Clinical Significance of Terminal Syringomyelia and Accompanying Congenital Anomalies of Neurosurgical Interest in Adult and Pediatric Patients with Tethered Cord Syndrome

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Abstract

Magnetic resonance imaging (MRI) can be used to examine tethered cord syndrome (TCS) and terminal syringomyelia (TS). Additionally, there is increasing evidence of an association between congenital anomalies and TCS. We aimed to identify the clinical and radiological characteristics of syringomyelia and other anomalies in pediatric and adult patients with TCS. This study included 54 TCS patients (mean age, 17.37 ± 15.83 years; 31 females) admitted to our department between 2010 and 2019. The patients were divided into two age groups: pediatric (<18 years; 63%) and adult (>18 years). Clinical findings, direct vertebrae radiographs, lower extremity radiographs, and spinal/cranial MRI findings were used to evaluate all patients. Computed tomography (CT) was performed to reveal the structure of the septum in patients with Diastematomyelia. Cranial ultrasonography or CT was performed if the fontanel was open or closed, respectively, in pediatric hydrocephalus cases. Pelvic ultrasonography and urodynamic tests were performed to evaluate other comorbid anomalies and urinary system pathologies. A thick filum terminale (73.3%) and diastematomyelia (44.4%) were found to cause spinal tension. The most common accompanying pathology was syringomyelia (78%). The common symptoms were urinary incontinence and bowel problems (71%), scoliosis (68%), and progressive lower extremity weakness (64.4%). It is difficult to distinguish the exact cause of symptoms in patients with TCS and TS. Due to the greater occurrence of other congenital spinal anomalies accompanying TCS, both preoperative symptoms and clinical findings are more severe in the pediatric group than in the adult group, and postoperative results may be more negative.

Keywords
- tethered cord
- syringomyelia
- diastematomyelia
- scoliosis
- congenital anomalies
- pediatric patients
- magnetic resonance imaging

Introduction

The term “tethered cord syndrome (TCS)” refers to a constellation of symptoms and signs of motor and sensory neuron dysfunction attributable to abnormally increased tension on
the spinal cord, and usually, this is accompanied by a low-placed conus medullaris. Tethered cord is associated with congenital malformations of the spine which are common in children. Congenital TCS occurs during embryonic development. In patients with TCS, tissue from the spinal cord to the sacrum—a known as the filum terminale—causes tension in the spinal cord.

In infants with this syndrome, the symptoms include skin discoloration, bristles, and dimple-shaped pits in the waist area. When diagnosed in childhood, TCS should be controlled before urological, orthopaedic, and neurological problems develop. If treatment is delayed, permanent problems (such as renal failure), uncontrollable bowel and urinary problems (such as urinary and fecal incontinence), scoliosis, and foot deformities are more likely to occur in the future. This may be secondary to other disorders, including meningocele, spinal lipomas, lipomatous filum, and split cord malformations. In adults, TCS occurs mostly due to adhesion that develops following trauma or spinal surgery.

The prevalence of syringomyelia is 8.4/100,000 persons. Syringomyelia is an abnormal cystic dilatation of the central canal of the spinal cord and occurs due to the accumulation of excessive cerebrospinal fluid (CSF). The CSF fuses to the ependymal layer adjacent to the central canal, causing payment. Patients may develop various neurological deficiencies secondary to untreated syringomyelia, some of which may persist despite surgical intervention. Early detection, frequent monitoring, and rapid treatment of the underlying etiology are crucial for minimizing potentially irreversible neurological defects.

Although many theories have been proposed regarding the formation and progression of syringomyelia, the underlying pathogenesis remains unknown. Syringomyelia can occur posttrauma and may be associated with Chiari’s malformations, intramedullary tumors, meningocele, meningocele, or TCS. It typically occurs in the cervical and/or thoracic segments. Terminal syringomyelia (TS) refers to segmental cystic dilatation of one-third of the caudal part of the spinal cord. With the increasing use of magnetic resonance imaging (MRI) and other advanced technologies, TS has become a remarkable finding in TCS. This study aimed to identify the clinical and radiological characteristics of syringomyelia and other anomalies in pediatric and adult patients with TCS. Also, the other goal we aimed for was to answer the following question: is the tethered cord the main cause of clinical worsening in patients with TCS? Or is the clinical worsening due to congenital anomalies that accompany them?

Materials and Methods

This study was approved by the ethics committee of our university.

Patient Population

We retrospectively examined the medical records of 54 patients with TCS who were surgically treated at the neurosurgery clinic of Afyonkarahisar Health Sciences University between January 2010 and December 2019. The patients were divided into two age groups: group 1 (pediatric patients, aged <18 years) and group 2 (adults, aged ≥18 years).

Inclusion Criteria

Inclusion criteria of this study are as follows:

- Patients diagnosed with TCS and treated surgically.
- Patients with newly developed or worsening neurological defects.
- Patients with poor quality of life (back pain, leg pain, cranky legs, etc.).

Exclusion Criteria

Exclusion criteria of this study are as follows:

- TCS patients without active complaints and not treated surgically.
- Chiari’s malformation with symptoms and requiring an operation.
- Adult patients who were fully quadriplegic or paraplegic.

Radiological Evaluations

Spinal and cranial MRI was performed on patients with TCS as part of our standard protocol. The imaging was conducted using a 1.5 Tesla General Electric Signa MRI scanner (General Electric Healthcare, Milwaukee, Wisconsin, United States). The vertebra and lower extremities were evaluated using radiography in all cases. Computed tomography (CT) (Toshiba Medical Systems Corporation, Japan) was performed to reveal the structure of the septum dividing the cord in patients with diastematomyelia. Cranial ultrasonography or CT was performed if the fontanel was open or closed, respectively, in pediatric patients with hydrocephalus. Pelvic ultrasonography and urodynamic tests were performed to evaluate other comorbid anomalies and urinary system pathologies.

