International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2020): 7.803

Surgical Management of Tethered Spinal Cord Syndrome in Adults - Our Experience

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Abstract: ***Background: Tethered spinal cord syndrome is a rare entity that usually presents with multiple neurological symptoms including lower extremity pain, backache, lower extremity muscle weakness and bowel bladder disturbance, but unlike children pain is a predominant clinical symptom. Materials and Methods: Case records of 10 patients aged ≥18 years who had undergone surgery with a diagnosis of TCS between 2013 and 2018 were reviewed. Patients who have underwent surgery earlier for tethered cord or for diastematomyelia/spinal dysraphism were excluded from the study. Results: Low backache was the most common presenting symptom. At the time of final follow - up, 7 (77.7%) patients had shown improvement in backache. Weakness improved by at least one grade in five (71.42%) patients. Bladder symptoms improved in two (40%) patients. Conclusion: In case of symptomatic patient with low - lying cord, surgical management with detethering of the spinal cord is an advisable option for better prognosis.***

Keywords: Adult tethered cord syndrome, Tethered cord, Detethering, Occult spinal dysraphism, Pain, Weakness, Bowel and bladder involvement

1. Introduction

STethered cord syndrome (TCS) is a complex clinicopathologic entity that is associated with varied but consistent symptomatology. [1], [2], [3] It was first described by Hoffman *et al*., in 1976 [4] and is due to relative failure of spinal cord ascent within the vertebral column during embryogenesis resulting in a "low - lying" conus medullaris. [5] Mechanical causes, as seen in radiology, known to be associated with tethered cord are thickened or lipomatous filum terminale, lipomas, epidermoid tumors, myelomeningocoeles, lipomyelomeningocoeles, and scar lesions that lack viscoelasticity and result in the fixation of the spinal cord. [6] It is postulated that TCS subsequently results from stretch - induced ischemia, depressed electrophysiological activity, and impairment of oxidative metabolism. [3], [5]

Most literature described this entity in children, with limited number of studies describing adult TCS. [3], [7] The true incidence of adult TCS is largely unknown. This unique and rare subgroup of patients presents with characteristic features of TCS, but unlike children, pain is a predominant clinical symptom. Surgical intervention is usually indicated based on an expected natural history of disease progression in the absence of treatment. Timely surgical intervention can prevent further deterioration and may even improve existing symptoms. In this background, we analyzed 10 consecutive patients aged ≥18 years, operated for tethered cord, and present our recommendations for treatment of such rare cases, based on our experience and review of literature.

2. Materials and Methods

The medical records of patients aged ≥18 years undergoing surgery with a diagnosis of TCS between 2013 and 2018, were retrospectively evaluated. Only patients (age ≥18 years) with conus level being below the L 1 - L 2 disk were included in the study. Patients who have underwent surgery earlier for tethered cord or for diastematomyelia/spinal dysraphism were excluded from the study.

The data was analyzed for type of presentation, radiological features, indications for surgery, and results in follow - up. Patients were followed up in outpatient department and telephonic interviews were also conducted. Mean follow - up period was 34 months (range 12 - 110 months).

A systematic search was performed using the Embase, Medline, and PubMed databases to identify case series about adult TCS using the key words 'adult tethered cord syndrome', 'tethered cord', and 'detethering and occult spinal dysraphism'. Searches were restricted to articles in English language and those published upto December 2016. Fourteen articles were thus found and analysis was done combining the observations of these studies, to find the rates of improvement in different symptoms.

Of the 10 patients included, 6 were men and 4 women. Patient's age ranged from 18 to 34 years [Table 1]. Typical cutaneous stigmata were present in only one patient, he had hyperpigmented patch over the lower back. Low backache was the most common presenting symptom in 9 patients, the average duration was 5.4 years (range 1 - 14 years). Other presenting symptoms are shown in [Table 2]. Bladder involvement was recorded in 5 patients. No patients had non - healing ulcers

Table 1

Features	Numbers	Percentage
Demographic profile		
Mean age	21.0years (SD - 4.85)	
Males	6	60
Female	4	40
Presentation		
Low back ache	9	90
Paresthesia	1	10
Numbness	2	20
Weakness of lower limbs	5	50
Bladder incontinence	5	50
Low – lying conus	10	100

Table 2

	No. of patients	Improved (%)	No change	Deteriorated
Backache	9	7 (77.7)	2	0
Motor weakness	5	3 (60)	1	1
Bladder symptoms	5	2 (40)	6	0

