

Tethered Cord Syndrome in Childhood: Special Emphasis on the Surgical Technique and Review of the Literature with Our Experience

Çocukluk Çağı Gergin Omurilik Sendromu: Cerrahi Tekniğe Vurgu ve Tecrübemiz Eşliğinde Literatür İncelemesi

Ilker SOLMAZ¹, Yusuf IZCI¹, Baki ALBAYRAK¹, Eralp CETINALP¹, Cahit KURAL¹, Goksin SENGUL²,
Cuneyt GOCMEZ¹, Serhat PUSAT¹, Yusuf TUZUN²

¹Gulhane Military Medical Academy, Department of Neurosurgery, Ankara, Turkey

²Ataturk University, Faculty of Medicine, Department of Neurosurgery, Erzurum, Turkey

Correspondence address: Yusuf IZCI / E-mail: yusufizci@yahoo.com

ABSTRACT

AIM: Surgery is the treatment of choice for children who had tethered cord syndrome (TCS). However, a detailed technique for the release of spinal cord is not described yet. The aims of this study are to present our series of TCS in children and to focus on the details of surgical technique.

MATERIAL and METHODS: Forty-nine children with tethered cord syndrome underwent surgical treatment for the release of spinal cord between 2004 and 2009. The mean age was 4.6 years (2 days-13 years). Twenty (40.8%) patients were female and 29 (59.2%) were male. Among the 49 children, 41 (83.7%) had different spinal malformations and 8 (16.3%) had no associated lesion. Sectioning of the filum terminale, cutting the arachnoid and fibrous bands, protection of the rootlets and correction of the associated malformations was the standard surgical method to release the spinal cord.

RESULTS: Neurological improvement was observed in 4 (8.2%) patients, while the neurological status was unchanged in the others. Cerebrospinal fluid fistula was the main complication and was observed in 3 patients. No mortality or neurological deterioration was encountered.

CONCLUSION: Spinal cord release with appropriate technique seems to be beneficial in maintaining neurological functioning in children with TCS.

KEYWORDS: Tethered cord syndrome, Child, Surgical technique

ÖZ

AMAÇ: Gergin omurilik sendromu (GOS) olan çocuklarda cerrahi öncelikli tedavi yöntemidir. Bununla birlikte omuriliğin serbestleştirilmesi ile ilgili detaylı bir teknik tanımlanmamıştır. Bu çalışmanın amaçları, GOS'lu çocuklar ile ilgili serimizi sunmak ve cerrahi tekniğin detayları üzerine odaklanmaktır.

YÖNTEM ve GEREÇLER: Kırk dokuz GOS'lu çocuk 2004-2009 yılları arasında omurilik serbestleştirilmesi amacıyla cerrahi tedavi görmüştür. Ortalama yaş 4,6 yıldır (2 günlük-13 yıl). Yirmi (%40,8) hasta bayan, 29 (%59,2) hasta erkektir. Kırkdokuz çocuk içerisinde 41 (%83,7) tanesinde farklı spinal malformasyonlar vardı, 8 (%16,3) tanesinde eşlik eden bir lezyon yoktu. Filum terminalenin kesilmesi, araknoid ve fibröz bandların kesilmesi, köklerin korunması ve eşlik eden malformasyonların tamiri omuriliğin serbestleştirilmesinde standart cerrahi metod olmuştur.

BULGULAR: Nörolojik düzelme 4 hastada (%8,2) görülmüş, diğerlerinde nörolojik tablo değişmemiştir. Beyin-omurilik sıvısı fistülü en önemli komplikasyondur ve 3 hastada görülmüştür. Ölüm ve nörolojik bozulma görülmemiştir.

SONUÇ: Omuriliğin uygun teknik ile serbestleştirilmesi GOS'lu çocuklarda nörolojik fonksiyonların idamesi için faydalı olacaktır.

ANAHTAR SÖZCÜKLER: Gergin omurilik sendromu, Çocuk, Cerrahi teknik

INTRODUCTION

The tethered cord syndrome (TCS) results from the fixation of the spinal cord in the vertebral canal due to the adhesions either of congenital or acquired origin. Primary TCS is seen in patients with intradural lipomas, lipomyelomeningocele, split cord malformations, dermal sinus tract, and neuroenteric

cysts whereas acquired TCS is associated with repair surgery for spina bifida aperta including myelomeningocele, meningocele as well as spinal tumors, traumas, and infections all of which may cause arachoiditis and in turn traction on the spinal cord (7,8,14,15,17,18,19,20,24). This leads to mechanical torsion and ischemia in distal spinal cord including conus

medullaris (CM) thus motor-sensory deficits and associated urological and orthopedic complications are seen in TCS (31,32).

