Multidisciplinary Combined Approach for Tethered Spinal Cord Syndrome: Radiology, Surgery and Physical Therapy

Arslanoglu A\textsuperscript{1}, Cirak B\textsuperscript{2}, Ilhan M\textsuperscript{3}, Aktekin N\textsuperscript{4}

Van Military Hospital, Department of Radiology\textsuperscript{1}, Van, Turkey
Pamukkale University, Departments of Neurosurgery\textsuperscript{2} and of Physical Therapy and Rehabilitation\textsuperscript{3}, Denizli, Turkey
Gazi University Faculty of Medicine, Department of Public Health\textsuperscript{4}, Ankara, Turkey

Abstract. The aim of this study is to formulate some objectives and to maintain the place of physical therapy in the management of tethered spinal cord syndrome by evaluating the clinical, radiological, and therapeutic approaches of our patients retrospectively. Eighteen patients with tethered spinal cord syndrome were analyzed retrospectively. All of the patients underwent surgical untethering and were put on a rehabilitation program after the surgery. Except three cases, all of the patients (83\%) improved their neurologic deficits at the end of the rehabilitation programme. Rehabilitation program, especially in the patients with motor deficit and bowel and bladder, dysfunction facilitates the outcome.

Key words: spinal dysraphism, rehabilitation, tethered spinal cord syndrome, untethering

Tethered spinal cord syndrome (TSCS) is a well-recognized entity involving the adult and pediatric populations [1,2]. TSCS is characterized by a low conus medullaris (below the L2 to L3 interspace). It occurs because of a tight filum terminale, adhesions from the myelomeningocele repair, other spinal surgical interventions, or in association with the lipomeningocele [1,3]. TSCS is more often encountered in children, but does also occur in adults [4]. Clinically it is characterized by any one or combination of the followings; motor and sensory dysfunction of the lower extremities, muscle atrophy, decreased or hyperactive reflexes, urinary and bowel incontinence, spastic gait, or orthopedic deformities [1-5]. In addition, the cutaneous lesions or subcutaneous lipomas in the lumbosacral region may be indirect signs of intraspinal pathology [1,3]. Although most of the reported cases of the TSCS are about lumbosacral tethering, some cervical and thoracic TSCS reports were also published [5,6]. Primary therapy for the cases having neurologic deficit is the surgical untethering of the spinal cord [5,7,8]. Eventhough surgical therapy is widely discussed the role of physical therapy and rehabilitation has not been discussed before. Rehabilitation of the TSCS patients is best attained by a coordinated team approach that is provided within a rehabilitation center. The team usually includes at least a psychologist, rehabilitation Nurse, physical therapist and some times orthist [9,10]. In this article we discussed the role of combined therapy of surgical untethering and physical therapy with radiologic evaluation for the TSCS.

Materials and Methods

Eighteen consecutive patients diagnosed clinically and radiologically as TSCS. Informed consent was obtained for each subject. All of them underwent a full neurologic, orthopedic, dermatologic, urologic examination, and radiologic evaluation including x-ray films, computerized tomography (CT), magnetic resonance imaging (MRI) of the spine. All of the patients were evaluated by a physical therapy and rehabilitation specialist (Physiatrist) and underwent surgical treatment according to their pathology. Filum has been freed, bony spurs have been resected, if the lipoma invaded or infiltrated the cord or the filum, it has been removed and meningocele or meningo(myelo)cele have been repaired.

The patients with TSCS have started rehabilitation as soon as being medically stable in the postoperative period. Proper positioning in bed was the utmost important in early care to prevent joint contractures and bed sores. Patients were avoided from bed sores by turning the patients sideways in every two hours. To prevent muscle imbalance, positioning of the patient was taught to parents. Foot positioning splints were used in each patient to prevent equinus deformity and pressure sores on the heel. During positioning of the patients in every two hours, legs were moved at each joint to full range; actively by patient and passively by the nurse to avoid from bed sores by turning the patients sideways in every two hours. To prevent muscle imbalance, positioning of the patient was taught to parents. Foot positioning splints were used in each patient to prevent equinus deformity and pressure sores on the heel. During positioning of the patients in every two hours, legs were moved at each joint to full range; actively by patient and passively by the nurse to prevent joint deformity. That was also important to decrease deep venous thrombosis risk. All of the patients received extension exercises besides muscle strengthening exercises to increase the muscle strength. The patients were trained to gain balance at gait and to stand still. Paravertebral and abdominal muscle strengthening exercises were done for body stability and balance. Patients were taken out of the bed to sit in a wheelchair or to walk alone or with the help of a physical therapist and nurse as soon as the vital signs are stable. Selection of the orthosis, splints and best fit shoes were done. Their uses were taught to parents and to the patients. All patients were evaluated by physiatrist, urologist, and orthopedist before discharge from neurosurgical clinic. One of the patients who had paresia has been hospitalized to the Physical Therapy and Rehabilitation Clinic for an extensive rehabilitation programme. They had two sessions of rehabilitation (each lasting one and a half hour), one in the