MRI showed evidence of low - lying (below L2) conus in all 10 patients [Figure 1], [Figure 2] and [Figure 3] and two patients had lipoma of the cord. There was no evidence of bony spur or fibrous septa causing tethering [Table 1]. At the time of final follow - up, 7 (77.7%) patients had shown improvement in backache. Weakness improved by at least one grade in five patients (71.42%). Bladder symptoms improved in two patients (40%) [Table 2]

FIG 1 - 4, show T2 sagital image, axial image and intraoperative pics of low lying tethered cord attached to intradural lipoma from S3 - S5

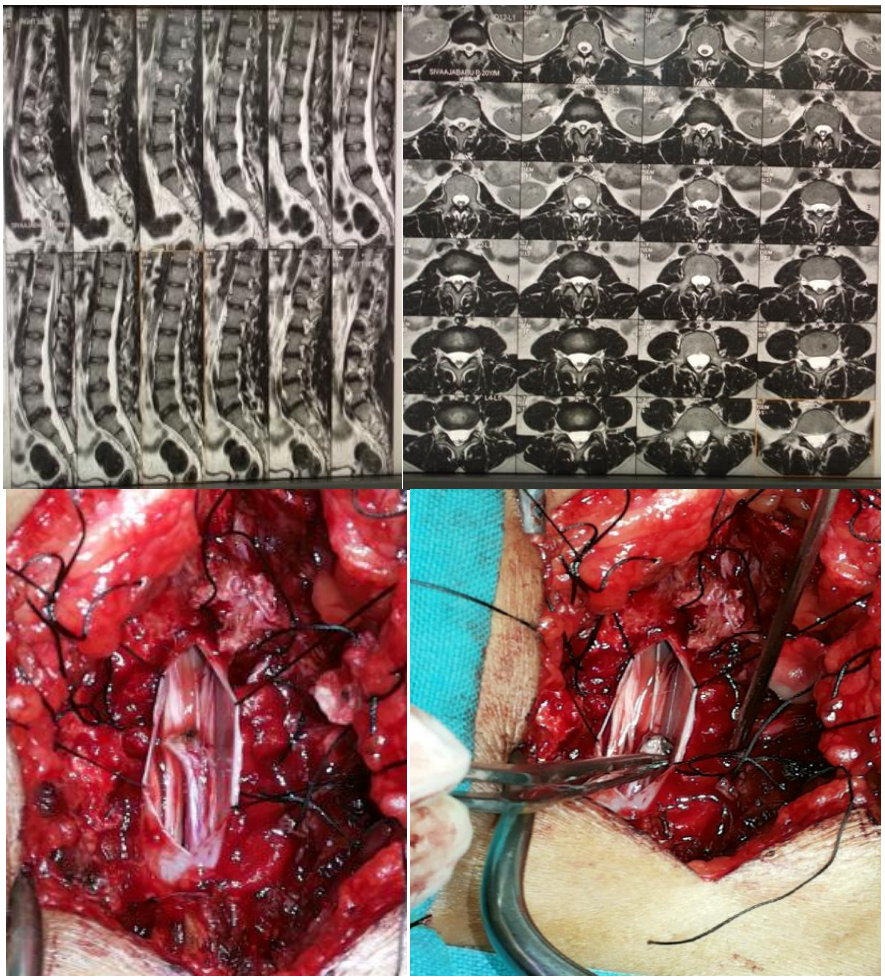
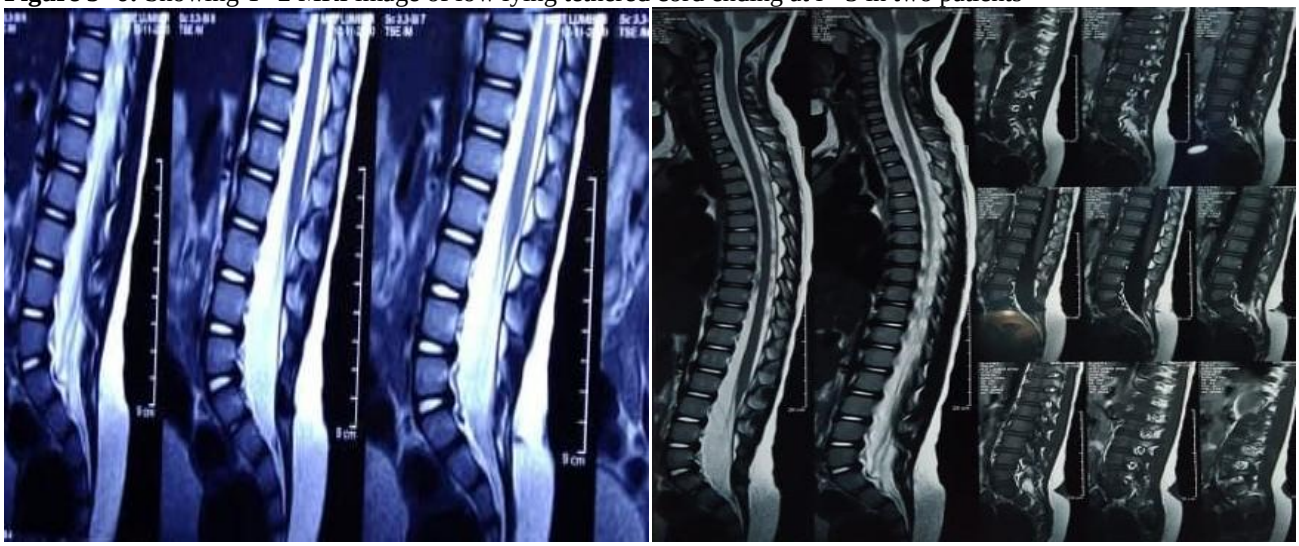


