrome: A Clinical Challenge. *J Pediatr Health Care*. 2014; 28(3):23-33.
12. Ailawadhi P, Kale SS, Agrawal D, Mahapatra AK, Kumar R. Primary tethered cord syndrome--clinical and urological manifestations, diagnosis and management: a prospective study. *Pediatr Neurosurg*. 2012; 48(4):210-5.
13. Lew SM, Kothbauer KF. Tethered cord syndrome: an updated review. *Pediatr Neurosurg*. 2007; 43(3):236-48.
14. Filippidis AS, Kalani MY, Theodore N, ReKate HL. Spinal cord traction, vascular compromise, hypoxia, and metabolic derangements in the pathophysiology of tethered cord syndrome. *Neurosurg Focus*. 2010; 29(1):9.
15. Stavrinou P, Kunz M, Lehner M, Heger A, Müller-Felber W, Tonn JC, et al. Children with tethered cord syndrome of different etiology benefit from microsurgery-a single institution experience. *Childs Nerv Syst*. 2011; 27(5):803-10.
16. Cools MJ, Al-Holou WN, Stetler WR Jr, Wilson TJ, Muraszko KM, Ibrahim M, et al. Filum terminale lipomas: imaging prevalence, natural history, and conus position. *J Neurosurg Pediatr*. 2014; 13(5):559-67.
17. Agarwalla PK, Dunn IF, Scott RM, Smith ER. Tethered cord syndrome. *Neurosurg Clin N Am*. 2007;18(3):531-47.
18. Yong RL, Habrock-Bach T, Vaughan M, Kestle JR, Steinbok P. Symptomatic Retethering of the Spinal Cord Following Section of a Tight Filum Terminale. *Neurosurgery*. 2011; 68(6):1594-601.
19. Haq I, Cruz-Almeida Y, Siqueira EB, Norenberg M, Green BA, Levi AD. Postoperative fibrosis after surgical treatment of the porcine spinal cord: a comparison of dural substitutes. Invited submission from the Joint Section Meeting on Disorders of the Spine and Peripheral Nerves, March 2004. *J Neurosurg Spine*. 2005; 2(1):50-4.