Virchow first used the term 'spina bifida occulta' to describe the lesions covered by skin and the term 'The horse maned woman' for hypertrichosis in 1875. Jones from the United Kingdom was the surgeon who first successfully released the tethered cord in 1891. Fuchs used the term 'myelodysplasia' for the clinical picture consisting of deep tendon reflex-sensorial disturbances, enuresis, and foot deformities in patients with spina bifida occulta. Recently, Yamada et al. reported that various neurological findings in TCS were resulting from caudal cord ischemia due to mechanical tension (31,32,33).

Since TCS occurs as a result of increased traction on spinal cord in caudal direction, treating underlying lesions or structures including arachnoid bands, primitive neural placode, thick or fibro-adipose FT, lipomyelomeningocele, diastometamyelia, and intradural lipomas with the release of tethered FT is the standard surgical treatment modality (1,5,9,11,12, 13,16,21,22,23,26,27,28,34).

In this study, we aimed to analyze the surgical outcomes of children who underwent surgery for TCS. We documented the symptoms and associated pathologies of the patients and focused on the details of surgical technique for each subgroup. The variations in surgical procedures were also pointed out, as well as the length and methods of follow-up.

MATERIAL and METHODS

Forty-nine children with TCS underwent surgical treatment for the release of the spinal cord in a period of 5 years. Twenty (40.8%) patients were female and 29 (59.2%) were male. The mean age was 55.67 months (ranged between 2-days and 13-years).

The symptoms were the cutaneous lesions in 30 patients (61.2%), urinary disturbances in 10 patients (20.4%), leg or foot weakness, numbness and/or spasticity in 9 patients (18.4%), foot deformity (pes cavus, claw toes) in 5 patients (10.2%), scoliosis in 5 patients (10.2%), and back pain in 5 patients (10.2%).

All patients underwent detailed radiological examination with magnetic resonance imaging (MRI) and computed

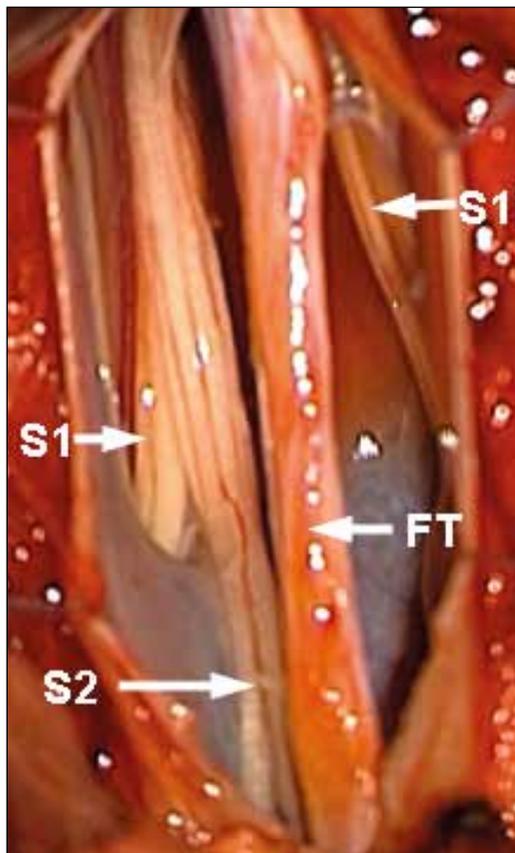


Figure 1: The intraoperative view of a patient with fatty filum terminale (FT). The filum was not thick, no vessel on the surface, but it was yellow in color. Vessels on the rootlets, especially on S1, are obvious. These vessels may confuse the surgeon. FT: Filum terminale, S1: S1 rootlets, S2: S2 rootlets.

