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Surgical Treatment Results of Adult Tethered Spinal Cord Syndrome

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ABSTRACT

AIM: To present symptoms and surgical results of patients operated on with the diagnosis of tethered cord syndrome (TCS) in adulthood.

MATERIAL and METHODS: In this retrospective study, 20 patients older than 18 years were evaluated. There were 17 female and 3 male patients. Patient demographics, clinical and surgical results were evaluated.

RESULTS: The mean age was 29.9 years and the mean follow-up period was 30 months. Of the 20 patients with low back and leg pain, 16 had relief. Two were partially benefited and 2 were unchanged. In nine patients, carrying heavy loads, excessive exercise, childbirth, and walking too long before the onset of symptoms were detected. Complaint of urinary incontinance was improved only in one patient. Three of them continued with frequent urination. There was no change in two of them. There was no improvement in sensory dysfunction and sphincter dysfunction.

CONCLUSION: In patients diagnosed with TCS in adulthood, symptoms may begin with physical activity, and clinical recovery is better when early surgery is performed.

KEYWORDS: Adult tethered cord syndrome, Tethered cord, Tethered cord syndrome, Incontinence

ABBREVIATIONS: TCS: Tethered cord syndrome, **MRI:** Magnetic resonance imaging, **SSPE:** Somatosensory evoked potential

INTRODUCTION

Tethered cord syndrome (TCS) is a disease characterized by progressive neurologic deficits occurring due to congenital or acquired spinal cord stretching (17). Although TCS frequently develops in the pediatric age group, cases in the adult age group have also been rarely reported (17,21).

TCS is characterized by skin lesions such as skin hypertrichosis, hemangiomatous color changes, dermal sinus tract and subcutaneous lipoma, lower extremity weakness, gait disturbance, sensory deficits, bladder dysfunction, and spina bifida. In addition lower extremity weakness,gait disturbance, sensory deficits, and spina bifida may also be observed. Its most common etiologic factors are short and thick filum terminale, diastematomyelia, intradural lipoma, and lipomyelomeningocele. Scoliosis and kyphosis deformity develop in approximately 30% of patients (9,17). Patients with TCS may remain symptom-free for years and may not need any treatment. Symptoms may rarely appear for the first time in adulthood.

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This study aimed to identify factors that cause the emergence of symptoms, how and when surgical planning should be done, the effects of accompanying spinal pathologies on surgical results, and the results of surgical treatment with all its components in patients diagnosed with TCS during adulthood.

MATERIAL and METHODS

We reviewed 20 patients diagnosed with adult TCS who were operated on and followed in our clinic between 2010 and 2021. Patients under 18 years of age, with disorders such as diastematomyelia, and lipomyelomeningocele, without adequate documentation, or with at least 6 months of follow-up were excluded from the study.

Patients with symptomatic TCS were operated on. Radiographic signs of TCS include the presence of a low-lying conus (typically below L2), a thickened filum terminale (>2 mm), and lipomatous infiltration within the flum (12).

All patients underwent hemilaminectomy for filum sectioned. The dura was opened and the filum was cut distally in all patients. A waterproof dura closure was performed (Figures 1-4).

Data were analyzed in terms of demographic characteristics, admission characteristics, and radiological findings. Pain, motor deficit, sensory deficit, and urinary symptoms were evaluated at follow-up.

