

Original Article

## Surgical Outcome of Adolescents and Young Children with Tethered Cord Syndrome: A Retrospective Analysis

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### ABSTRACT

**Objective:** In Spinal dysraphism, neural tube defect is one of the leading causes of congenital central nervous system disorders and contributes to a high proportion of disability and mortality. This study aimed to evaluate the outcome of the detethering of tethered cord syndrome.

**Materials & Methods:** The study was conducted at the Neurosurgery Department of Hayatabad Medical Complex Peshawar. A total of 110 patients were included in our study. Clinical examination was recorded for back swelling, dermal stigmata, scars, lower limb neurological function, sensory deficits, reflexes. Also, look for any deformities like kyphoscoliosis or limb deformities and the presence/absence of neuropathic ulcers. Imaging studies were recorded for diagnosis, the level of spinal tethering, presence/absence of syrinx, and spine deformities. During the surgical operation, the diagnosis of the lesion was confirmed and the interventional procedure was recorded.

**Results:** One hundred ten patients were admitted during the study period with 57 (52.2%) females and 53 (47.8%) males with a mean age of 9.3 years (SD  $\pm$  4.3). Most patients had presented with pain in lower limbs (60.9%) and backache was (34.8%). The most common operative complication was CSF leak in 29 patients (26.1%) while wound infection occurred in 14 (13%) cases.

**Conclusion:** Tethered cord syndrome due to occult spinal dysraphism is a rare disease with significant neurologic consequences for the younger generation. Early clinical diagnosis and good surgical treatment could prevent the development of these complications.

**Keywords:** Spinal Dysraphism, Spina bifida Occulta, Tethered Cord Syndrome.

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### INTRODUCTION

The tethered spinal cord was described as characterized by low lying conus and thickening of the filum terminale. This condition was first described by Hoffman in a landmark paper which described the related characteristics of the

disease and its outcome to surgical release of the tethering.<sup>1</sup> Occult spinal dysraphism is a well-known form of neural tube defects with a relatively rare occurrence and late clinical presentation. It was described by Virchow in 1875 with a recorded first instance of surgical correction performed by Jones in 1891.<sup>2</sup> Similarly tethered cord syndrome was described as lower spinal cord dysfunction due to constant traction upon the conus medullaris.<sup>3</sup>

Multiple aetiological factors have been described which are associated with the development of neural tube defects like, genetic factors, dietary deficiencies, socioeconomic, environmental, and maternal factors<sup>13,15</sup>. Open spinal dysraphism accounts for up to 95% of all neural tube defects while occult spinal dysraphism accounts for the remaining 5% of the lesions. Low antenatal obstetric coverage of childbearing women in our society and very few clinical manifestations at birth has delayed the diagnosis of occult spinal dysraphism.<sup>4,5,6</sup>

Occult spinal dysraphism lesions have been further described into different varieties which include meningocele, lipomyelomeningocele, diplomyelia or diastematomyelia, thickening of filum terminale, post-operative intradural adhesions, and adherence of the placode following myelomeningocele repair.<sup>7</sup> All the above mention varieties of the lesions require open untethering of the cord to improve neurologic manifestations or stop its progression.<sup>8,9</sup>

Over the three decades, much advancement has been achieved regarding the prevention, earlier diagnosis, and treatment of this disorder, to stop the progression of neurologic deterioration. Antenatal dietary supplementation with folic acid has reduced the incidence globally, frequent clinical visits to the obstetric clinics and increasing expertise in antenatal ultrasound, availability of high-frequency ultrasound, antenatal and early post-natal MRI, dynamic or phase-contrast MRI has helped in earlier

diagnosis, and improvement in micro-neurosurgical techniques has led to improved outcome for these patients.<sup>10-14</sup>

However, the rarity of this clinical condition warrants repeats clinical research to improve upon the already achieved developments and as a reminder for the neurosurgical and spine surgical community to remain vigilant regarding the earliest possible diagnosis and good surgical management. In a developing country like Pakistan, the high prevalence of these disorders, low availability of obstetric, advanced diagnostic, and tertiary care neurosurgical facilities, and decreased awareness among the primary health care physicians warrants widely available current research about this entity of disorders.<sup>4,5,15</sup>

Our main focus in this study is to know about surgical outcomes of tethered cord syndrome present to our department Hayatabad medical complex Peshawar in mention duration. Which will be helpful to prevent disability.

## MATERIALS & METHODS

This was a retrospective study which was conducted at the department of neurosurgery Hayatabad medical complex Peshawar from 1<sup>st</sup> January 2013 to 31<sup>st</sup> December 2018. The data of the patients was included as anonymous and used with the permission of the administration.