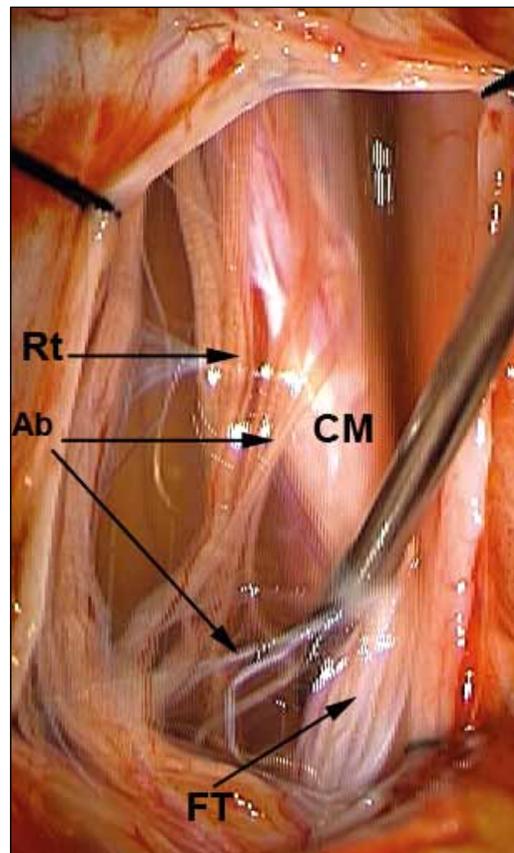


Figure 2: The intraoperative view of a patient with thick filum terminale. The thin arachnoid bands were attached to the spinal cord and caused tethering. Ab: Arachnoid bands, FT: Filum terminale, CM: Conus medullaris, Rt: Rootlets.

tomography. Among 49 children who underwent cord release, 16 (32.6%) had lipomeningocele, 14 (28.6%) had split cord malformation, 7 (14.3%) had dermal sinus tract, 2 (4.1%), had dermoid tumor and 2 (4.1%) had previous myelomeningocele operation. No associated lesion was found in 8 (16.3%) patients.

Sectioning of the FT, cutting the arachnoid and fibrous bands, releasing the spinal cord and correction of the associated malformation was the summary of surgical technique that was performed in all patients.

Surgical technique:

Patients are positioned prone under general anesthesia with supporting rolls on each side. The routine procedure is the L5 laminectomy, and additional partial or complete S1 laminectomy is added in order to expose the dura and then to identify FT. Laminotomy may also be selected for the exposure of the FT, especially in young children. In cases of spina bifida occulta, there is no need for laminectomy or laminotomy. Normally, the dural sac ends at the second sacral vertebra. In some circumstances, the spinal cord may continue until the S1 or S2 levels by giving some sacral rootlets, the laminectomy should be performed up to this level (Figure 1). The dura should be opened in the midline and tacked by four sutures bilaterally. Following the dural opening, FT, arachnoid bands, and rootlets should be first observed (Figure 2). Microsurgery with microinstruments should be used after the dural opening. FT is a fibrovascular tag containing a large vessel, which becomes smaller across its course in the lumbar subdural space. However, this vessel is not a reliable landmark FT because similar vessels can be found on the rootlets or no vessel may be seen on the FT. The most important issue at this time is the differentiation of the neural elements from extraneural structures. Rootlets and arachnoid bands are mostly confused by the surgeons (Figure 3). The rootlets at the sacral levels are directed to both sides and may be identified by their size and situation. The arachnoid bands are attached to the dura and rootlets (Figure 4). Both of them are thin and white in color. The intraoperative electrophysiological monitoring may be useful for safe surgery, but in the absence of this assortment, good neuroanatomical knowledge is required to preserve the neural structures. The rootlets are retracted laterally by microdissector in order to cut the arachnoid adhesions by microscissors and to expose the FT. In most cases, FT is thicker than normal, violet in color and is attached to the dura posteriorly in the midline leaving barely no free subarachnoid space for cerebrospinal fluid (CSF) passage. FT is coagulated and cut after the identification. The practical way to assess the degree of tethering pre-operatively is the immediate cranial movement of FT right after releasing. This sudden cranial movement of the superior-edge of FT resembles the movement of the string of a violin. In addition to sectioning the FT, all connective tissues attached to the caudal part of the spinal cord and CM should be released. FT and nerve roots must be free from the surrounding tissues. In cases of dermal sinus, the tracts may be attached to the FT or

other fibrous bands, therefore, these structures should also be cut in order to release the spinal cord. Another useful measure of preventing re-tethering would be performing dura-plasty in order to create a potent space allowing passage of CSF between lumbosacral rootlets and dura matter. The only way to prevent a recurrence in a detethered cord is to be certain that the neural elements remain free circumferentially with a patent CSF circulation.