Surgical Treatment and Follow-up

All patients were treated following standard surgical principles. Surgical treatment was aimed at primary spinal malformation, and surgery was performed upward from the most caudal area of the cord. This rule did not apply to diastematomyelia. The standard postoperative follow-up periods for our surgical patients are 1 week, 1 month, 3 months, 6 months, and 1 year. However, the patients included in this study were followed-up for at least 2 years (average, 3.5 years).

Surgical Procedure

Under general anesthesia and intraoperative neurophysiological monitoring (IONM), patients were positioned prone. Midline intrusion was performed at the S1 to S2 level. After the paraspinal muscles were sequenced, the laminectomy was performed using a high-speed drill or Kerrison’s rongeur. The ligamentum flavum and adipose tissue were then removed. The microscope was placed in the operation area. The dura mater was opened from the midline and fixed to the para-vertebral muscles with sutures. After exposing all the nerve
roots, filum terminale, and arachnoid bands, the filum terminale was selected using IONM. The filum terminale contains large vessels, is whitish, and looks lighter than roots. The IONM probe was used to determine whether the tissue was neural, as this helped avoid cutting one of the roots instead of the tense filum terminale. The roots were pulled back sideways, and the filum terminale was cut. All connective tissues and the conus medullaris connected to the caudal part of the spinal cord were released. After hemostasis was achieved watertight, duraplasty was performed using 5.0 sutures. Using fibrin adhesive products, anatomical layers are tightly closed. If accompanied by diastematomyelia, the bone septum or fibrous band were resected before the untethering procedure.

Statistical Analysis
Statistical analyses were performed using IBM SPSS Statistics for Windows, version 25 (IBM Corp., Armonk, New York, United States) and Microsoft Excel. Standard descriptive statistics (mean ± standard deviation) was calculated, and Wilcoxon’s signed ranks test was used to compare groups. Statistical significance was set at \( p < 0.05 \).

Results

Study Population
Of the 54 patients included in this study, 31 (57.5%) were women and 23 (42.5%) were men. Groups 1 (pediatric group) and 2 (adult group) accounted for 63% (n = 34) and 37% (n = 20) of the patients, respectively. The mean age of the patients was 17.37 ± 15.83 years (range, 48 hours–71 years).

Clinical Outcomes
The most common symptoms in group 1 were urinary and/or fecal incontinence/retention (64.5%), followed by walking disorders and leg pain. In group 2, lower back pain (70%), followed by leg pain (65%), and urinary incontinence/retention (30%) were most common.

The most common clinical findings in groups 1 and 2 were bladder/bowel dysfunction (64.5 and 30%, respectively) and weakness in the lower extremity (58.8 and 30%, respectively).

Radiological Outcomes
The terminal end of the conus medullaris was located at the L2–5 and L5–S2 levels in 74 (n = 40) and 15% (n = 8), respectively, of the 54 patients. Six (11%) of the patients were anatomically normal (T12–L2). However, when evaluated from a single-level point of view, the tethered cord was most commonly detected at the L5 level.

When the images of all the cases were examined in detail, the most common pathologies associated with TCS were syringomyelia (77.7%), a short, thick filum (74%), diastematomyelia (57.4%), scoliosis (64.8%), and spina bifida (38.8%). However, when we examined the groups separately, the most common accompanying pathologies were syringomyelia (79.1%), a short, thick filum (73.5%), and diastematomyelia (64.7%) in group 1, and a short, thick filum (75%), diastematomyelia (45%), and syringomyelia (40%) in group 2.

Accompanying pathologies were detected in 82.4% of the patients in group 1; however, no congenital anomaly or pathology was detected in 50% of the patients in group 2. Table 2 details the accompanying pathologies in the adult and pediatric groups.

There was a significant difference in the incidence of syringomyelia between groups 1 and 2 (\( p < 0.05 \)). Syringomyelia levels, the widest dimensions, and syrinx indexes were measured in patients with syringomyelia. A statistically significant difference was found between the preoperative and postoperative measurements (\( p < 0.05 \)). Detailed data for all groups are provided in Tables 3 and 4.

Table 1 Presenting symptoms and clinical findings in the neurological examinations of patients with tethered cord syndrome

| Presenting symptoms | Pediatric group (n = 34) | Adult group (n = 20) |
|---------------------|-------------------------|---------------------|
|                     | No. of patients | Percentage | No. of patients | Percentage |
| Urinary incontinence/retention | 22 | 64.5 | 6 | 30 |
| Low back pain | 13 | 38.2 | 15 | 75 |
| Leg pain | 16 | 47 | 13 | 65 |
| Restless leg syndrome | 6 | 17.6 | 6 | 30 |
| Unsteady gait (in those who reached walking age) | 19 | 55.8 | 12 | 60.1 |
| Numbness | 16 | 47 | 5 | 25 |
| Bladder and/or bowel dysfunction | 22 | 64.5 | 6 | 30 |
| Muscular weakness (motor deficit) | 20 | 58.8 | 6 | 30 |
| Sensory deficit | 16 | 47 | 5 | 25 |
| Muscular atrophy | 6 | 17.6 | 3 | 15 |
| Skin lesions | 14 | 41.2 | 5 | 25 |
Accompanying nonneurosurgical pathologies included club feet (18.5%), congenital hip dislocation (9.2%), and kidney anomalies (7.4%). This ranking was not different between the groups (Table 2).