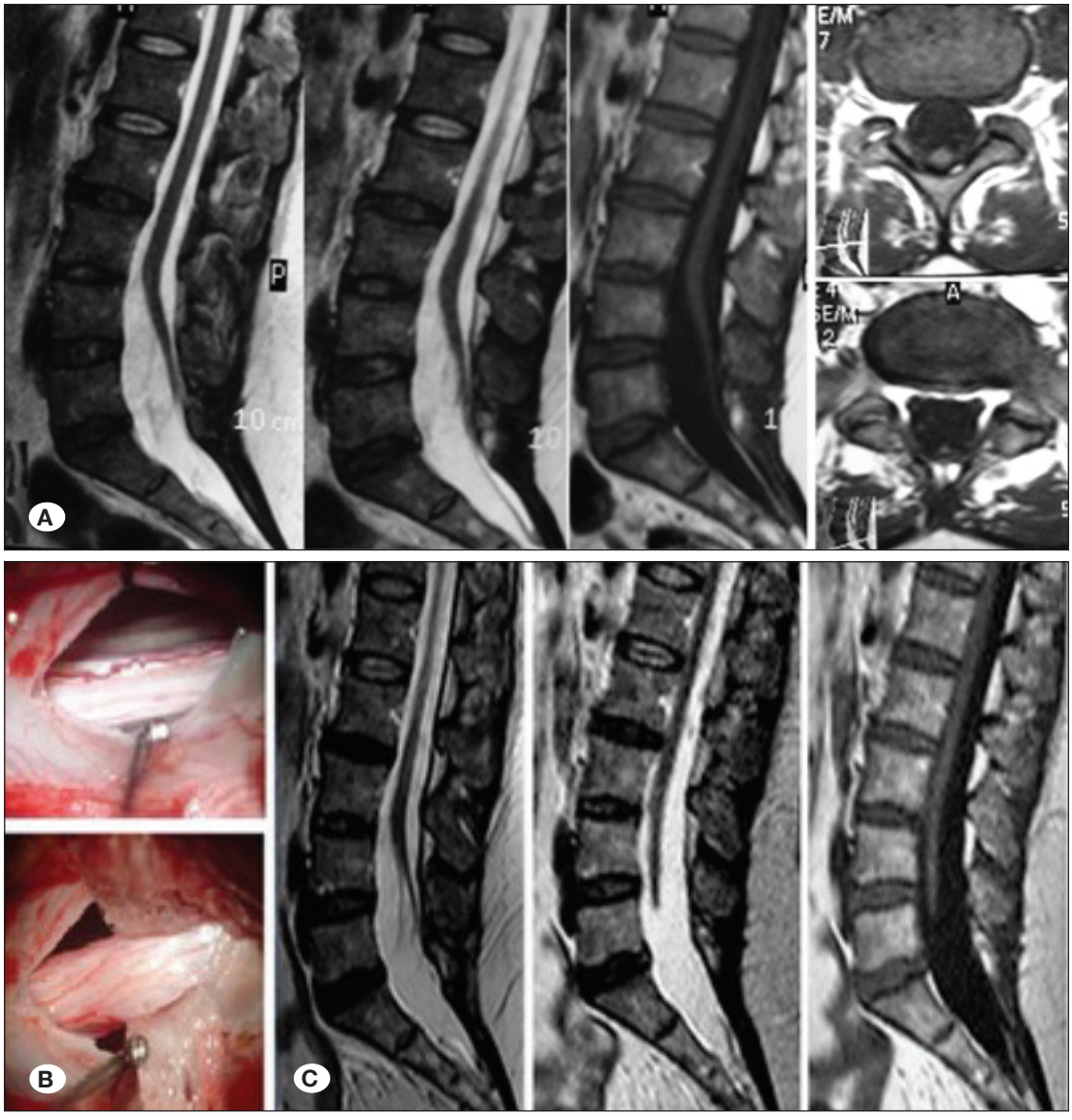

Figure 1: A) A 35 year-old female patient with bilateral leg pain, numbness, and frequent urination for 3 years. T2W and T1W sagittal, T1W axial MRI images show the TCS. The conus appears to end in the L4-5 levels. **B)** Operative view of filum terminale. **C)** Postoperative 10 years control. There are no symptoms. The control sagittal MRI images (left; supine, middle, and right; prone position) showed that there is no tethering.

Figure 2: A) A 49 year-old female patient, complaining of pain in the lower back and legs. T2W and T1W sagittal MRI images show the TCS. **B)** She has no complaints except for mild low back pain in the 5th year postoperative. T2W and T1W sagittal MRI show that the spinal cord position is the same as the preoperative view.