MRI was performed in all patients at the end of third month after surgery to assess the level of the conus medullaris. The MRI was then repeated every year. Computed tomography was not used in follow-up. Postoperatively, all patients with neurological deficits received physical therapy.

The patients were followed-up neurologically and radiologically in postoperative period with a mean follow-up period of 2.3 years (range, 3 month to 4 years).

RESULTS

Neurological improvement was observed in 4 (44.4%) of 9 patients, while unchanged in the others. Urological disturbances improved in 2 (20%) of 10 patients. Back pain was improved in all patients. However, scoliosis and foot deformities were unchanged postoperatively. CSF fistula was the main complication and observed in 3 (6.1%) patients. Skin lesions such as dermal sinus tracts, dimples or subcutaneous masses were corrected during the surgery, but the hypertrichosis was unchanged following surgery. No mortality or neurological deterioration was recorded. The CM became free and observed as not attached to other structures in the follow-up imaging studies in all cases. Re-tethering was observed in 12 (24.5%) patients and most of them had lipomeningocele preoperatively. Re-operation was not planned for these patients because no neurological deterioration was observed.

Although this is a retrospective review of the patients' data with no control group, spinal cord release seems to be beneficial in maintaining neurological functioning in children with TCS.

DISCUSSION

From 2004 to 2009, a total of 49 children underwent surgery for untethering of the spinal cord. Of these, 41 (83.7%) patients had concomitant diagnosis of malformation, such as lipomeningocele, split cord or myelomeningocele. Surgery of TCS with such malformations showed some variations and the technique varies according to the underlying lesion. Not only the section of FT but also the dissection and cleaning of arachnoid bands around the rootlets are important for an effective detethering procedure. It is interesting to note that only 9 patients of 49 in the TCS patient population had neurological symptoms related to tethered cord and 4 of them showed clinical improvement after surgery. However, urinary functions were unchanged in most of the patients. MRI was the main radiological follow-up method in our series.

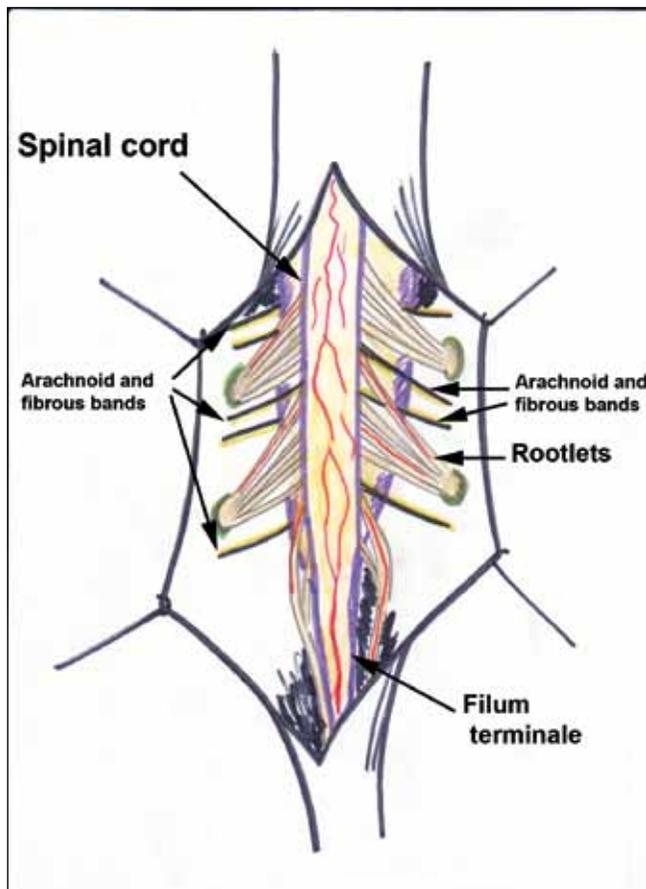


Figure 3: The line drawing shows spinal cord, filum terminale, rootlets and arachnoid and fibrous bands which are attached and tethered the spinal cord.

