# **Research Article**

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# Clinico pathological study of tethered cord syndrome

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#### **ABSTRACT**

**Background:** The disorder remains a challenge for both diagnostic and therapeutic measures. Today there is no question, however, that neurosurgery plays an important role in its treatment and indications for surgery have now widened rather than narrowed. Early diagnosis to prevent neurological deterioration is an urgent requirement. The objective of the study was, the current study focuses on tethered cord syndrome by age group, of the more common causes of the condition, as well as the adult presentation of occult TCS, its presentation and management.

**Methods:** This is a prospective study conducted on 44 patients who presented to outpatient department of Neurosurgery, Osmania Medical College/Hospital, Hyderabad between August 2011 to February 2014. All the patients were diagnosed based on clinical features investigations like X-ray spine and brain screening apart from routine investigations to assess fitness for surgery. All patients were counseled regarding the diagnosis, its treatment options, possible complications including intra operative bleeding, infection, cerebrospinal fluid leak/fistula formation, wound dehiscence, possibility of improvement, worsening, persistence of symptoms, retethering of cord and after obtained informed consent were included in the study and subjected to surgery.

Results: Infants (<1 year) are the most common age group of presentation as was observed in other series. There is no significant difference in incidence of tethered cord syndrome among males or females with a slight inclination towards males as was observed in other studies. Swelling over lower back with associated cutaneous lesion since birth are the most common presentations among children. Neurological deficits, bladder incontinence, bone deformities associated with tethering are commonly seen in elderly children or adults presenting late and do not subside / improve easily once they set in. Primary Tethered cord syndrome is more common than acquired. Back pain is generally a feature of presentation in elderly children and adults and probably the only symptom which subsides after detethering. Conclusions: MRI is the best diagnostic tool for identification of tethering and associated anomalies. Myelomeningocele and Lipomyelomeningocele are the most common associated anomalies with tethered cord syndrome. Proper identification of filum terminale, arachnoid bands and rootlets by electrophysiological monitoring intra operatively and good anatomical knowledge and proper establishment of CSF flow are essential for preventing retethering.

Keywords: Clinico pathological study, Tethered cord syndrome, Presentation, Management

### **INTRODUCTION**

Classically Tethered Cord Syndrome has been defined as a spectrum of congenital anomalies resulting in an abnormally low position of the conusmedullaris that may lead to neurological, musculoskeletal, urological, or gastrointestinal abnormalities.<sup>1</sup>

The term "tethered spinal cord" originated from the article in 1976 by Hoffman et al wherein the authors described 31 patients with elongated cords whose

symptoms improved following the sectioning of the filum terminale.<sup>2</sup> Most commonly TCS is related to spinal dysraphism. The signs and symptoms correlate with the radiological definition in which the conusmedullaris is anatomically lower than the L-2 vertebra or below the L1-2 disc space.<sup>3,4</sup> More recently, there have been descriptions of TCS in which patients are described to have the conusmedullaris in a normal position on imaging but presenting with signs and symptoms consistent with TCS.<sup>5,6</sup> Most of the patients with normal conus position but TCS are reported to have associated findings such as cutaneous stigmata, vertebral abnormalities, intra dural lipoma, and neurological abnormalities on examination.

In this patient population, symptoms of pain and bowel or bladder incontinence appeared to be responsive to detethering.<sup>7</sup>

Primary tethered cord syndrome seen in patients with myelomeningocele, lipomyelomeningocele, split cord malformation, dermal sinus tract, neuroenteric cyst, may be entirely different from secondary forms related to trauma, previous myelomeningocele repair or dysraphism in general.<sup>8</sup>

#### **METHODS**

This is a prospective study conducted on 44 patients who presented to outpatient department of Neurosurgery, Osmania Medical College/Hospital, Hyderabad between August 2011 to February 2014.

Patients presenting with various symptoms of tethered cord syndrome along with myelomeningocele, lipomyelomeningocele, split cord malformation, dermal sinus tract, neuroenteric cysts etc. were classified as having Primary Tethered Cord Syndrome.<sup>8</sup>

Patients developing features of tethered cord syndrome following repair of spina bifida aperta including myelomeningocele, meningocele, infections, and trauma are referred to as Acquired or Secondary Tethered cord syndrome.<sup>8</sup>

Patients who presented with features of ruptured myelomeningocele and who did not want to get operated or did not want to be included in the study were excluded.

All the patients were diagnosed based on clinical features investigations like X-ray spine and brain screening apart from routine investigations to assess fitness for surgery.

All cases with conusmedullaris at or above L 2 were considered to be at normal level. <sup>1</sup> In cases with conus at normal position thickness of filum terminale greater than 2 mm was considered abnormal although this finding remains controversial. <sup>1</sup> Any other associated anomalies were noted.