Correspondence to: Atilla Arslanoglu, M.D., e-mail: atilla02002@yahoo.com
morning and the other in the afternoon. Male patients who have urinary retention were trained for intermittent catheterization and they were encouraged to regular toilet visits to gain the regular bowel motility. Female patients were also trained for intermittent catheterization with the aid of a nurse. All of the patients were prescribed stool softeners, and the ones who had problem in bowel motility were prescribed rectal enema or suppository to stimulate anal sphincters. One of the family members was trained by the physical therapist and nurses all along the rehabilitation programme to help the patient during the outpatient period. For children, mother was trained for rehabilitation. Patients were controlled every 2 weeks by physical therapist and every month by physiatrist after discharge.

The distribution of Karnofsky Performance Scale (KPS) was compared between study groups by Wilcoxon signed-rank test. All analyses were performed with the SPSS statistical package version 10.0. The level of statistical significance was set to p < 0.05. The results are presented as mean standard deviations (sd).

**Results**

Thirty-three per cent (6) of the patients were under the age of 10. There were 7 (39%) male and 11 (61%) female patients. All except one of the patients have had cutaneous stigmata: 6 hypertrichosis, 7 hyperpigmentation, 4 sacral dimple. The ending point of the medulla was on L3 in 5 patients, L4 in 4 patients, L5 in 5 patients, and S1 in 4 patients (Fig. 1).

![Fig. 1. MRI (sagittal T1-weighted) of the spine revealed tethered cord and lipomyeloschisis. The ending point of the medulla is on L4 (left side-tuned picture).](image)

Most frequently encountered complaint was paresia at one or two lower extremities (12 patients), especially in the ages lower than 12 month paresia was the only symptom that the families had noticed. All of the adults have had back pain other than their complaints listed on the Table I. Two patients with meningocele have been reoperated with the diagnosis of retethering following the surgery for meningocele repair. All of the patients have started rehabilitation in the early postoperative period. But they continued the rehabilitation programme in outpatient basis following discharge from the neurosurgery clinic up to 2 to 6 (mean 3) months. They were followed up at 2 months interval up to 5-36 months (mean 15 months). At the time of discharge only 17% (3) of the patients had improvement in their preoperative neurologic deficits. At the end of rehabilitation programme, 83% (15) of the patients had improvement in their neurologic status. Only three patients (17%) had not any improvement or deterioration of their signs at the end of the physical therapy programme even though their programme has been extended up to 6 months postoperatively. Two of these three patients had invasion of the cord by lipoma and the resection of the lipoma was not total. Statistically significant differences were found between pre-KPS and last-KPS (p=0.001). As a a whole 83 % (15) patients have a mean 22.44 point increase in their KPS (Table 1).

**Discussion**

The TSCS is a congenital abnormality characterized by an abnormal low position of the conus medullaris due to a variety of tethering causes, which hamper the normal fetal ascensus medullae. Radiologically tethered cord means the fixation and limitation of the movements of the spinal cord due to different reasons. The pathogenesis of TSCS cannot be explained by an embryological disorder only. The start of the clinical symptoms and signs at an advanced age is indicative of secondary factors; most probably subsequent ischemia due to traction of the tethered cord during growth. Other reasons for the functional disturbances are direct damage to the cord by factors such as diastometamyelia and lipomyeloschisis [7,9-12]. According to Merx et al. [13] spinal cord tethering can be distinguished in three types generally:

- **Pathological filum terminale:** the simplest form; a short filum attaching the sacrum causes a low conus.
- **Intradural lipomas:** The lipoma of the filum, and conus, the lipomyeloschisis are in this group.
- **Diastometamyelia:** Fixation of the cord due to a bony fibrous or fibrocartilagenous septum extending from the ventral part of the spinal canal to the posterior splitting the cord.