Figure 5- 6: Showing T- 2 MRI image of low lying tethered cord ending at l - 3 in two patients



Twelve articles were reviewed and their findings are presented in [Table 3]. Maximum improvement rates were seen in backache, while least in bladder disturbances [Table 4].

Table 3

Authors and year	Follow-up	Clinical outcome					Complications
		Back pain	Leg pain	Weakness	Altered sensation	Bladder dysfunction	
Garces-Ambrossi *et al.*[8] (2009)	18 months	81% showed significant improvement		69% better	79% better	47% better	CSF leak 7%, surgical site infection 10%
Romagna *et al.*[9] 2013	Mean follow-up 16 months	77% better, rest same	47% better, rest same	3.7% better	7.4% better	No improvement	CSF fistula 11%
Pang and Wilberger[7] 1982	6 months-11 year	100% better		87% better		38% better, 54% same, 8% worse	4% meningitis/death
Iskandar *et al.*[10] 1998	7 days-7 years (mean-4 years, median 2 years)	81% better, 15% same, 4% worse		48% better, 33% same, 4% worse		61% better, 33% same, 6% worse	15% subcutaneous CSF collection, 3% CSF leakage
Yamada and Lonser[11] (2000)	2-19 years in patients with spinal dysraphism, 2-24 years in those without dysraphism	Patients with spinal dysraphism-100% better, others-most better	Patients with spinal dysraphism-100% better, others-100% better	Patients with spinal dysraphism-'modest' improvement, others-'marked' improvement	Patients with spinal dysraphism-'modest' improvement	8 incontinent patients 'stable', 56% normalized	Not described
Gupta *et al.*[12] 1999	Not described	89% better		Not described	Not described	33% better	Not described
Akay *et al.*[13] (2000)	12-42 months (mean-18.7 months)	100%	100%	Not reported	Not reported	Sphincter problem persisted in one patient	0% neurological complication
Huttmann *et al.*[14] 2001	6 months-12 years (mean 8 years)	91% better, 4% same, 4% worse		26% better, 69% stable, 6% worse (at 6 m)		1 patient worse	19% subcutaneous CSF collection, 4% wound infection, 2% extradural hematoma, 5% revision CSF leakage and wound infection, 2% permanent neurological deficit
van Leeuwen *et al.*[15] 2001	51 of 57 patients at 6 months	56% better, 39% same, 6% worse			10% better, 86% same, 9% worse	14% better, 86% same, 0% worse (bladder/bowel)	12% CSF leakage, 2% 'difficult wound healing'
	40 of 57 patients at 2 years	48% better, 45% same, 8% worse			15% better, 80% same, 5% worse	15% better, 83% same, 3% worse (bladder/bowel)	
Phi *et al.*[16] 2004	3-123 months (mean 43 months)	14% better, 14% worse, 72% same	11% better, 11% worse, 78% same		11% better 89% same		31% overall, 6% reoperation for CSF leakage, 13% new feet hypoesthesia, 6% new back pain, 6% big toe weakness
Quinones Hinojosa *et al.*[17] 2004	1.5 months-3.2 years (mean 6.8 months, median 3 months)	62.5% better, 25% same, 12.5% worse		50% better, 12.5% same, 37.5% worse	50% better, 30% same, 20% worse	60% better, 30% same, 10% worse	0% worse
Lee *et al.*[2] 2006	Median follow- up of 27.5 months (range 1-125 months, mean 41.5 months)	78% better, 19% same, 3% worse	83% better, 10% same, 7% worse	64% better, 27% same, 9% worse	45% better, 50% same, 5% worse	50% better, 45% same, 5% worse	15% CSF leakage, 5% wound infection, meningitis in 1 patient, urinary tract infection in 1 patient, worsened foot weakness in 1 patient