**Table 2** Frequency of accompanying congenital anomalies according to the age distribution of patients with tethered cord syndrome who underwent surgery

| Pathology of accompanying nonneurosurgical anomalies | All patients (n = 54) | Pediatric group (n = 34) | Adult group (n = 20) |
|------------------------------------------------------|----------------------|-------------------------|----------------------|
| Neurosurgical anomalies                              | No. of patients | Percentage | No. of patients | Percentage | No. of patients | Percentage |
| Syringomyelia                                         | 42 | 77.7 | 27 | 79.1 | 8 | 40 |
| Short thick filum terminale                           | 40 | 74 | 25 | 73.5 | 15 | 75 |
| Diastematomyelia                                      | 31 | 57.4 | 22 | 64.7 | 9 | 45 |
| Kyphoscoliosis                                        | 35 | 64.8 | 25 | 73.5 | 9 | 45 |
| Hydrocephalus                                         | 20 | 37 | 18 | 58 | 2 | 8 |
| Chiari’s malformations*                                | 12 | 22.2 | 10 | 29.4 | 2 | 1 |
| (Generic) spina bifida                                | Occulta spina bifida | 21 | 38.8 | 15 | 44 | 14 | 4 |
|                                                        | Meningocele | 6 | 11.2 | 5 | 14 | 1 | 5 |
|                                                        | Meningomyelocele | 3 | 5.6 | 2 | 6 | 1 | 5 |
|                                                        | Myelochisis | 3 | 5.6 | 3 | 9 | 0 | 0 |
| Spinal lipoma                                         | 8 | 14.8 | 5 | 14.7 | 3 | 15 |
| Vertebral fusion defect                                | 23 | 42.5 | 20 | 58.8 | 3 | 15 |
| Only tethered cord without accompanying pathology      | 16 | 29.6 | 6 | 17.6 | 10 | 50 |

Pathology of accompanying nonneurosurgical anomalies

| Orthopaedic deformities                                | Club feet | 17 | 31.4 | 12 | 35.2 | 4 | 2 | 20 | 10 |
|                                                        | Congenital hip dislocation | 5 | 9.2 | 4 | 11 | 1 | 5 |
|                                                        | Varus–valgus deformity | 2 | 3.7 | 1 | 2 | 1 | 5 |
| Anorectal anomalies                                    | 14 | 25.9 | 14 | 41.1 | 0 | 0 |
| Cardiac defect                                         | 2 | 6 | 2 | 6 | 0 | 0 |
| Other                                                  | Kidney anomalies | 17 | 31.4 | 10 | 30 | 6 | 7 | 2 | 10 |
|                                                        | Inguinal hernia | 3 | 5.5 | 1 | 3 | 2 | 10 |
|                                                        | Hydrocele and undescended testicle | 3 | 5.5 | 2 | 6 | 1 | 5 |
|                                                        | Tarlov’s cyst | 3 | 5.5 | 2 | 6 | 1 | 5 |
|                                                        | Dermal sinus tract | 3 | 5.5 | 2 | 6 | 1 | 5 |
|                                                        | Advanced thinning of the thoracic cord | 1 | 1.8 | 1 | 3 | 0 | 0 |

*Small-sized and nonintrusive Chiari’s malformations.

**Treatment and Follow-up**

Untethered surgery was performed on all patients, and only one patient underwent both untethered surgery and syrinx drainage.

Of the 34 patients included in group 1, the motor, sensory, and urinary functions were intact in 41, 41, and 35% of patients, respectively, during the preoperative period. The remaining patients had functional disorders to varying degrees. Compared with the preoperative period, 70, 70, and 54.5% of the patients with the aforementioned disorders showed functional improvement in motor, sensory, and urinary functions, respectively. Of the 20 patients included in group 2, the motor, sensory, and urinary functions were intact in 65, 70, and 65% of patients, respectively. Following surgery, 85.7, 83.3, and 50% of patients in group 2 experienced functional improvements in motor, sensory, and urinary functions, respectively. The clinical findings did not
Table 3  Comparison of preoperative and postoperative measurements of syringomyelia and the accompanying pathology of patients with tethered cord syndrome in the pediatric group