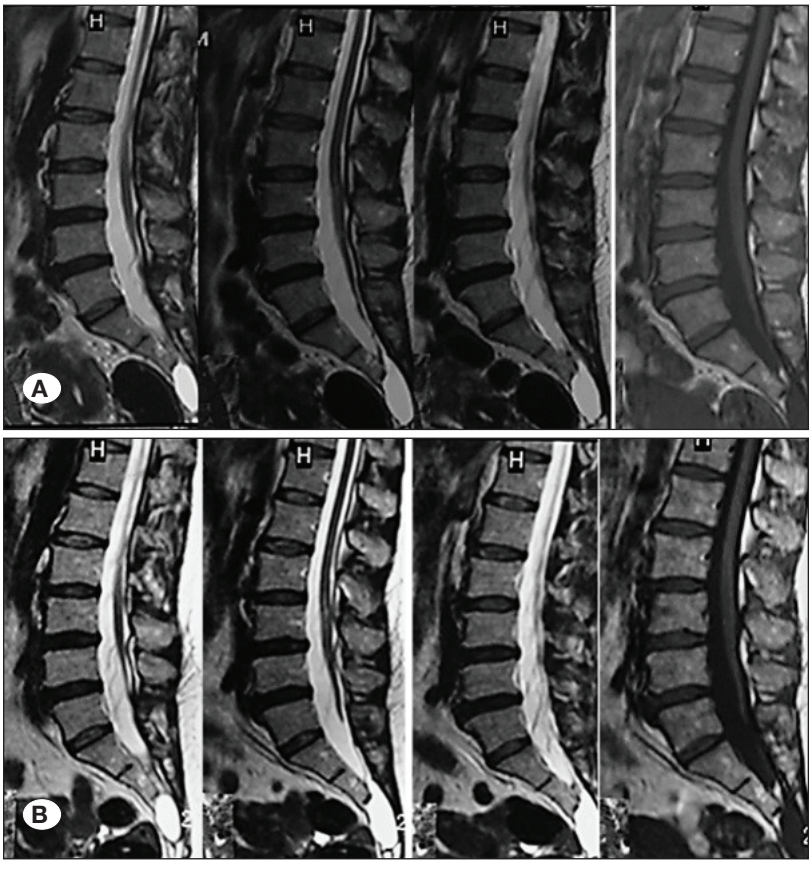

Figure 3: A) A 21 year-old female patient, complaining of pain and numbness in the lower back and legs. T2W and T1W sagittal MRI show that the spinal cord ends L4-5 levels **B)** Leg pain gone and back pain and numbness reduced. T2W and T1W sagittal MRI show that the spinal cord untethered.