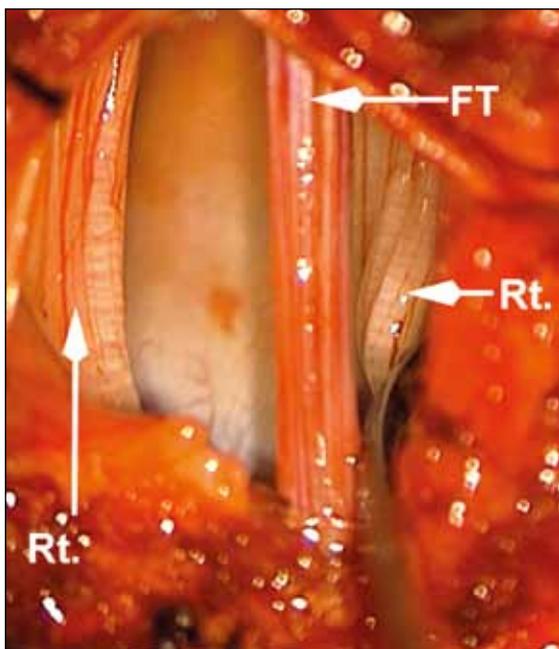


Figure 4: The intraoperative view of a patient with thick filum. Sacral rootlets are separated from the filum and directed toward the left and right sacral foramina. **FT:** Filum Terminale, **Rt:** Rootlet.

TCS is a rare intraspinal anomaly, caused by abnormal spinal cord fixation and resultant low-lying and immobile conus medullaris. The incidence of TCS is estimated to be 0.05 to 0.25 per 1000 births. It may result from many causes, including intraspinal lipoma, lipomyelomeningocele, diastematomyelia, spina bifida occulta, tight or thickened filum terminale, or scarring from previous myelomeningocele repair (3,4,10,15,17,18,25). In addition to pain, neurological dysfunction, and urological dysfunction, pediatric TCS is frequently associated with scoliosis (5,10,16,18,19,30). In accordance with the previous reports in the literature, lipomeningocele and split cord malformations together constitute 60% of all tethered cord cases in our series (2,10,11,12,13,14)

The syndrome is usually diagnosed in childhood and the presentation is variable and may be insidious. A TCS is suspected in patients with an unexpected spastic gait, a neurogenic bladder, bowel dysfunction, lower extremity weakness, scoliosis, or foot deformities. Although the presentation of TCS is well recognized and the course is usually progressive, it continues to pose significant diagnostic and surgical controversies (23,26,27,28,29). In our series, skin lesions were the most prominent symptoms, followed by neurological disturbances, urinary disorders, and scoliosis.

Before the management of pediatric patients with TCS, some questions should be answered. Should the treatment be aggressive? Should it be investigated and operated on as soon as possible? Is it reasonable to perform prophylactic surgery on patients who are neurologically intact? (14) In general, we perform aggressive treatment to children with TCS because they would have severe pain and advanced neurological deficits in the future. But the aggressive treatment could result in worsening of their neurological deficits. Pre-operative studies are necessary to plan the treatment in order to avoid operative morbidity or complications. However, if the children with TCS have associated malformations, impending neurological deficits and/or progression of their existing neurological deficits, it is reasonable and prudent to operate as soon as possible.

There are various controversial opinions about operating on patients who are neurologically intact for prophylactic purposes. However, if tethering pathology is confirmed through a radiological investigation, a prophylactic spinal cord release procedure seems reasonable and can be recommended if the patient is limited in exercise or work. There are many reports describing the surgery for tethered cord release in children (4,14,10,18,22,24,26,27,28). All of these studies summarized the surgical technique as repair of associated lesion and cutting the filum terminale. But some surgical details are missed in these reports. By the present study, we tried to describe the surgical technique as detailed as possible. Accurate localization of the level of CM preoperatively, identification and confirmation of the FT, preservation of the rootlets are the surgical pearls of our technique, while arachnoid and fibrous bands which attached to the FT are the pitfalls for surgeon. The surgical