| Age/sex | Preoperative | Postoperative | Accompanying neurosurgical congenital anomalies |
|---------|--------------|---------------|------------------------------------------------|
|         | Level        | Widest diameter (mm) | Syringomyelia area index (%) | Level | Widest diameter (mm) | Syringomyelia area index (%) | |
| 1 7 years/F | NA | NA | NA | NA | NA | NA | None |
| 2 4 years/F | T6–L4 | 0.8 x 1.4 | 0.06 | Unchanged | Unchanged | Unchanged | None |
| 3 4 years/F | T9–12 | 1.3 x 1.9 | 0.09 | Disappeared | Disappeared | Disappeared | None |
| 4 4 years/M | NA | NA | NA | NA | NA | NA | None |
| 5 9 years/M | T10–L1 | 7 x 5 | 0.52 | Disappeared | Disappeared | Disappeared | None |
| 6 4 years/F | NA | NA | NA | NA | NA | NA | None |
| 7 12 years/M | T10–L2 | 0.8 x 1.6 | 0.06 | Unchanged | T10–L2 | 0.6 x 1.1 | 0.04 | DTM + HC + SC |
| 8 17 years/M | T8–12 | 1.3 x 1.9 | 0.09 | Disappeared | Disappeared | Disappeared | None |
| 9 7 years/M | T9–L3 | 4.9 x 5.6 | 0.5 | Unchanged | Unchanged | Unchanged | DTM |
| 10 8 years/F | T9–L3 | 4.9 x 5.6 | 0.5 | Unchanged | Unchanged | Unchanged | DTM + SC |
| 11 14 years/F | T12–L2 | 2 x 1.2 | 0.1 | Disappeared | Disappeared | Disappeared | DTM + SC |
| 12 11 years/F | NA | NA | NA | NA | NA | NA | None |
| 13 3 years/F | T9–L1 | 2.4 x 2.8 | 0.21 | T10–L1 | 1.4 x 2.1 | 0.1 | DTM + OSB + HC + SC |
| 14 12 years/F | L2–5 | 2.7 x 1.5 | 0.19 | L2–3 | 2 x 1.2 | 0.1 | DTM + SC |
| 15 7 years/F | L1–S1 | 1.8 x 2 | 0.11 | L2–4 | 1 x 1.2 | 0.1 | DTM + SC |
| 16 1 years/M | T7–9 | 7 x 5.9 | 0.47 | Unchanged | Unchanged | Unchanged | DTM + SC |
| 17 2 years/F | NA | NA | NA | NA | NA | NA | None |
| 18 14 years/M | L2–S1 | 1 x 2.4 | 0.08 | L2–4 | 0.8 x 1.2 | 0.06 | DTM + MMS + HC + SC |
| 19 10 years/F | T9–L4 | 2 x 1.4 | 0.1 | T12–L2 | 2 x 1.2 | 0.1 | DTM + MMS + SC |
| 20 3 years/M | NA | NA | NA | NA | NA | NA | None |
| 21 2 years/M | L1–S1 | 2.4 x 2.8 | 0.21 | L2–5 | Unchanged | Unchanged | DTM + MMS + HC + CH + SC |
| 22 2 years/F | T9–12 | 1.3 x 1.9 | 0.09 | T11–12 | Unchanged | Unchanged | DTM + MMS + HC + CH + SC |
| 23 3 years/F | L3–4 | 1 x 1.5 | 0.1 | Unchanged | Unchanged | Unchanged | DTM + MMS + HC + CH + SC |
| 24 2 days/M | T12–L3 | 3.3 x 2.8 | 0.32 | T12–L1 | 2 x 1.8 | 0.11 | DTM + MMS + HC + CH + SC |
| 25 2 months/F | L2–3 | 1.4 x 2.1 | 0.1 | Unchanged | Unchanged | Unchanged | DTM + MMS + HC + CH + SC |
| 26 1 week/M | L1–3 | 2.1 x 1.8 | 0.12 | Unchanged | Unchanged | Unchanged | DTM + MMS + HC + CH + SC |
| 27 1 days/M | L1–3 | 1.5 x 1.7 | 0.09 | Unchanged | Unchanged | Unchanged | DTM + MMS + HC + CH + SC |
| 28 3 days/F | T10–L4 | 2.4 x 3.9 | 0.24 | T10–L1 | 1.3 x 1.9 | 0.1 | DTM + MMS + HC + CH + SC |
| 29 5 days/F | NA | NA | NA | NA | NA | NA | MMS + HC + SC |
worsen in any of the patients. A statistically significant difference in improvements was found between the two groups ($p < 0.05$).

The mean and shortest follow-up periods were 51.15 ± 26.85 months and 18 months, respectively. Due to CSF fistula and wound infection, and two patients were operated on again in groups 1 and 2, respectively. The average hospitalization time was 4.15 ± 2.88 days (∗Tables 5 and 6).

**Discussion**

**Presenting Symptoms**

Abdallah et al$^9$ reported lower back pain (68%), leg pain (60%), urinary incontinence or retention (52%), muscular weakness (52%), and numbness (20%) in adult patients with TCS. Similarly, the patients in our study presented with lower back pain (75%), leg pain (65%), unsteady gait (60%), restless leg syndrome (30%), urinary incontinence or retention (30%), and numbness (25%).

Sadrameli et al$^{11}$ found that urinary incontinence or retention, followed by lower back/leg pain and lower extremity weakness, was the most frequent symptom in the pediatric population. In the pediatric group in our study, the most frequent clinical symptoms were urinary and/or stool incontinence, gait disturbance, and lower back/leg pain.

**Clinical Findings**

Abdallah et al$^9$ reported bladder dysfunction (52%), motor deficit (52%), sensory deficit (32%), and muscular atrophy (12%) in adults with syringomyelia. In our study, the patients presented with bladder dysfunction (30%), motor deficit (30%), sensory deficit (25%), skin lesions (25%), and muscular atrophy (15%).

Erkan et al$^{12}$ reported that 62.5, 37.5, 68.8, 12.5, 43.8, 21.9, and 46.9% of pediatric patients presented with lower extremity muscular weakness, bilateral long-tract signs, sensory deficits, lower back pain, urinary incontinence, fecal incontinence, and progressive scoliosis, respectively. Several studies have also found similar results for presenting symptoms. In our study, we found urinary and/or fecal incontinence; lower extremity weakness; sensory deficits; gait impairment; pain in the back, waist, and legs; skin lesions and increased hair growth; restless leg; and leg atrophy and foot asymmetry in 71, 64.4, 60, 55.5, 37.7, 64.4, 31, and 17.7%, respectively, of the pediatric population.

**Associated Malformations**

In their study of 34 patients, Beaumont et al$^{13}$ found that tethered cord is most frequently caused by a thickened or fatty filum (70%). Less common causes included lipoma, meningocele, myelomeningocele, and diastematomyelia. In their study on 132 TCS patients, Erkan et al$^{12}$ concluded that tethering was caused by a thick filum terminale (12%), lumbosacral lipomas (25%), diastematomyelia (31%), repaired lipomyelomeningocele sites (16%), and diastematomyelia associated with terminal lipomas (16%). In a study of 30 patients published by the same researcher in 2000,$^{14}$...
spinal cord tethering was found to be caused by a thick filum terminale (40%), diastematomyelia (43.3%), repaired lipomyelomeningocele site (13.3%), and diastematomyelia associated with a terminal lipoma (3.3%). Abdallah et al. found that the comorbidities accompanying the tethered cord were diastematomyelia (44%), vertebral fusion anomalies (44%), and spinal cord malformation (32%). In our study, a short, thick phylum (74%, n = 40), diastematomyelia (57.4%, n = 20), and kyphoscoliosis (64.8%) were the most common accompanying and/or causative pathologies for TCS. This ranking did not change when stratified by age group; however, the probability of occurrence changed (►Table 2).