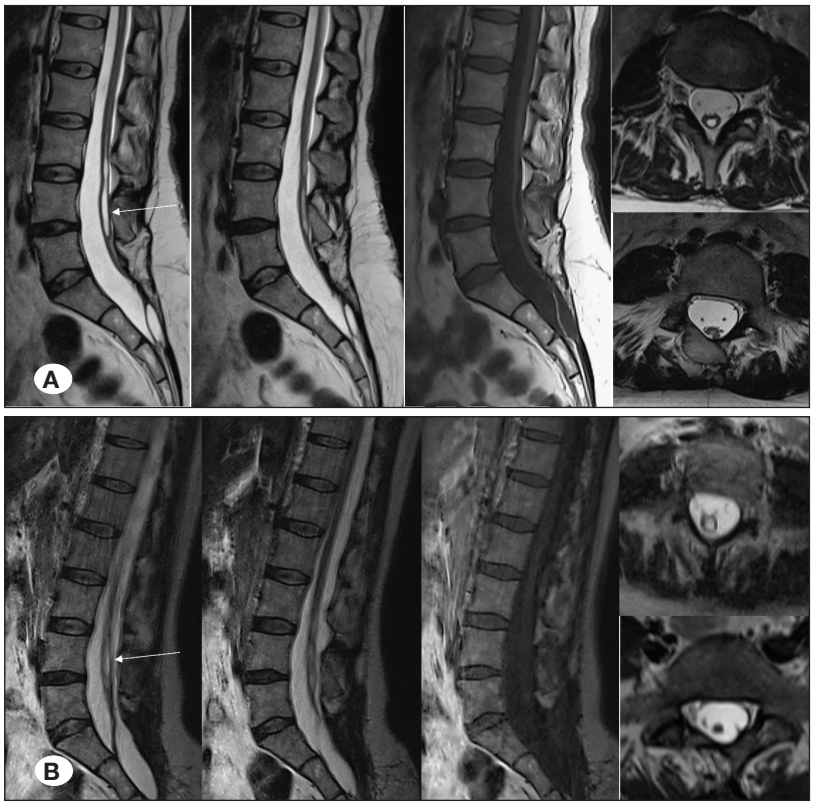

Figure 4: A) A 46 year-old female patient was operated 3 times for tethered cord. Complaints of pain, numbness and frequent urination in the lower back and legs. No significant loss of strength. T2W and T1W sagittal, and T2W axial MRI show that the spinal cord ends L5-S1 levels. **B)** Postoperative 3rd month: She describes mild low back pain. His numbness continues although it has decreased. There was no change in frequent urination. T2W and T1W sagittal, and T2W axial MRI show that the spinal cord untethered and syringomyelia decreases. However, there is a new band starting from the conus and extending to the anterior surface of the canal.

Statistical Analysis

The data were analyzed in SPSS 22 software. Frequency, percentage, mean value, standard deviation, and highest and lowest (min-max) values were used for descriptive statistics. For statistical analysis of categorical data, Fisher's Exact Test was applied for values below five. The statistical significance of the difference was accepted as $p<0.05$.

The institutional ethics committee of Erciyes University approved this study (No: 2023/111).

RESULTS

Of the 20 patients included in the study, 17 (85%) were female and 3 (15%) were male. The mean age of the patients was 29.8 ± 7.7 (min-max: 21-52) years. The main symptoms and findings of the patients are given in Table I. All patients had low back, hip and leg pain at presentation. Loss of sensation in the form of numbness in the legs was present in 9 patients. There was a history of trauma in 3 patients and a history of heavy back exercise or exaggerated walking was obtained before the onset of symptoms in 9 of the patients. Sphincter dysfunction was observed in 6 patients and spinal deformities such as kyphosis or scoliosis in 4 patients. Two patients had a loss of strength in the extremities (one patient had a right foot drop and the other patient had 3/5 loss of strength in dorsal flexion in the left ankle).

Conus was most frequently observed at the L5 and L4 levels. It was less frequently at the L3 level. Conus levels of the patients are given in Table II. Sixteen (80%) of the patients with back, hip, and leg pain had complete recovery. Complete recovery was seen in only one of the patients with urinary incontinence, while complete recovery was not seen in sensory and sphincter dysfunction (Table III).

When the patients' history of heavy physical activity before the onset of symptoms was evaluated, it was observed that there was a significant relationship between the history of heavy physical activity and the onset of symptoms ($p<0.05$).

In our series, among the six patients with back and leg pain who were operated on in the late period (≥3 months), two had completely resolved symptoms, two had partial improvement, and two had no improvement. Among them, two had spinal lipoma, three had concomitant scoliosis, and one had a club foot. Although early surgery was performed on this patient, the motor strength did not improve. Late surgery and the presence of additional spinal pathologies seem to have a negative effect on surgical results ($p<0.05$).

DISCUSSION

TCS is a pathology with a wide clinical spectrum, ranging from the lower extremity motor and sensory loss to urinary and urogenital system dysfunction due to conus medullaris traction. In the pediatric age group, the diagnosis is relatively easier with accompanying midline closure defects, such as meningocele, myelomeningocele, spina bifida, lipomyelomeningocele, and intradural spinal teratoma. Previous studies reported that TCS is associated with foot deformities in 32-63%, scoliosis in 16-29%, and dermal manifestations in 46-98% of patients (1,3,4,17).

Menezes et al. noted that most adults with newly diagnosed TCS have unrecognized neurocutaneous abnormalities and neurological deficits and that findings of non-dermatomal sacral or perineal pain, bladder dysfunction, and neurological deficits should not be confused with hip or degenerative lumbosacral disease, as addressing the primary pathology usually leads to successful outcomes (11).