CSF - Cerebrospinal fluid

Table 4

Symptom	Total number of patients	Number of patients who improved (O/o)
Backpain	290	233 (80.3)
Weakness	236	153 (64.8)
Altered sensation	191	110 (57.6)
Bladder dysfunction	274	112 (40.9)

3. Discussion

In normal fetal development before the 15 - mm stage, the cord extends to the lower end of the sacrum. [7] Thereafter, the vertebral column lengthens caudad more rapidly than the spinal cord, and the conus ascends the canal to reach the L - 3 level at about 30 weeks of gestation. At the 30 - mm stage, the terminal conus formed from the caudal cell mass regresses to become the filum terminale, which normally

remains thin and slightly redundant to allow for ascent of the conus. If at this stage the conus is trapped at a low level by an abnormally stout and short filum, a lipoma, a sagittal septum, fibrous adhesions, or the fibroneural stalk of an occult myelomeningocele, normal ascent would be arrested.

After the excellent work that Yamada *et al*. [3], [6], [18], [19], [20] have performed over the last 20 years, it seems that in both human and experimental animal models of TCS there is dysfunction in neuronal mitochondrial terminal oxidase in the electron transport chain. Furthermore, it has been shown by Schneider *et al*., using laser Doppler flowmetry, that there was a relative decrease in spinal cord blood flow *in vivo* in patients with pediatric TCS and following cord release, blood flow normalized to control levels. [21] The pathophysiology of adult - onset TCS is not very clear. It is still unclear that why do some patients with TCS remain asymptomatic till adulthood. Pang and Wilberger [7] postulated that the degree of traction on the conus is what determines the age of symptom onset.

Adult TCS is now being increasingly encountered by neurosurgeons of developing world and is perhaps more common than earlier thought. Increase in incidence can be attributed to easy availability of imaging modality like MRI than it was 2 decades. The incidence of occult spinal dysraphism (OSD) is unknown and, it is possible that some patients may remain asymptomatic and a diagnosis is never made, while in others with the congenital syndrome, may develop progressive deficits in adulthood, which can be caused by traction on the spinal cord due to sudden movement. [7], [18], [22], [23], [24] These may include bending movements, the lithotomy position during childbirth, movement occurring during road traffic accidents, and others.

The inclusion criteria for adult TCS appear to differ between reports, highlighting the lack of consensus. Gupta *et al*., [12] excluded patients with post myelomeningocele repair, although they have acknowledged that patients with this type of lesion formed the largest group of adult patients presenting with TCS. On the other hand, Phi *et al.*, [16] have excluded from their review patients who had underwent meningomyelocele repair or lipoma removal earlier, but included two cases of "retethering" referred from other institutions. In contrast, patients with post myelomeningocele repair have been included in other studies of adult TCS. But in our study we did not include patients, who were operated for spina bifida in childhood or had any structural lesion in conus, as seen on imaging. In our series, we included patients in whom the conus was low - lying and no identifiable cause (other than thickened filum) was found.

The clinical presentation of our patients was similar to those reported in previous studies. [25], [26], [27] Similarities and differences exist between the adult and pediatric populations, and in many regards certain disparities between the two may be attributed to the young child's inability to communicate symptoms such as pain, sensory changes, urinary urgency, or incomplete voiding. [28] Adult patients predominantly present with pain, which is one of the major difference between adult and pediatric TCS.

The issue of surgery in newly diagnosed adults with OSD is still controversial. [29] It is commonly believed that children who have a congenital tethered cord benefit from surgical detethering because it prevents neurological deterioration. [30] The same rationale is extended to adult TCS as well. It is believed that patients with primary tethered cord will sooner or later experience worsening of neurological deficits if they do not undergo spinal cord untethering. [24], [31], [32] Better postoperative outcomes have been reported when this condition is treated promptly after the appearance of deficits. [33], [34] This is supported by the good results following surgery in adult patients.

Other approach may be close follow - up including monitoring of motor power and urodynamic studies. Klekamp [23] studied 85 adult TCS patients and concluded that surgery in adult patients with a TCS should be reserved for those with symptoms and a conservative approach is warranted in adult patients without neurological deficits. As of now, there is no conclusive evidence in literature favoring either surgery or conservative management of adults with OSD without any deficit.