To date, the largest study on the association of spinal anatomical disorders with TCS and TS has been conducted by Erkan et al. This study reported progressive kyphoscoliosis, hemivertebrae, block vertebrae, and an unsegmented bar in 50, 67, 13, and 20% of patients, respectively. In our study, we detected kyphoscoliosis in 64.8% of patients, and hemivertebrae, butterfly vertebrae, block vertebrae, and other fusion defects in 42.5% of patients.

### Syringomyelia

Several authors have linked the pathogenesis of syringomyelia with fluid accumulation and changes in local spinal blood flow and oxidative metabolism. Syringomyelia that occurs in the distal third of the spinal cord is called TS, and several studies have linked TS to tethered cord. In a study involving 90 patients with occult spinal dysraphism, TS was detected by MRI in 27% of the cases. In this study, TS often appeared in the tense filum terminale and was accompanied by anorectal anomalies (67%), meningocele manqué (54%), and diastematomyelia (38%). Syringomyelia was found to be below the T6 level in all patients, except in one patient who had holocord syringomyelia. In a similar study, Erkan et al. found that of 132 patients with tethered cords, 32 (24%) had TS. According to Iskandar et al., 38, 34, and 28 of syrinxes were below the T8 level and covered the lower thoracic (T8–T12), lumbar (L1–S1), and thoracolumbar (T8–L4) regions, respectively.

In our study, syringomyelia was detected in 42 (77.7%) of the 54 patients. Only one patient had widespread

### Table 4

| Age/sex | Preoperative | Postoperative | Accompanying neurosurgical congenital anomalies |
|---------|--------------|---------------|-----------------------------------------------|
|         | Level        | Widest diameter (mm) | Syrinx area index (%) | Level | Widest diameter (mm) | Syrinx area index (%) | |
| 1       | NA          | NA             | NA                        | NA    | NA             | NA                        | None |
| 2       | NA          | NA             | NA                        | NA    | NA             | NA                        | None |
| 3       | T11–L1      | 4.4 x 5.2      | 0.49                      | Disappeared | Disappeared | Disappeared | None |
| 4       | T9–12       | 3 x 2.1        | 0.16                      | Unchanged | 1.7 x 1.2      | 0.11                      | None |
| 5       | NA          | NA             | NA                        | NA    | NA             | NA                        | None |
| 6       | T8–12       | 0.8 x 1.6      | 0.06                      | T10–12 | 0.8 x 1.1      | 0.06                      | None |
| 7       | T7–9        | 1.3 x 1.9      | 0.09                      | Disappeared | Disappeared | Disappeared | None |
| 8       | NA          | NA             | NA                        | NA    | NA             | NA                        | None |
| 9       | T12–L2      | 4.9 x 5.6      | 0.5                       | Disappeared | Disappeared | Disappeared | None |
| 10      | NA          | NA             | NA                        | NA    | NA             | NA                        | DTM + LPM + SC |
| 11      | T10–L2      | 3.2 x 1.9      | 0.16                      | T10–12 | 1.5 x 1.7      | 0.10                      | DTM + SC |
| 12      | NA          | NA             | NA                        | NA    | NA             | NA                        | DTM + SC |
| 13      | NA          | NA             | NA                        | NA    | NA             | NA                        | DTM + SC |
| 14      | NA          | NA             | NA                        | NA    | NA             | NA                        | DTM + SC |
| 15      | NA          | NA             | NA                        | NA    | NA             | NA                        | DTM + SC |
| 16      | NA          | NA             | NA                        | NA    | NA             | NA                        | DTM + SC |
| 17      | NA          | NA             | NA                        | NA    | NA             | NA                        | DTM + SC |
| 18      | NA          | NA             | NA                        | NA    | NA             | NA                        | UN + DTM + LPM + HC + SC |
| 19      | L1–3        | 2.5 x 1        | 0.16                      | L1–3  | 1 x 0.8        | 0.06                      | UN + DTM + MMS + LPM + SC |
| 20      | T10–L4      | 2.4 x 1.5      | 0.12                      | T10–L1 | 1.8 x 0.6      | 0.07                      | UN + MMS + HC + SC |

Abbreviations: DTM, diastematomyelia; F, female; HC, hydrocephalus; LPM, lipoma; M, male; MMS, meningocele myelocele schisis; NA, not available; OSB, occulta spina bifida; SC, scoliosis.
Table 5 Preoperative and postoperative symptoms, clinical findings in examinations, length of hospital stay, follow-up periods, and complications in pediatric patients with tethered cord syndrome