### Inclusion Criteria

We included a total of 110 patients both male and female patients from the age range 0 to 18 years of children diagnosed case of tethered cord syndrome due to spinal dysraphism in the mentioned period of duration. The institute ethical committee for research evaluation was obtained for approval of the proposed study.

### Exclusion Criteria

Spina bifida cystica and previously operated patients were excluded from the study.

## Data Collection

The data was collected on a predesigned proforma. All cases who presented to the outpatient department were diagnosed using clinical methods and diagnostic imaging studies such as MRI & CT scans of the spine were performed. Data was collected about the demographics, clinical symptoms, duration of symptoms, and developmental history.

## Clinical Management

Clinical examination was recorded for back swelling, dermal stigmata, scars, lower limb neurological function, sensory deficits, reflexes. Also, look for any deformities like kyphoscoliosis or limb deformities and the presence/absence of neuropathic ulcers.

Imaging studies were recorded for diagnosis, the level of spinal tethering, presence/absence of syrinx, and spine deformities. During the surgical operation, the diagnosis of the lesion was confirmed and the interventional procedure was recorded. The data was entered and analyzed using SPSS version 20.0 and is presented using charts, tables, and graphs.

## Follow-up

After surgery, the patients were followed up to the date of discharge. During the postoperative course, the development of complications, any additional procedures performed for complication management, and the total length of stay were recorded.

## RESULTS

We have analyzed a total of 110 patients having tethered cord syndrome and got the following results.

## Age and Gender Distribution

The mean age of patients was 9.3 years (SD  $\pm$  4.3). One hundred ten patients with 57 (52.2%) females and 53 (47.8%) males were included over the five years.

## Clinical Presentations

The most common presenting complaints were lower limbs pain (60.9%), backache (34.8%), symptoms of urinary tract dysfunction (incontinence, retention, difficulty voiding) (30.4%), and weakness of the lower limbs with gait problems (26.1%). A majority of patients also complained of the presence of back swelling which was present since birth as shown in Table 1.

**Table 1:** Clinical Symptoms.

Variables	Frequency	Percentage (%)
Lower limbs pain	67	60.9%
Backache	38	34.8%
Urinary symptoms	33	30.4%
Weakness Lower limbs & gait problems	29	26.1%

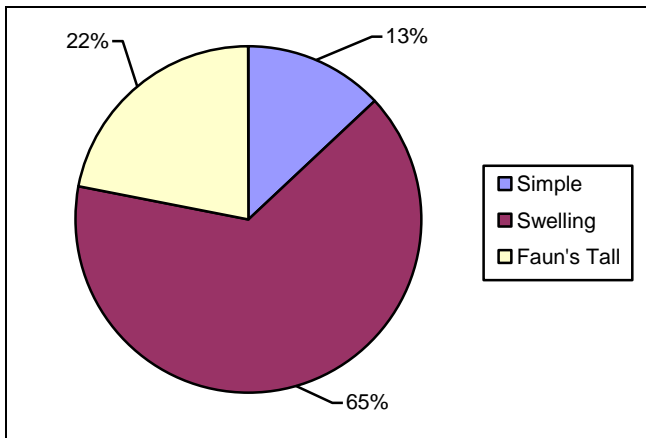
## Physical Findings

On clinical examination the findings included back swelling (65.2%), wasting of the lower limbs (30.4%), ulceration of the feet (21.7%), faun's hairy tail (21.7%), skin dimpling over the lumbosacral area, or the swelling (13%) as shown in Figure (1) and decreased power grade of the lower limbs in these patients. Kyphoscoliosis was present in only two (8.7%) patients. The clinical diagnoses included lipomyelomeningocele (LMMC) (34.8%), diplomyelia (21.7%), meningocele (21.7%), filum terminale thickening (4.3%), and congenital dermal sinus (8.7%) as given in Table 2 and represent in Figure 2.