plan for TCS must be tailored according to the preoperative radiological diagnosis and modified as necessary according to intraoperative pathological findings (17). Many differences in the mode of onset, clinical manifestations, and outcome exist between pediatric and adult patients with TCS and this warrants a more detailed analysis in children. The most problematic technical consideration in surgery for the release of the tethered cord is how to release the spinal cord, to preserve the neural elements and to rebuild the dural sac to maintain normal CSF circulation (14). Opening the dura, identification and section of the FT and dural closure is the routine technique to release the spinal cord in the literature (6,16,19,21,28). Although this technique is simple, it is not enough to reach a satisfactory spinal cord release. In addition, the FT can be identified by its typical dorsal midline location, its slightly bluish discoloration, its anteriorly located vessels, and the fat that often infiltrates it (6). But this is not always true. In our technique, we first focused on the cause of tethering and solved this problem. Then, we directed to the FT and rootlets to release the spinal cord. In cases of split cord malformations, there may be two FTs for both hemicords and these FTs should be identified before sectioning. An anteriorly located vessel may not be a reliable marker for FT and the FT may not be located midline in cases of split cord malformations. In addition, sectioning of FT is not enough to release the cord because arachnoid bands and fibrous stalks might also tether the spinal cord. Therefore, we suggested meticulous arachnoid dissection and preservation of neural elements. Identification and protection of the rootlets are crucial for maintaining the neural stability of the patient. The surgeon should identify these rootlets and follow them until the foramen, and differ from the bands either by electrophysiological techniques or microtechniques. The FT could be confirmed by neurophysiological monitoring, but we used microscope and microinstruments to identify the FT. After the confirmation of FT, it is transected distally and the end point of CM is closed with microsutures to decrease the scar formation and prevent the re-tethering.

The operative results for pain are most gratifying (14). Most of our patients had no pain after surgery, and sensorimotor deficits also improved favorably with surgical release of the conus. Many authors with experience in childhood TCS have commented on the poor operative results in terms of urinary dysfunction (14,19). In childhood TCS patients, sensory deficits, musculoskeletal atrophy, and voiding difficulty were improved after early untethering procedures. However, pain relief and sensorimotor deficits were improved after untethering in adults TCS patients. In the school-aged children, adolescents, and young adults with TCS, untethering is an appropriate procedure when there are neurological symptoms and signs and a confirmed tethering effect based on neuroimaging (21,29). Likewise, our results for bladder dysfunction were disappointing. Of the 49 patients with TCS, the pre-operative sensorimotor deficits improved after surgery in 4 patients (8.2%) while the others remained neurologically stable postoperatively.

CONCLUSIONS

We recommend surgical management for TCS in childhood. Intradural arachnoid bands and fibrous stalks may attach to the FT and CM, and confuse the surgeon. Not only the FT but also these bands should be cut and removed in order to preserve the CM and sacral rootlets. The clinical outcome is excellent if the tethering structures are completely released. Early diagnosis and adequate surgical release are the keys to a successful outcome in children with TCS.

REFERENCES

1. Ackerman LL, Menezes AH: Spinal congenital dermal sinuses: A 30-year experience. *Pediatrics* 112:641-647, 2003
2. Akay KM, Izci Y, Baysefer A: Dorsal bony septum: A split cord malformation variant. *Pediatr Neurosurg* 36:225-228, 2002
3. Akay KM, Izci Y, Baysefer A, Timurkaynak E: Split cord malformation in adults. *Neurosurg Rev* 27:99-105, 2004
4. Al-Holou WN, Muraszko KM, Garton HJ, Buchman SR, Maher CO: The outcome of tethered cord release in secondary and multiple repeat tethered cord syndrome. *J Neurosurg Pediatr* 4:28-36, 2009
5. Brooks BS, El Gammal T, Hartlage P, Beveridge W: Myelography of sacral agenesis. *AJNR Am J Neuroradiol* 2:319-323, 1981
6. Bui CJ, Tubbs RS, Oakes WJ: Tethered cord syndrome in children: A review. *Neurosurg Focus* 23:E2, 2007
7. Cheung AC, Kalkanis SN, Ogilvy CS: Paraplegia after tethered cord surgery: An uncommon combined anomaly of spinal arteriovenous fistula and sacral lipoma: Case report. *Neurosurgery* 57:E598, 2005
8. Daszkiewicz P, Barszcz S, Roszkowski M, Maryniak A: Tethered cord syndrome in children - impact of surgical treatment on functional neurological and urological outcome. *Neurol Neurochir Pol* 41:427-435, 2007
9. Hsu AR, Hou LC, Veeravagu A, Barnes PD, Huhn SL: Resolution of syringomyelia after release of tethered cord. *Surg Neurol* 72:657-661, 2009
10. Izci Y, Apaydin O, Ozdem T, Cerrahoglu K: The bony malformations of lumbosacral junction and sacrum: Case reports and literature review. *Tohoku J Exp Med*. 201:277-281, 2003
11. Izci Y, Taskaynatan MA: Management of lower back pain in young Turkish recruits. *Mil Med* 169:824-828, 2004
12. Izci Y, Gonul M, Gonul E: The diagnostic value of skin lesions in split cord malformations. *J Clin Neurosci* 14:860-863, 2007
13. Izci Y, Gurkanlar D, Gönül E: An unusual type of split cord malformation. *J Clin Neurosci* 14:383-386, 2007
14. Kang JK, Yoon KJ, Ha SS, Lee IW, Jeun SS, Kang SG: Surgical management and outcome of tethered cord syndrome in school-aged children, adolescents, and young adults. *J Korean Neurosurg Soc* 46:468-471, 2009
15. Kershner DE, Binhammer RT: Lumbar intrathecal ligaments. *Clin Anat* 15:82-87, 2002
16. Kothbauer KF, Novak K: Intraoperative monitoring for tethered cord surgery: An update. *Neurosurg Focus*. 16:E8, 2004