| Preoperative clinical presentation | Preoperative clinical examinations | Surgery | Postoperative outcome | Length of hospitalization (d) | Complication, tracking time (mo) |
|-----------------------------------|-----------------------------------|---------|-----------------------|-----------------------------|---------------------------------|
| 1 Lower back and leg pain, unsteady gait | All intact | UN | Improved | Intact | Intact | Intact | 5 | None, 81 |
| 2 UI, leg pain, unsteady gait | U, Ni | UN | Improved | Intact | Intact | Improved | 2 | None, 25 |
| 3 UI, numbness | M, Ni; U, Ni | UN | Improved | Improved | Intact | Improved | 3 | None, 61 |
| 4 Leg pain, unsteady gait | All intact | UN | Improved | Intact | Intact | Intact | 7 | None, 26 |
| 5 Leg pain, numbness | All intact | UN | Improved | Intact | Intact | Intact | 4 | None, 91 |
| 6 Lower back pain, unsteady gait | All intact | UN | Improved | Intact | Intact | Intact | 2 | None, 54 |
| 7 UI, low and leg pain, unsteady gait | U, Ni | UN + DTM + 3VS | Partially improved | Intact | Intact | Improved | 2 | None, 82 |
| 8 UI, leg pain, numbness | M, Ni; S, Ni; U, Ni | UN + DTM | Improved | Intact | Partially improved | Intact | 2 | None, 41 |
| 9 Low and leg numbness | All intact | UN + DTM | Improved | Intact | Intact | Intact | 4 | None, 35 |
| 10 UI, unsteady gait, numbness | M, Ni; S, Ni; U, Ni | UN + DTM | Improved | Improved | Improved | Partially improved | 2 | None, 19 |
| 11 UI, low and leg pain, numbness | M, Ni; S, Ni; U, Ni | UN + DTM | Partially improved | Improved | Partially improved | Unchanged | 2 | None, 20 |
| 12 Leg pain, unsteady gait | All intact | UN + DTM + 3VS | Partially improved | Intact | Intact | Intact | 2 | None, 31 |
| 13 Low pain | All intact | UN + DTM + 3VS | Partially improved | Intact | Intact | Intact | 2 | None, 84 |
| 14 UI, low and leg pain, numbness | M, Ni; S, Ni; U, Ni | UN + DTM | Improved | Improved | Improved | Improved | 4 | None, 87 |
| 15 UI, low and leg pain, numbness | M, Ni; S, Ni; U, Ni | UN + DTM | Improved | Improved | Improved | Improved | 4 | None, 104 |
| 16 Numbness, unsteady gait | All intact | UN + DTM | Partially improved | Intact | Intact | Intact | 2 | None, 54 |
| 17 Low and leg pain, unsteady gait | All intact | UN + DTM | Partially improved | Intact | Intact | Intact | 2 | None, 67 |
| 18 UI, low and leg pain, numbness | M, Ni; S, Ni; U, Ni | UN + DTM + MMS + 3VS | Improved | Improved | Improved | Improved | 3 | None, 21 |
| 19 UI, leg pain, numbness | M, Ni; S, Ni; U, Ni | UN + DTM + MMS | Improved | Improved | Improved | Improved | 3 | None, 112 |
| 20 Low pain, unsteady gait | All intact | UN + DTM + MMS | Partially improved | Intact | Intact | Intact | 3 | None, 32 |
| 21 UI, numbness unsteady gait | M, Ni; S, Ni; U, Ni | UN + DTM + MMS + 3VS | Improved | Improved | Improved | Partially improved | 8 | CSF fistula, 39 |
| 22 UI, numbness unsteady gait | M, Ni; S, Ni; U, Ni | UN + DTM + MMS + V-P | Unchanged | Unchanged | Unchanged | Unchanged | 4 | None, 89 |
(Continued)
| Preoperative clinical presentation | Preoperative clinical examinations | Surgery                          | Postoperative outcome                          | Length of hospitalization (d) | Complication, tracking time (mo) |
|-----------------------------------|-----------------------------------|----------------------------------|-----------------------------------------------|------------------------------|---------------------------------|
|                                  |                                   |                                  | Clinical presentation | Motor | Sensory | Urinary |                                  |                              |
| 23 Unsteady gait, numbness        | M, Ni; S, Ni; U, Ni               | UN + DTM + MMS + V-P              | Partially improved | Improved | Improved | Unchanged | 4                              | None, 72                      |
| 24 UR                             | M, Ni; S, Ni; U, Ni               | UN + DTM + MMS + V-P              | Partially improved | Improved | Improved | Unchanged | 14                             | Wound infection, 45           |
| 25 UR                             | M, Ni; S, Ni; U, Ni               | UN + DTM + MMS + V-P              | Unchanged          | Unchanged | Unchanged | Unchanged | 5                              | None, 45                      |
| 26 UR                             | M, Ni; S, Ni; U, Ni               | UN + DTM + MMS + V-P              | Unchanged          | Unchanged | Unchanged | Unchanged | 3                              | None, 26                      |
| 27 UR                             | M, Ni; S, Ni; U, Ni               | UN + DTM + MMS + V-P              | Unchanged          | Unchanged | Unchanged | Unchanged | 5                              | None, 52                      |
| 28 UR                             | M, Ni; S, Ni; U, Ni               | UN + DTM + MMS + V-P              | Unchanged          | Unchanged | Unchanged | Unchanged | 2                              | None, 40                      |
| 29用人单位，不稳当，不稳当，不稳当，不稳当 | M, Ni; S, Ni; U, Ni               | UN + MMS + V-P                   | Improved           | Improved | Intact   | Intact    | 6                              | None, 30                      |
| 30 UR                             | M, Ni; S, Ni; U, Ni               | UN + MMS + LPM + V-P              | Improved           | Intact   | Intact   | Intact    | 6                              | None, 74                      |
| 31 UR                             | M, Ni; S, Ni; U, Ni               | UN + MMS + LPM + V-P              | Improved           | Improved | Improved | Improved | 3                              | None, 28                      |
| 32 Unsteady gait, numbness, unsteady gait | M, Ni; S, Ni; U, Ni               | UN + MMS + LPM + V-P              | Unchanged          | Partially improved | Partially improved | Unchanged | 4                              | None, 18                      |
| 33 Low and leg pain, unsteady gait | M, Ni; S, Ni; U, Ni               | UN + LPM                         | Partially improved | Intact   | Intact   | Intact    | 2                              | None, 56                      |
| 34 Unsteady gait, numbness, unsteady gait | M, Ni; S, Ni; U, Ni               | UN + LPM                         | Improved           | Improved | Improved | Improved | 2                              | None, 49                      |