**Table 2:** Physical Findings.

Variables	Frequency	Percentage
Lower Limbs Power		
0	4	4.3%
1	4	4.3%
4	24	21.7%
5	76	69.6%
Feet ulcers	24	21.7%
Wasting	33	30.4%
Faun's tail	24	21.7%
Skin Dimpling	14	13.0%
Swelling	72	65.2%
Kyphoscoliosis	9	8.7%
Dermal Sinus	9	8.7%
Diplomyelia	24	21.7%
Filum Terminale fibrosis	5	4.3%
Lipomyelomeningocele	38	34.8%

**Dermal Signs**

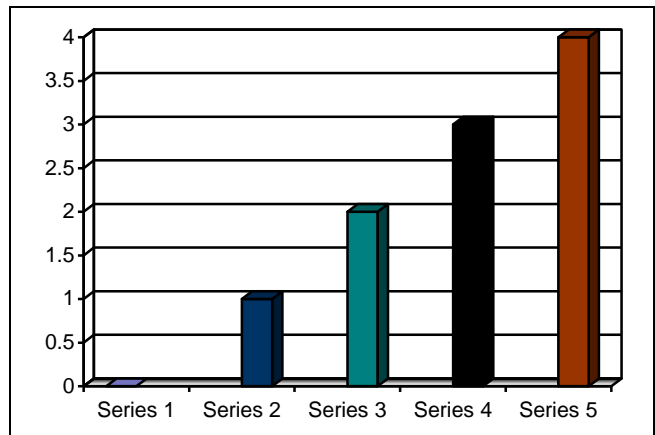


**Figure 1:** Dermal Signs.

### Postoperative Complication

Postoperative complications included cerebrospinal fluid (CSF) leak in 13 (12%) patients and infection of the wound in (5.5%) patients. Only two patients underwent reoperation for Dural closure and two patients were treated for CSF leak through the insertion of a lumbar drain. The mean length of stay for the patients was found to be 5.7 days (SD ± 3.0).

**Diagnoses**



**Figure 2:** Diagnosis.

**Table 3:** Postoperative Complications.

Complication	Frequency	Percentage
CSF leak	29	26.1%
Wound Infection	14	13.0%
<b>Additional Procedures</b>	-	-
Lumbar Drainage	9	8.7%
Reoperation/dura repair	9	8.7%

### DISCUSSION

Spina bifida occulta is a rare entity in the spectrum of neural tube malformation which may accompany tethering of the spinal cord causing a myriad array of symptoms and signs. The tethering produces symptoms by various mechanisms, among which ischemic insults to the spinal cord and nerve roots are one of the most favored explanations. The incidence of tethered cord syndrome is reported to be in the range of 0.05 to 0.25 per 1000 births.<sup>16</sup> The clinical features of occult spinal dysraphism and associated tethered cord syndrome are insidious and in some instances, they may be absent until the neurological sequelae become apparent. The symptoms of the tethered cord syndrome may sometimes appear later in adolescence or adult life and it has been shown that untethering by surgery has very satisfactory results in terms of neurological improvement.<sup>16-18</sup> The insidious

nature of the tethering of the spinal cord warrants a heightened clinical vigilance for both the general physicians as well as specialists.<sup>2</sup> Iskandar et al have shown the obvious advantage of surgical intervention for tethered cord syndrome in terms of neurological and functional improvements, despite the wide debate about the feasibility of the intervention in adolescence and adult patients.<sup>19</sup> Therefore, it is apparent that surgical intervention especially in the presence of symptomatic tethered cord syndrome has clear benefits over the wait and watch approach.<sup>2,20</sup>

The spectrum of spina bifida occulta varies in frequency concerning the individual lesions, however, diplomyelia and lipomyelomeningocele collectively form about 60% of the total.<sup>16,20</sup> Quite similar are the findings in our study where more than 56% of patients comprised of Lipomyelomeningocele (34.8%) and diplomyelia (21.7%). The majority of pediatric patients present within an age range of 7 to 12 years (mean 9 years in our study) with complaints of pain occurring in legs, hips, or back (95.7% collectively in our study) followed by dysfunction of the bowel or bladder (30.4%), various degrees of motor deficits, neuropathic ulceration of the feet, orthopedic deformities and dermal stigmata in the form of hair tuft over the lumbosacral area, dimples and swelling caused by the underlying lipomata. These findings have been noted in the studies by Das et al, Sarris et al, Muthukumar et al, Solmaz et al., Koo et al., Phi et al. and Iskandar et al.<sup>2,7,11,12</sup> Findings of the present study concur with the findings of the above studies. These consistencies in the findings of various studies denote that the presenting symptoms of Spina bifida occulta and Tethered cord syndrome are widely constant, despite differences in patterns of appearance, and can be relied upon in a clinical perspective.