The symptoms in the TSCS are generally evaluated in six groups; pain (back pain or radicular pain), sensorial abnormalities, motor deficits, bowel and bladder dysfunctions, orthopedic deformities including scoliosis and pes cavus deformities and cutaneous stig mata [14-17]. Most of the patients have more than one symptom. In childhood, most of the cases present with motor deficits, bowel and bladder dysfunctions including incontinence, enuresis, and constipation. But in adults; pain, sensorial and motor deficits are the leading symptoms [3,4,15,18,19]. Although most of the patients with TSCS have cutaneous stig mata, patients and families do not care of it so much. Diagnosis of the TSCS depends on the evaluation of the symptomatology together with neurologic and physical finding [13,18]. Direct x-ray studies are helpful if there is any bony abnormality such as scoliosis, hemivertebra, fusion defects of the vertebrae, spina bifida or a bony spur causing diastomatomyelia [17]. Myelographic findings of TSCS were first described in 1963 by Gryspeerdt [16] with myelographic imaging later supplemented by CT. In children myelography carried some difficulties, including puncture of the cord, encephalopathy caused by contrast administration, anesthesia to obtain cooperation of the child, and exposure to ionizing radiation. To avoid these risks ultrasonography has been used as an alternative diagnostic tool and it found usefull especially in the children under the age of 1 year of age since ossification of the posterior elements of the spine is incomplete. After the advent of MRI, it became the most important diagnostic tool in the evaluation of TSCS [20,21]. The inherent advantages of the MRI are that it is noninvasive and there is no ionizing radiation. Poor bone definition has always been described as...
TSCS is a neurologic disorder consisting of multisystem symptomatology related to the dermatology, urology, neurology, orthopedics, physical therapy and rehabilitation and neurosurgery clinics. Because of this, multisystem evaluation of the patient by each of the related clinic helps to delineate the extent of the deficits and guides the therapy and also helps to predict the postoperative improvement of the symptomatology. Besides multisystem evaluation of the patient, treatment must be completed by the adjuvant rehabilitation program starting from the preoperative state through the postoperative period. In our study we have observed that; to be a child, to have a lesser degree of motor deficit, and to get a good outpatient help from the family is a good prognostic indicator. In outpatient rehabilitation programme families hesitate more about the motor failures than the bowel incontinence or urinary retention. Because of this most of the patients could not improve in bowel or bladder function failures as it is in the preceding reports. Another thing we have observed is that the reason for the children does well in motor deficits is that, mothers care more about the deficit of their child more than the deficit of an adult patient.

The Karnofsky Performance Scale Index allows patients to be classified as to their functional impairment. This can be used to compare effectiveness of different therapies and to assess the prognosis in individual patients. The lower the Karnofsky score, the worse the survival for the most serious illnesses. According to our results, we found statistically significant differences between pre-KPS and last-KPS of our patients \( p=0.001 \). As a whole, 83% (15) of our patients have a mean 22.44 (from 62.22 to 86.66) point increase in their KPS (Table 1).

Postoperative rehabilitation must include passive exercises in the early postoperative period. Initiations of active exercises as soon as the vital signs are stable, and taking the patient out of the bed will help the neurologic deficits improve more than the surgeon expected. Rehabilitation program in our patients was very helpful especially when there was motor deficit.

Major limitation of MRI when compared to CT. MRI is very useful in visualizing the conus, assessing the thickness of the filum terminale, and identifying traction lesions in a patient clinically suspected of having TSCS \([13,16,17]\).

In the treatment of TSCS all of the articles to date were about the surgical untethering of the spinal cord \([7,8,18,22,25]\). In fact only surgical untethering is not enough every time in every patient. A physical therapy and rehabilitation program started just at the time of diagnosis and extended through the postoperative period will help the patients to improve their deficits more than expected.

The Karnofsky Performance Scale Index allows patients to be classified as to their functional impairment. This can be used to compare effectiveness of different therapies and to assess the prognosis in individual patients. The lower the Karnofsky score, the worse the survival for the most serious illnesses. According to our results, we found statistically significant differences between pre-KPS and last-KPS of our patients \( p=0.001 \). As a whole, 83% (15) of our patients have a mean 22.44 (from 62.22 to 86.66) point increase in their KPS (Table 1).