Intraoperative monitoring was not used in our study due to its unavailability. However, there are reports that intraoperative electromyography may be useful. Pang and Wilberger and Haro *et al.*, [7], [35] have argued that intraoperative spinal cord monitoring is indispensable to safe operation because functional neural elements are often embedded within the lipomatous tissues. In absence of EMG monitoring, our surgical strategy was to detether filum at the lowest possible and preferably L_5 - S_1 level. Before detethering, filum was always inspected and all roots adherent to it were dissected free and then detethering was achieved. In none of our patients fresh neurological deficits appeared. However, in one of the patients we encountered increase in weakness. Here a small root adherent to filum was accidentally sectioned.

Newly acquired deficits in adults may be reversible following timely surgery. [24] Not only the developed deficits are reversed, but potential complications in the coming years are obviated by the surgery. Yamada *et al*., [19] has reported improvement in pain and motor function in all patients in their series, but they only included those with a tethered cord caused by a fibrous or lipomatous filum terminale. Pang and Wilberger [7] have stated that a majority of patients were free of pain following surgery. Iskandar *et al.*, [10] found improvement in pain status in over 80% of their patients. In our series, more than 80% of patients had improvement in backache and 78% had improvement in motor weakness.

The surgical complication rate is generally low. However, few studies have reported that it is slightly higher than that in children with the same disease, if one has to compare the data with those of pediatric series. [36] In our study, we did not encounter any immediate postoperative wound site - related complication. Long - term results were encouraging, justifying the need of surgery.

There is risk of retethering after detethering. However, the incidence of retethering and the indications for repeat

surgery remain controversial. In the pediatric age group, Archibeck *et al.,* [37] have suggested that retethering is relatively common with 52% of patients requiring revision surgery by the age of 5 years in their series. In contrast, the incidence of retethering in adults seems to be significantly lower. Huttmann *et al.,* [14] has reported that over a mean follow - up period of 8 years, only 16% of patients required repeat detethering surgery. In the current study, no patient required repeated detethering procedures during the follow - up period.

In case of symptomatic patient with low - lying cord, detethering is an advisable option. However, in asymptomatic person with incidentally diagnosed low - lying cord there is lack of unanimity in opting for surgery or conservative follow - up [Figure 4]. Our philosophy is to explain the patient that option of surgery exists in contrast to conservative follow - up. In case of no radiological abnormality being detected and if the patient is not an active athlete, of child - bearing age, and does not have any other risk factors, an option of conservative follow can be exercised. During follow - up we monitor neurological status (sensory, motor, and bowel/bladder), urodynamic studies and MRI are repeated at regular intervals. In case of any new deficit or finding on investigations, surgery is advocated. However, we at times advocate surgery at the first presentation to the poor patients who are unable to afford the cost of repeat radiological/urological and clinical follow - up costs. However, a prospective long - term outcome study for such patients is warranted to provide a definitive guideline.

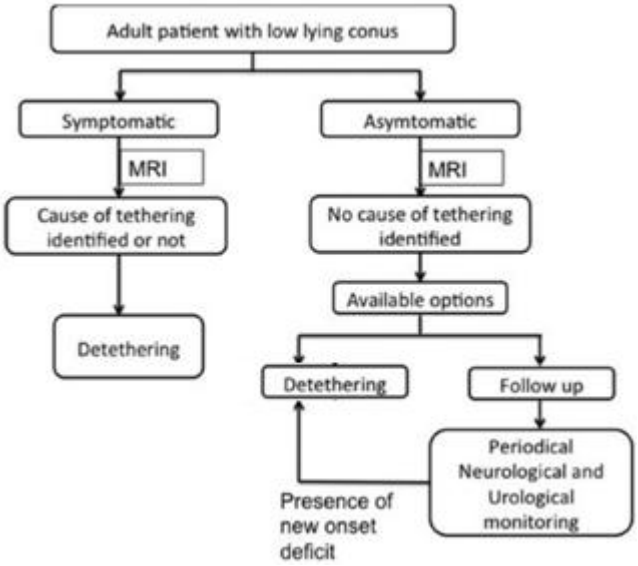


Figure 7: Our Management Algorithm for Adult Tethered CORD Syndrome

Use of electrophysiological monitoring during detethering procedure has become a standard of care. We are not having this facility in our hospital and so it was not used. Moreover, all patients were not subjected to regular pre - and postoperative urodynamic evaluation. It is due to prohibitive cost of this investigation for some of the extremely poor patients and its non- availability in some parts of the country 10 years back. However, nowadays it is an integral part of our treatment protocol for such patients. We lacked data of patients treated conservatively in our outpatient department. Prospective follow - up of these patients as cohorts can provide us with comparative results regarding surgery versus conservative treatment.

Pain is the most common symptom in adults with TCS. These patients are likely to benefit from surgery. The issue of surgery in asymptomatic adults with low - lying conus is controversial. On the basis of our experience, we have proposed our guidelines.

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