Magnetic Resonance Imaging is the diagnostic tool of choice for defining the specific lesion of Spina bifida occulta and the level of tethering of the spinal cord.<sup>2,16,22</sup> Das et al, have

also described the efficacy of ultrasonography for defining Spina bifida occulta in very young neonates. However, for older patients, MRI has superseded CT scan as it delineates the soft tissue pathology clearly and defines the tethering level.<sup>2</sup> In addition to defining the spinal dysraphism, MRI was used by Erfani et al and Gharedaghi et al, for evaluating the presence or absence of syrinx and its resolution after surgery and also for the determination of congenital spinal orthopedic deformities such as congenital scoliosis.<sup>23,24</sup> MRI was the principal investigation after clinical diagnosis in our study. However, we tried to further confirm the lesion during surgery along with its spinal level which was shown by MRI during preoperative workup. This strategy helped us additionally in confidently classifying the lesion according to their type as found on open surgery.

Open surgical release of the tethered cord is the standard method of intervention in symptomatic cases of Tethered cord syndrome due to Spina bifida occulta. There has been increasing use of release and repair of Tethered cord syndrome under magnification and with the use of motor and sensory evoked potentials.<sup>16,22</sup> It has also been shown that these two modalities help improve the postoperative results in terms of functional improvement and complications avoidance. Four principle surgical methods are recommended by a review by Das et al.<sup>2</sup> namely: laminectomy & detethering, lipoma excision, resection of a bony spur, and division of filum terminale.

We used resection of the lipoma/spur and release of the Dural sac from any adhesions as well as a repair under magnification. Motor or sensory evoked potentials were not used. A very limited untethering approach was used for those who had minimal or no motor deficits and only the Dural sac was released taking care of the nerve rootlets.

The postoperative course of these patients usually is a dramatic improvement in the symptoms of pain with only a minority of patients

experiencing a transient worsening of sensory or motor deficits. We, however, followed the patients for the occurrence of early postoperative complications. The two major postoperative complications which affect the early postop course were CSF leak (26.1%) and wound infection (13%) mention in Table 4. No patient experienced a worsening or appearance of new motor deficit in our study, although it has been reported in 3.9% to 5% of patients of untethering surgery.<sup>22</sup>

Wound infections responded quickly by instituting barrier dressing methods and the use of intravenous antibiotics according to the sensitivity patterns. The mean hospital length of stay was found to be 5.7 days. Only two (8.7%) instances of reoperation for the repair of the dural defect in cases of CSF leak were found and two (8.7%) cases of CSF leak were relieved with insertion of a lumbar drain above the region of the dura repair. Most of the CSF leaks occurred in patients who had a meningocele sac repaired at operation. This signifies the fact that the dura in this subset of patients is very thin and closing sutures usually hold poorly. A meticulous closure technique is therefore advised. Studies by Solmaz et al and Galhom et al, employed similar techniques of untethering and repair of the thecal sac with complications of CSF leak and wound infections (20% CSF leak and 10% wound infection rates) similar to our study. The mean hospital stay was similar to the above studies as well as a study by Shweikeh et al, where the mean hospital stay was 3.5 to 5.1 days.<sup>25</sup>

## CONCLUSION

Early diagnosis and optimal treatment strategy development is the key to a successful outcome in patients with Tethered cord syndrome due to Spina bifida occulta. Earlier diagnosis is only possible if a high degree of clinical suspicion is observed, especially by the primary care physicians and neurosurgeons. Further studies are

required to further elucidate the association of various clinical correlates with occult spinal dysraphism and tethered cord syndrome and to establish their diagnostic and prognostic role.

## RECOMMENDATIONS

Early diagnosis of tethered cord syndrome has a better prognosis. So general public and general practitioners should know about these conditions and their signs/ symptoms. Proper surgical steps to detethering of the spinal cord are essential in spina bifida to prevent future neurological complications.

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## Additional Information

**Disclosures:** Authors report no conflict of interest.

**Ethical Review Board Approval:** The study was conformed to the ethical requirements.

**Human Subjects:** This was the retrospective study. The data of the patients was included as anonymous and used with the permission of the administration.

**Conflicts of Interest:**

In compliance with the ICMJE uniform disclosure form, all authors declare the following:

**Financial Relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.

**Other Relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

### **AUTHORS CONTRIBUTIONS**

<b>Sr.#</b>	<b>Author's Full Name</b>	<b>Intellectual Contribution to Paper in Terms of:</b>
1.	Ayaz Ahmad	Study design and methodology.
2.	Muhammad Ali Noman	Paper writing and data calculations.
3.	Muhammad Mukhtar	Data collection and calculations.
4.	Imran Khan, Muhammad Mushtaq	Analysis of data and interpretation of results etc.
5.	Shahid Ayub	Literature review and referencing.