Postoperative rehabilitation must include passive exercises in the early postoperative period. Initiations of active exercises as soon as the vital signs are stable, and taking the patient out of the bed will help the neurologic deficits improve more than the surgeon expected. Rehabilitation program in our patients was very helpful especially when there was motor deficit.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age-Sex</th>
<th>Radiologic findings</th>
<th>Neurologic findings</th>
<th>Pre-KPS</th>
<th>Last-KPS</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5y-F</td>
<td>Syrinx, lipoma, tight filum</td>
<td>Slight monoparesia</td>
<td>60</td>
<td>80</td>
<td>Good</td>
</tr>
<tr>
<td>2</td>
<td>11y-M</td>
<td>Syrinx, increased subcut, fat</td>
<td>Incontinence</td>
<td>80</td>
<td>90</td>
<td>Good</td>
</tr>
<tr>
<td>3</td>
<td>7y-F</td>
<td>Scoliosis, sacral cyst, widened medullar canal</td>
<td>Foot deformity, distal monoparesia</td>
<td>60</td>
<td>90</td>
<td>Good</td>
</tr>
<tr>
<td>4</td>
<td>5y-M</td>
<td>Diastomatemayila, widened medullar canal</td>
<td>Paraparesia, incontinence</td>
<td>40</td>
<td>100</td>
<td>Good</td>
</tr>
<tr>
<td>5</td>
<td>4y-M</td>
<td>Meningocele</td>
<td>Sensoriel deficit</td>
<td>80</td>
<td>100</td>
<td>Good</td>
</tr>
<tr>
<td>6</td>
<td>4y-M</td>
<td>Diastomatemayila, tight filum, increased subcut, fat</td>
<td>Monoparesia, bilat. foot deformity</td>
<td>50</td>
<td>80</td>
<td>Good</td>
</tr>
<tr>
<td>7</td>
<td>4m-F</td>
<td>Lipomyelocele, lipoma invasion of the cord</td>
<td>Paraparesia</td>
<td>70</td>
<td>70</td>
<td>Same</td>
</tr>
<tr>
<td>8</td>
<td>4y-F</td>
<td>Meningocele, filar lipoma, widened canal, increased subcut, fat</td>
<td>Sensorial deficit, incontinence</td>
<td>70</td>
<td>90</td>
<td>Good</td>
</tr>
<tr>
<td>9</td>
<td>30y-M</td>
<td>Diastomatemayila, lipomyelocele, scoliosis</td>
<td>Sensoriel deficit</td>
<td>80</td>
<td>90</td>
<td>Good</td>
</tr>
<tr>
<td>10</td>
<td>2y-M</td>
<td>Syrinx, lipomyelocele, increased subcut, fat</td>
<td>Monoparesia</td>
<td>60</td>
<td>90</td>
<td>Good</td>
</tr>
<tr>
<td>11</td>
<td>7m-F</td>
<td>Lipomyelomeningocele, syrinx, increased subcut, fat</td>
<td>Paraparesia</td>
<td>60</td>
<td>60</td>
<td>Same</td>
</tr>
<tr>
<td>12</td>
<td>22y-F</td>
<td>Lipomyelocele, increased subcut, fat</td>
<td>Monoparesia</td>
<td>50</td>
<td>90</td>
<td>Good</td>
</tr>
<tr>
<td>13</td>
<td>21y-F</td>
<td>Lipomyelocele, increased subcut, fat</td>
<td>Sensorial deficit</td>
<td>80</td>
<td>90</td>
<td>Good</td>
</tr>
<tr>
<td>14</td>
<td>29y-F</td>
<td>Lipomyelocele, lipoma invasion of the cord</td>
<td>Paraparesia, incontinence</td>
<td>60</td>
<td>60</td>
<td>Same</td>
</tr>
<tr>
<td>15</td>
<td>4y-M</td>
<td>Lipomyelocele, increased subcut, fat</td>
<td>Monoparesia</td>
<td>60</td>
<td>100</td>
<td>Good</td>
</tr>
<tr>
<td>16</td>
<td>9m-F</td>
<td>Lipomyelocele, syrinx, increased subcut, fat, sacral cyst</td>
<td>Paraparesia</td>
<td>50</td>
<td>90</td>
<td>Good</td>
</tr>
<tr>
<td>17</td>
<td>12y-F</td>
<td>Sacral agenesis, sacral lipomyelocele</td>
<td>Distal paraparesia</td>
<td>60</td>
<td>90</td>
<td>Good</td>
</tr>
<tr>
<td>18</td>
<td>8m-F</td>
<td>Lipomyelocele, increased subcut, fat</td>
<td>Monoparesia</td>
<td>50</td>
<td>100</td>
<td>Good</td>
</tr>
</tbody>
</table>

(KPS= Karnofsky Performance Scale; Pre-KPS= Preoperative KPS; Last-KPS= KPS on last control; y= years; M= male; F= female.)

References