In patients with asymptomatic TCS without abnormal examination findings until a certain age, phisical causes such as trauma affecting the spine, sitting for too long and/or awkwardly, making inverted movements, carrying heavy loads, excessive exercise, childbirth, and walking for too long may lead to the onset of symptoms (14). In our study, nine patients (45%) had similar conditions that would cause the emergence of symptoms. Thus, these symptoms should be carefully assessed at the anamnesis stage.

Umur et al. suggested that TCS may be considered as a clinical picture of lumbar disc disease (18). Furthermore, if there is a clinical picture of disc disease but no pathologic appearance is observed on MR examinations (both in terms of disc herniation and thick and fatty filum terminale), somatosensory evoked potential (SSPE) examination should be performed to determine the presence of conduction block or delay that may indicate cord tethering. Zhang et al. reported a definite correlation between TCS and lumbar disc degeneration and lumbosacral angle enlargement, suggesting that the spine reduces the high tension of the spinal cord through disc degeneration (22).

A very important issue in TCS is the differential diagnosis of asymptomatic low-lying conus. TCS is a combination of neurological, urological, orthopedic, and dermatologic signs and symptoms with radiographic evidence of a thickened flum and low-lying conus. Low-lying conus may be asymptomatic for years and is a radiologic diagnosis. The manifestation of TCS symptoms in the absence of a low-lying conus has been referred to as occult TCS (12).

In symptomatic cases, if the low-lying conus and a thick filum terminale is detected, there is a consensus on relieving the tension by cutting the filum terminale, as there is no debate about the presence of tension. Gupta et al. reported in their study that surgical treatment should be performed immediately after diagnosis in all adult patients with TCS even in the absence of any neurological deficit (5). However, they also emphasized that surgery should be performed in accordance with the intraspinal pathology and that complete removal of the lipoma, especially in patients with intraspinal lipoma, may increase the neurological deficit. Ide et al. also reported that spinal shortening osteotomy in TCS minimizes perioperative complications by indirectly reducing the tension in the spinal cord; however, in addition to SSEP and motor-evoked potentials, the use of intraoperative high-resolution ultrasound is essential (7). In our study, all newly diagnosed patients underwent surgery at an early stage. In two patients with intraspinal lipomas, total lipoma excision was not considered, but their sizes were only reduced to allow cord movement.

Table I: Main Preoperative Symptoms and Signs

Symptoms	Frequency	
	n	%
Low back, hip and leg pain	20	100
Heavy exercise or exaggerated walking	9	45
Trauma history	3	15
Leg numbness, loss of sensation (sensory deficit)	9	45
Bladder dysfunction and urinary incontinence	6	30
Skin findings	4	20
Spinal deformity	4	20
Limb strength loss	2	10

Data expressed as % (n/total).

Table II: Frequency of Patients According to Conus Levels

L2		L3		L4		L5		S1	
n	%	n	%	n	%	n	%	n	%
4	20	1	5	6	30	6	30	3	15

Data expressed as % (n/total).

Tablo III: Change in Symptoms and Signs After Surgery

Symptoms and signs	Full recovery		Partial recovery		No recovery	
	n	%	n	%	n	%
Low back, hip, leg pain (n=20)	16	80	2	10	2	10
Sensory deficit (n=9)	0	0	4	44.4	5	55.6
Urinary incontinence (n=6)	1	16.7	3	50	2	33.3
Sphincter defisit (n=6)	0	0	3	50	3	50
Motor deficits (n=2)	0	0	0	0	2	100

Fisher's exact test and percentage of columns were used. Data expressed as % (n/total).

