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Clinicoradiological, pathological, and surgical outcome in patients with tethered cord syndrome: A prospective study

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ABSTRACT

Background: Tethered cord syndrome (TCS) is a stretch-induced functional disorder of the spinal cord in which the caudal part is anchored by an inelastic structure. It is believed that, if the viscoelasticity of the filum is lost or compromised by either fatty infiltration or abnormal thickening, caudal tension and traction may cause undue stress upon the conus, resulting in TCS. It typically occurs in children, and it is rare in adults. **Aims and Objectives:** The study was conducted to know the clinicoradiological, pathological, and surgical outcomes in patients with TCS. **Materials and Methods:** This is a prospective observational study done in the premier institute of central India from May 2022 to April 2024. This study included patients who exhibited clinical and/or radiological features of congenital tethered cords. This encompassed both patients with tethered cord and those with TCS. Patients who failed to give consent and refused to participate in the study were excluded from the study. **Results:** A total of 30 patients were recruited for the study. The median age of the patients was 6–12 months (range). The gender distribution M:F was 63.3:36.6. Based on 30 cases studied, most of the patients (75%) had improvement in urinary symptoms, 57.1% of patients had improvement in bowel symptoms, 80% of patients had improvement in pain symptoms, 75% had improvement in motor deficit, and 83.3% had improvement in sensory deficit. **Conclusion:** Our prospective, cross-sectional study aimed at complete detethering of the spinal cord and the nerve roots along with excision of filum terminale and dural reconstruction to maintain adequate cerebrospinal fluid space around the spinal cord to prevent retethering.

Key words: Tethered cord syndrome; Tethered cord syndrome filum terminale; Filum terminale spinal dysraphism; Spinal dysraphism meningomyelocele; Meningomyelocele Diastematomyelia

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INTRODUCTION

Tethered cord syndrome (TCS) is a stretch-induced functional disorder of the spinal cord in which the caudal part is anchored by an inelastic structure. It is believed that, if the viscoelasticity of the filum is lost or compromised by either fatty infiltration or abnormal thickening, caudal tension and traction may cause undue stress upon the conus, resulting in TCS.[1] It is believed that this abnormal inelastic filum interferes with normal cord ascent and results in a low-lying conus medullaris (that is, a conus below the L1–L2 interspace). This is classically believed to be the hallmark of the TCS. This can be secondary to a heterogenous group of disorders, such as spinal lipomas, lipomatous filum, split cord malformations (SCMs), and meningomyelocele. It typically occurs in children, and it is rare in adults.[2] Congenital tethered cervical spinal cord is a very rare entity and is usually due to a dermal sinus tract stalk entering the subarachnoid space and attaching to neural elements. In cervical dysraphism, tethering of the neural structures to nearby dural or intrasaccular structures may be present.

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The term filum terminale syndrome was first used by Garceau in 1953 in describing three patients.[3] In 1976, Hoffman et al., coined the term "tethered spinal cord" in patients with a low-lying conus medullaris with a thickened filum.[4]

Adult-onset cases are rare compared to that in children.[5] The risks for spinal dysraphism include folic acid deficiency,[6] maternal age (very young or very old), toxic medications, obesity, multiple gestations, anti-epileptic medications, zinc deficiency, and excessive tea consumption during the 1st trimester of pregnancy. The abnormally low position of the conus medullaris may lead to neurological, musculoskeletal, urological, orthopedic, or gastrointestinal abnormalities.[7] Although TCS is a well-known entity, it continues to pose challenges regarding diagnosis and management.[8]

In our study, we analyze the different presentations of TCS, their surgical outcome, various prognostic factors that can predict the outcome of surgery, and determine the incidence of various post-operative complications[9,10] in such patients.

Aims and objectives

The study was conducted to know the "clinico-radiological, pathological and surgical outcome in patients with tethered cord syndrome."

MATERIALS AND METHODS

Study design

Prospective study.

Study population

The above study included 30 consecutive patients who were admitted to the Department of Neurosurgery at J.A. Group of Hospitals, G.R. Medical College in Gwalior, Madhya Pradesh, over a 24-month period.

Inclusion criteria

Patients who exhibited clinical and/or radiological features of congenital tethered cord. This encompassed both patients with tethered cord and those with TCS.