Abbreviations: 3VS, third ventriculostomy; CSF, cerebrospinal fluid; d, day; DTM, diastematomyelia; LPM, lipoma; M, motor; MMS, meningomyelocele; mo, month; Ni, nonintact; S, sensory; U, urinary; Ui, urinary incontinence; UN, untethered; UR, urinary retention; V-P, ventriculoperitoneal shunt; wk, week; y, year.
Table 6  Preoperative and postoperative symptoms, clinical findings in examinations, length of hospital stay, follow-up periods, and complications in adult patients with tethered cord syndrome

| Age/sex | Preoperative clinical presentation | Examinations | Surgery | Postoperative outcome | Length of Hospitalization (d) | Complication, tracking time (mo) |
|---------|---------------------------------|--------------|---------|-----------------------|-------------------------------|---------------------------------|
| 1       | 71 years/M                       | Lower back and leg pain, unsteady gait | Intact    | UN                    | Improved                      | Intact                          | Intact                          | Intact                          | 3                               | None, 18                         |
| 2       | 43 years/F                       | Lower back and leg pain, restless leg syndrome | Intact    | UN                    | Partially improved            | Intact                          | Intact                          | Intact                          | 3                               | None, 87                         |
| 3       | 19 years/M                       | Leg pain, restless leg syndrome, UI | Intact    | UN                    | Improved                      | Intact                          | Intact                          | Intact                          | 3                               | None, 42                         |
| 4       | 62 years/M                       | Lower back and leg pain, numbness       | U, Ni; motor; Ni; S, Ni | UN                    | Improved                      | Improved                        | Improved                        | Improved                        | 3                               | None, 31                         |
| 5       | 42 years/F                       | Lower back and leg pain                | U, Ni; motor; N; | UN                    | Partially improved            | Improved                        | Improved                        | Improved                        | 3                               | None, 65                         |
| 6       | 49 years/F                       | Restless leg syndrome, unsteady gait | Intact    | UN                    | Improved                      | Intact                          | Intact                          | Intact                          | 4                               | None, 19                         |
| 7       | 19 years/F                       | Restless leg syndrome, UI              | Intact    | UN                    | Improved                      | Intact                          | Intact                          | Intact                          | 4                               | None, 42                         |
| 8       | 56 years/F                       | Restless leg syndrome, unsteady gait   | Intact    | UN                    | Improved                      | Intact                          | Intact                          | Intact                          | 5                               | None, 18                         |
| 9       | 54 years/F                       | Lower back pain, unsteady gait         | Intact    | UN                    | Partially improved            | Intact                          | Intact                          | Intact                          | 3                               | None, 39                         |
| 10      | 18 years/M                       | UI, lower back and leg pain, unsteady gait, numbness | U, Ni; motor; Ni; S, Ni | UN                    | Partially improved            | Improved                        | Improved                        | Improved                        | 3                               | None, 25                         |
| 11      | 22 years/M                       | Lower back and leg pain, unsteady gait | Intact    | UN + DTM + LPM        | Partially improved            | Intact                          | Intact                          | Intact                          | 2                               | None, 98                         |
| 12      | 18 years/F                       | UR, lower back and leg pain, unsteady gait, numbness | U, Ni; motor; Ni; S, Ni | UN + DTM                    | Improved                      | Improved                        | Improved                        | Improved                        | 12                              | CSF fistula, 48                  |
| 13      | 19 years/M                       | Leg pain, unsteady gait                | Intact    | UN + DTM              | Improved                      | Intact                          | Intact                          | Intact                          | 6                               | None, 82                         |
| 14      | 23 years/F                       | Lower back pain, restless leg syndrome | Intact    | UN + DTM              | Improved                      | Intact                          | Intact                          | Intact                          | 3                               | None, 20                         |
| 15      | 19 years/M                       | Lower back pain, unsteady gait         | Intact    | UN + DTM              | Improved                      | Intact                          | Intact                          | Intact                          | 6                               | None, 62                         |
| 16      | 34 years/F                       | Lower back, leg pain, unsteady gait    | Intact    | UN + DTM              | Improved                      | Intact                          | Intact                          | Intact                          | 3                               | None, 24                         |
| 17      | 21 years/M                       | Lower back and leg pain, unsteady gait | Intact    | UN + DTM              | Partially improved            | Intact                          | Intact                          | Intact                          | 3                               | None, 21                         |
| 18      | 19 years/M                       | Lower back and leg pain, unsteady gait | U, Ni; motor; Ni; S, Ni | UN + DTM + LPM + 3VS       | Partially improved            | Improved                        | Improved                        | Improved                        | 6                               | None, 62                         |
| 19      | 21 years/F                       | UI, lower back and leg pain, numbness | U, Ni; motor; Ni; S, Ni | UN + DTM + MMS + LPM       | Unchanged                      | Unchanged                       | Unchanged                       | Unchanged                       | 10                              | CSF fistula, 98                  |
| 20      | 23 years/F                       | UI, lower back and leg pain, numbness | U, Ni; motor; Ni; S, Ni | UN + MMS + TV5            | Partially improved            | Improved                        | Partially improved              | Unchanged                       | 15                              | Wound infection, 71              |

Abbreviations: 3VS, third ventriculostomy; CSF, cerebrospinal fluid; DTM, diastematomyelia repair; F, female; LPM, lipoma excision; M, male; M; MMS, meningocele myelocele schisis repair; Ni, nonintact; S, sensory; TVS, terminal ventriculostomy; U, urinary; UI, urinary incontinence; UN, untethered; UR, urinary retention.
syringomyelia (C7–T10) and was diagnosed with a tethered cord accompanying the spinal mass. In the remaining 41 patients, syringomyelia was below the T6 level. In our study, 75% of the 54 patients with TCS had TS. Moreover, syringomyelia in 22, 36, and 40% of the patients were localized in the thoracic (T6–T12), lumbar (L1–S1), and thoracolumbar (T6–L5) regions, respectively.