When the conus medullaris remains below the first lumbar vertebra, the spinal cord is stretched and the resulting mechanical, ischemic, and metabolic changes are involved in the formation of TCS. The main pathophysiologic event responsible for the findings in TCS is the mechanical effect of stretching (9,10). A series of metabolic events initiated by the mechanical effect is responsible for the emergence of neurologic deficits. The spinal cord itself is affected by stretching rather than the nerve roots. Chronic conus traction decreases local blood flow, depletes Adenosine Triphosphate (ATP) stores in the cell, and disrupts mitochondrial activity and tissue ischemic development. If the mitochondria are not irreversibly damaged, the elimination of spinal cord stretching will improve oxidative metabolism and neurologic findings (10,17). In our study, the onset of symptoms first occurred with physical activity in nine patients due to sudden spinal cord stretching and the remaining in ischemia.

Van Leeuwen et al. followed 57 adult patients who underwent surgery for TCS for 2 years postoperatively (20). During these follow-ups, 15 patients had improved preoperative loss of muscle strength, 38 had no significant improvement in muscle weakness, 2 patients had mild deterioration, and 2 patients had significant deterioration in muscle function. They reported that patients with diastematomyelia and lipomyelomeningocele or those with rapidly developing neurologic and motor loss preoperatively were at risk for progressive motor dysfunction postoperatively. In conclusion, they emphasized that surgery is effective during the early period of TCS. They also emphasized that long-term follow-up should be performed to demonstrate the efficacy of surgery, considering that the improvement of neurologic deficits postoperatively occurs over time. Novik et al. also reported that successful pain control results can be obtained with spinal cord stimulation in TCS when neurological deficits are stable and pain is the primary cause (13). In our study, pain significantly improved postoperatively, and sensory deficit and sphincter defect partially improved, but strength loss did not improve.

Hüttmann et al. followed 54 patients with adult TCS for 8 years after surgical intervention and reported that neurological dysfunction remained the same in 85% of patients compared to the preoperative period and that the success of the operation depended on early diagnosis and early and complete tethered cord surgery (6). In this study, symptoms have also been reported to recur 5 years postoperatively in 80% of patients who underwent partial surgery. Rinaldi et al. reported that early surgical intervention in TCS is necessary for a good neurologic recovery, the pain complaint improves best with surgical treatment, and sphincter dysfunction usually does not improve (15). Lee et al. revealed that surgical treatment in adult TCS was safe and effective in the improvement of pain and neurologic deficits; however, the chances of a successful second surgery were not good (8).

The longer time elapsed after the onset of symptoms, the lower the success of surgery in the long term (19). Therefore, overlooking midline defects during childhood will affect the progression of neurologic problems during adulthood. Moreover, early surgical intervention stops the progression of neurologic symptoms but is not that effective in the return of lost functions. In addition, rehabilitation programs are also necessary to control postoperative pain complaints and reverse neurological losses (2,16,19). In our series, low back and leg pain were completely resolved in all six patients who were operated on within 3 months in the group of asymptomatic patients. One patient with urinary incontinence also completely recovered. Partial improvement was observed in four patients with sensory deficits.

CONCLUSION

TCS is traditionally believed to be a pediatric disorder defined by the onset of neurological deficits in early childhood. Although this condition originated during embryogenesis, a poorly characterized subset of patients with TCS had subsequently become symptomatic during adulthood.

Spinal pathologies accompanying tethered cords should be considered when planning surgical treatment for adult patients with TCS. Predisposing factors leading to the onset of symptoms should be carefully considered. Thus, the favorable effects of early diagnosis and surgery on treatment outcomes should always be considered. Considering that postoperative recovery of the neurologic condition takes years, long-term follow-up of patients is absolutely necessary to demonstrate the efficacy of surgery.

Declarations

Funding: No financial support was utilized during the study.

Availability of data and materials: The datasets generated and/or analyzed during the current study are available from the corresponding author by reasonable request.

Disclosure: Authors declare no conflict of interest.

AUTHORSHIP CONTRIBUTION

Study conception and design: MM
Data collection: MM, RKK
Analysis and interpretation of results: MM, HU, NAD
Draft manuscript preparation: MM, AS
Critical revision of the article: MM, RKK
Other (study supervision, fundings, materials, etc...): MM, AK
All authors (MM, HU, NAD, AS, AK, RKK) reviewed the results and approved the final version of the manuscript.

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