All patients were counseled regarding the diagnosis, its treatment options, possible complications including intra operative bleeding, infection, cerebrospinal fluid leak/fistula formation, wound dehiscence, possibility of improvement, worsening, persistence of symptoms, retethering of cord and after obtained informed consent were included in the study and subjected to surgery.

All patients who presented with features of spinal dysraphism with hydrocephalus were subjected to CSF diversion procedures followed by detethering of cord.

After obtaining informed consent for surgery patients were taken up for surgery (detethering of cord) under general anesthesia.

#### **RESULTS**

The incidence of tethered cord syndrome is not well documented worldwide. <sup>9</sup> But as per Solmaz et al it is estimated to be around 0.05 to 0.25 per 1000 births. <sup>8</sup>

The following observation was made as part of our study.

#### Age

The age incidence in the 44 cases (n=44) studied was as follows:

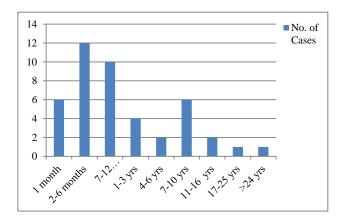


Figure 1: Incidence of age group in present study.

The mean age of presentation was 42.86 months. Maximum cases were seen in the age group of 2-6 months and few cases were seen in the age group of 17-25 years.

#### Sex

Of the 44 patients 21 patients (47.72%) were female and 23 patients (52.27%) were male (Figure 2).

#### Clinical features

22 patients presented with swelling over lumbo-sacral region at birth (50%) and among them, 7 patients had

lipomyelomeningocele and 15 patients had myelomeningocele.

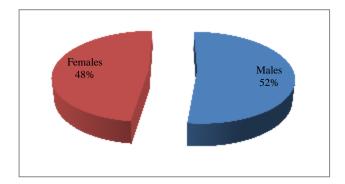


Figure 2: Pie chart showing sex ratio in present study.

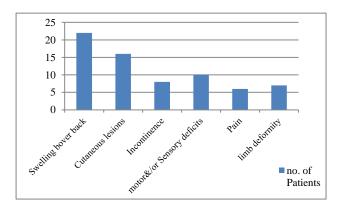


Figure 3: Distribution of study subjects as per their clinical features.

16 patients had associated cutaneous lesions (n=16) (36.36%). Of these, 5 patients presented with dermal sinus (31.25%), 4 with hyper-trichosis (25%), 3 with trophic ulcers over feet (18.75%), and 4 with atretic meningocele (25%).

8 patients presented with urinary incontinence (n=8) (18.18%).

#### MRI findings

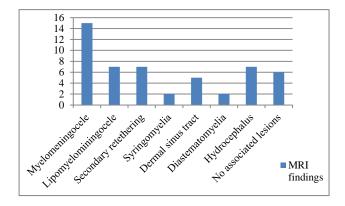


Figure 4: Columns showing MRI findings in present series.

15 patients presented with features of myelomeningocele (34.09%), 7 patients had associated Lipomyelomeningocele (15.90%). 5 patients had dermal sinus tract (n=5) (11.36%). 2 patients had Syrigomyelia (4.54%), 2 patients had diastematomyelia (4.54%). 7 patients had features of retethering like scarring (15.90%). 6 patients had no associated lesions (n=6) (9.09%).

#### **Complications**

No significant bleeding was observed in any of the cases. Among the 44 patients who underwent surgery, cerebrospinal fluid leak was observed in 2 patients (4.54%) who subsided with conservative management. Only 1 patient (2.27%) presented with infection among the 44 patients. No neurological deterioration was observed in any of the patients. Retethering was not observed in any of the patients.

#### Surgicaloutcome

Following detethering and surgery of associated anomalies the following observations were made:

Among the 10 patients presenting with motor or sensory deficits; 5 patients presented with upper motor signs among these 4 showed improvement over a period of time. There was no significant improvement in patients presenting with lower motor (2) or mixed signs of upper or lower signs (3). Of the 6 patients only 2 showed improvement over a patients (n=10) presenting with neurological deficits. Back pain subsided in all the 6 patients (100%). Unfortunately incontinence did not improve in any of presenting patients. Lower limb deformities persisted after the surgery. Trophic ulcers improved in 1 patient (33%) among the 3 patients presenting.

#### **DISCUSSION**

# Age

The youngest patient operated was 7 days old and the oldest was 27 years (range 7 days-27 years) who presented with retethering of cord who underwent surgery for myelomeningocele in infancy. In studies conducted by Hoffman et al<sup>1</sup> the range was 6 days-18 yr.

# Sex

Among the 44 patients (100%) studied 21 patients (47.72%) were females and 23 (52.27%) were males. The observations made by Solmaz et al  $^8$  were 40.8% females and 59.2% were males.

This showed that males are slightly more affected in cases of tethered cord syndrome which is in accordance with contemporary studies (Table 1).

Table 1: Table comparing sex distribution in present series to contemporary series.