### Treatment and Follow-up

When not accompanied by TS and other anomalies, TCS is typically treated by surgically cutting the tense filum terminale and liberating the cord. Congenital neurosurgical lesions (including meningocele, diastematomyelia, and intradural lipoma) are primarily removed surgically. However, researchers differ on treatment strategies for TS. In the past, syringomyelia was treated with chemotherapy and radiotherapy. The natural evolution of the tethered cord and coexisting syringomyelia are often interrelated, as the proper treatment of the tethered cord reduces the syringomyelic cavity. Syrinx drainage in TS is also controversial. Erkan et al. divided patients into two groups according to the surgical protocol as follows: (1) those who underwent the procedure to release the tethered cord (group I, n = 16), and (2) those in whom this procedure was combined with additional syrinx drainage (group II, n = 14). After a year of follow-up, patients in group II showed better clinical outcomes than those in group I (78 vs. 45%, respectively). Additionally, improvements in motor, sensory, and urinary deficits were observed in 50, 50, and 30% of patients in group I and in 78, 92, and 70% of patients in group II, respectively. In a study linking the tense cord syndrome to aortic coarctation, Hsu et al. found that the syrinx decreased after the liberalization of the tethered cord. Ng and Seow reported that in a patient whose tethered cord preceded lumbar syrinx formation—as demonstrated by serial radiographic imaging—the syrinx resolved after surgical untethering. In a 3.5-year follow-up study of 34 patients, Beaumont et al. examined patients in the following two groups: (1) the TCS group (TCS, n = 24) and (2) the TCS group with TS (TCS + TS, n = 10). The incidence of TS was 29%; only one patient underwent surgical drainage of the syrinx, and all the other patients underwent only tethered cord release. All patients who were asymptomatic preoperatively remained asymptomatic postoperatively. In the TCS + TS group, all patients either improved clinically after tethered cord release or improved and became asymptomatic. In the TCS group without TS, most patients improved or became asymptomatic. However, a very small number of these patients experienced no change or a worsening of symptoms. Patients who did not have a preoperative syrinx did not develop a syrinx postoperatively. In a limited number of patients, postoperative MRI demonstrated either no change or a reduction in the size of the syrinx.

Of the 42 patients with TCS included in our study, the tethered cord was released and the syrinx was drained in only one patient. In the postoperative follow-up, 37% of the 27 patients with syringomyelia in group 1 exhibited no changes in the size of the syringomyelia. The size of the syringomyelia decreased significantly and the syrinx was completely lost in 44.4 and 18.6% of the patients, respectively. Postoperative follow-up of syringomyelia patients in group 2 showed no changes in the size of the syringomyelia in eight patients and a significant decrease in size in 50% of the patients. The syrinx was completely lost in 37.5% of the patients (Tables 3 and 5).

Several researchers have focused on the relationship between TCS and TS. This study attached importance to the clinical symptoms of patients with TCS and evaluated the relationship between TCS and TS. However, our clinical findings suggest that compared with TS, accompanying congenital neurosurgical anomalies may be more important in TCS and aggravate the associated symptoms to a greater degree.

To prove this, we compared criteria such as symptoms, accompanying pathologies, and benefit from surgery in pediatric and adult patients. The clinical findings and symptoms were milder and more tolerable in adult patients than in pediatric patients. Moreover, the clinical findings and symptoms were milder and more tolerable in adult patients than in pediatric patients. Adult patients also achieved more favorable postoperative outcomes than pediatric patients.

### Limitations

This study has a few limitations. Though we included all documented TCS cases in our hospital over a 10-year period, the sample size was relatively small (n = 54). Moreover, the sample did not represent a wide geographical area, as all the patients were from Afyonkarahisar and surrounding locations.

This was a single-center study, and other institutes may follow different approaches. Moreover, the study was retrospective. Further prospective randomized studies with larger sample sizes and longer follow-ups are required to improve the generalizability of our results.

### Conclusion

When TCS is accompanied by TS and/or other congenital anomalies, the patient’s symptoms and clinical findings are more severe. Additionally, the possibility of postoperative recovery after surgery decreases. However, TCS alone is not accompanied by as many dramatic findings as previously thought. Although an important syndrome, TCS is a tolerable and highly curable syndrome. A multidisciplinary team consisting of a neurosurgeon, orthopaedist, urologist, radiologist, and physiotherapist should follow TCS patients closely to detect early clinical or radiological findings.

### Ethics Consideration

Approval for this study dated June 11, 2020, and numbered 2020/486 (2011-KAEK-2) was obtained from Afyonkarahisar University of Health Sciences Local Ethics Committee.

### Conflict of Interest

None declared.
Acknowledgment
We would like to thank Prof. Dr. Adem Aslan for sharing his vast experience with us.

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Table 7 Overall results comparison of operated patients with tethered cord syndrome in adult and pediatric groups

| Parameter                                         | Group 1                          | Group 2                          |
|---------------------------------------------------|----------------------------------|----------------------------------|
| Number of patients                                | 34                               | 20                               |
| Most common symptoms (%)                          | Urinary incontinence, 64.5%      | Lower back and leg pain, 70%     |
| Most common clinical findings                     | Bladder dysfunction, 64.5%       | Bladder dysfunction, 30%; motor deficit, 30% |
| Accompanying neurosurgical congenital anomalies (%)| 82%                              | 50%                              |
| Pathology of most common accompanying neurosurgical anomalies (%) | Syringomyelia, 79.4%; short thick file terminale, 73.5% | Short thick file terminale, 75%; diastematomyelia, 45% |
| accompanying syrinx (%)                           | 79.4%                            | 40%                              |
| Postoperative healing of syrinx (%)               | 63%                              | 87.5%                            |
| Postoperative improvement of symptoms (%)         | 88%                              | 95%                              |
| Results of postoperative clinical examination (%)  | Motor, 70%; sensory, 70%; urinary, 54.5% | Motor, 85.3%; sensory, 83.5%; urinary, 50% |