17. Lee GY, Paradiso G, Tator CH, Gentili F, Massicotte EM, Fehlings MG: Surgical management of tethered cord syndrome in adults: Indications, techniques, and long-term outcomes in 60 patients. *J Neurosurg Spine* 4:123-131, 2006
18. Matson DD, Woods RP, Campbell JB, Ingraham FD: Diastematomyelia (congenital clefts of the spinal cord); diagnosis and surgical treatment. *Pediatrics* 6:98-112, 1950
19. McLone DG, La Marca F: The tethered spinal cord: Diagnosis, significance, and management. *Semin Pediatr Neurol* 4: 192-208, 1997
20. Prabhu VC, Tomita T: Spinal cord tethering associated with amniotic band syndrome. *Pediatr Neurosurg* 31:177-182, 1999
21. Quiñones-Hinojosa A, Gadkary CA, Gulati M, von Koch CS, Lyon R, Weinstein PR, Yingling CD: Neurophysiological monitoring for safe surgical tethered cord syndrome release in adults. *Surg Neurol* 62:127-133, 2004
22. Radmanesh F, Nejat F, El Khashab M: Dermal sinus tract of the spine. *Childs Nerv Syst* 26:349-357, 2010
23. Rendeli C, Ausili E, Tabacco F, Focarelli B, Massimi L, Caldarelli M, Tamburrini G, Di Rocco C: Urodynamic evaluation in children with lipomeningocele: timing for neurosurgery, spinal cord tethering and follow-up. *J Urol* 177:2319-2324, 2007
24. Rogers HM, Long DM, Chou SN, French LA: Lipomas of the spinal cord and cauda equina. *J Neurosurg* 34:349-354, 1971
25. Souweidane MM, Drake J M: Retethering of sectioned fibrolipomatous filum terminales: Report of two cases. *Neurosurgery* 42:1390-1393, 1998
26. Talamonti G, D'Aliberti G, Collice M: Myelomeningocele: Long-term neurosurgical treatment and follow-up in 202 patients. *J Neurosurg* 107(5 Suppl):368-386, 2007
27. Tavafogni V, Ghandchi A, Hambrick GW Jr, Udverhelyi GB: Cutaneous signs of spinal dysraphism. Report of a patient with tail-like lipoma and review of 200 cases in the literature. *Arch Dermatol* 114:573-577, 1978
28. van der Meulen WD, Hoving EW, Staal-Schreinemacher A, Begeer JH: Analysis of different treatment modalities of tethered cord syndrome. *Childs Nerv Syst* 18:513-517, 2002
29. von Koch CS, Quinones-Hinojosa A, Gulati M, Lyon R, Peacock WJ, Yingling CD: Clinical outcome in children undergoing tethered cord release utilizing intraoperative neurophysiological monitoring. *Pediatr Neurosurg* 37:81-86, 2002
30. Yamada S, Zinke DE, Sanders D: Pathophysiology of "tethered cord syndrome". *J Neurosurg* 54:494-503, 1981
31. Yamada S, Iacono RP, Androde T, Mandybur G, Yamada BS: Pathophysiology of tethered cord syndrome. *Neurosurg Clin N Am* 6:311-323, 1995
32. Yamada S: Tethered cord syndrome in adults and children. *Neurol Res* 26: 717-718, 2004
33. Yamada S, Won DJ: What is the true tethered cord syndrome? *Childs Nerv Syst* 23: 371-375, 2007
34. Zide B, Constantini S, Epstein FJ: Prevention of recurrent tethered spinal cord. *Pediatr Neurosurg* 22:111-114, 1995