Series	Sex	% Of cases
Solmaz et al <sup>8</sup>	Male	59.2%
	Female	40.8%
Present series	Male	52.27%
	Female	47.72%

#### Clinical features

Most of the patients presented with more than one clinical feature. Majority of the patients presented with

complaints of swelling over lower back since childhood (50%) in comparison to contemporary study by Solmaz et al who found the incidence to be 46%. But for cutaneous lesions, the incidence of neurological deficits, pain, deformity and incontinence in present study was in accordance with study conducted by Solmaz et al. Neurological deficits were more common in children presenting more than 6 months of age which is in accordance with study conducted by Hertzler et al according to whom 62.5% patients who presented before 6 months of age were asymptomatic when compared to 29% of those presenting at more than 6 months of age. Koyangi et al reported that no child in their study population was asymptomatic after 5 years of age. 10

Table 2: Table comparing incidence of clinical features in present study to contemporary study.

Clinical features	Cutaneous us lesion	Incontinence	Neurological deficits	Deformity	Pain	Swelling
Solmaz et al <sup>8</sup>	61.2%	20.41%	18.4	10.2%	1.0.2%	46%
Present series	36.36%	18.18%	22.72%	15.9%	13.63%	50%

### MRI findings

MRI was done as modality of choice for evaluating cases of tethered cord syndrome. MRI was used to identify the level of conusmedullaris, thickness of filum and nay other associated anomalies by screening whole spine including carnio vertebral junction. There was certain variation in data of present series with the findings in corresponding date of Solmaz et al who had studied tethered cord exclusively in children hence probably the difference. Myelomeningocele (34.9%) was the most

common presentation in present series followed by lipomyelomeningocele (15.90%), scarring (15.90%), and dermal sinus tract (11.36%), an associated lesions (9.09%), Syrigomyelia (4.54%) and diastematomyelia (4.54%). Of the 44 patients included in the study, basing on history, clinical features and MRI findings 38 patients were diagnosed as having primary tethering and 7 patients as having acquired tethered cord syndrome. Primary tethering is a more common entity than acquired which is in accordance to study conducted by Solmaz et al.<sup>8</sup>

Table 3: Table comparing MRI finding of present series with contemporary study.

Lipo	meningocele	Dermal sinus tract	Split cord malformation	Syringo Myelia	Previous surgery / scarring	No other associated lesions
Solmaz et al <sup>8</sup>	32.6%	14.3%	28.6%	2.1%	4.1%	16.3%
Present series	15.90%	11.36%	4.54%	4.54%	15.90%	9.09%

# **Complications**

Among the 44 cases operated 7 patients had history of previous surgery in childhood / infancy with mean age of 15 years (who required redo surgery) with the youngest patient presenting with retethering at 7 years in present study. So probably a longer period of follow up may be necessary. Though majority of the patients did not develop any significant complications they should be kept in mind and the patient and attendees need to be counseled regarding these. No patient expired among the patients studied.

Table 4: Table comparing incidence of complications in present series to contemporary series.

Compli -cation	Blee- ding	Infe- ction	New deficits	CSF leak	Reoper- ation
Kang et al <sup>11</sup>	00	00	00	05 (6.7%)	04 (5.3%)
Present series	00	01 (2.27%)	00	02 (4.54%)	00

#### Surgical outcome

Unfortunately incontinence did not improve in any of presenting patients (n=8) which is the same in studies done by Solmaz et al Kang et al Hertzler et al Kanev et al in a large retrospective study noted that urological dysfunction showed no improvement even with extended follow up but motor and sensory symptoms did demonstrate significant recovery which is in accordance with present series. 1,8,11,12

In present series patients presenting with upper motor neuron signs improved in comparison to those who had lower or mixed signs and no significant improvement was observed in sensory deficits. This indicates that once a neurological deficit set in it is difficult or probably the clinical deficits persisting spite of detethering. Hence an early surgery needs to be contemplated in all cases presenting with features of tethered cord syndrome.

Table 5: Table showing comparison of surgical outcome with contemporary studies.

Out- come	Neurological improvemen	Inconfi-	Kack	Defor- mities
Solmaz et al <sup>8</sup>	In 4 of 9 patties (44.4%)	No improve- ment	Subsided in all	Persi- sted
Present series	4 of 10 patients (4%)	No improve- ment	Subsided in all	Persi- sted

# CONCLUSION

MRI is the best diagnostic tool for identification of tethering and associated anomalies. Myelomeningocele and Lipomyelomeningocele are the most common associated anomalies with tethered cord syndrome. Proper identification of filum terminale, arachnoid bands and rootlets by electrophysiological monitoring intra operatively and good anatomical knowledge and proper establishment of CSF flow are essential for preventing No significant complications retethering. encountered during the present study. All cases presenting with tethered cord features should be operated as early as possible in spite of patients being asymptomatic as delay in surgery could lead to serious neurological deficits which do not easily subside once they appear.

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Ethical approval: The study was approved by the

institutional ethics committee

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