Exclusion criteria

Exclusion criteria included patients with severe co-morbid illness, absconding or leaving against medical advice, unwillingness for surgery, and non-cooperation.

Study procedure

Patients with clinical and radiological features of TCS who met the inclusion criteria were included in the study.

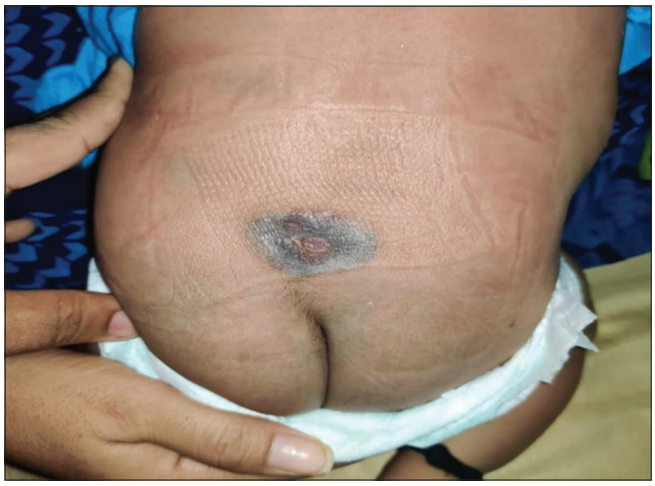

Figure 1: Cutaneous stigmata: Skin dimple

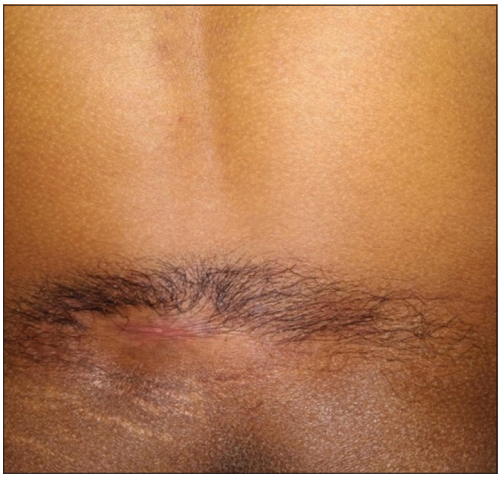

Figure 2: Cutaneous stigmata showing tuft of hair

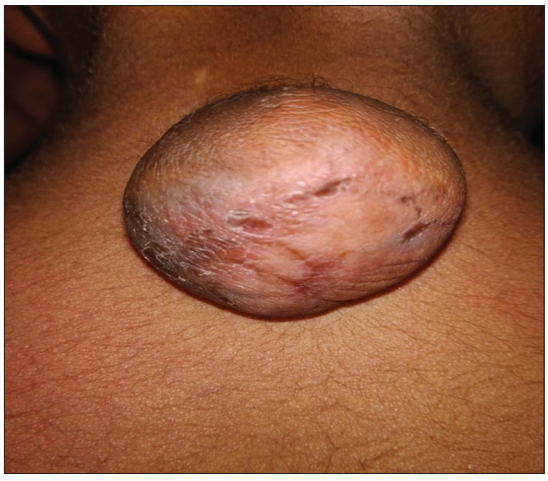

Figure 3: Picture showing dorsal myelomeningocele

Data collection

The study received Medical Ethical Committee approval, and data were collected for all admitted patients according to pro forma.

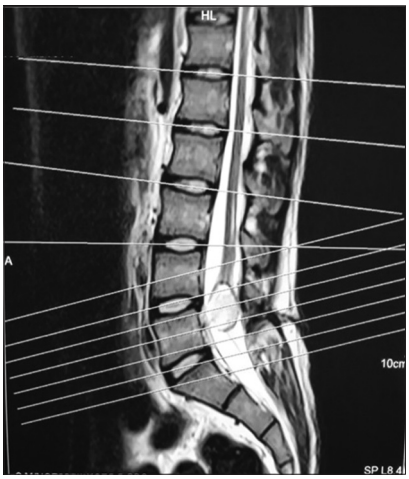

Figure 4: T2 sagittal magnetic resonance imaging showing dermoid cyst with low-lying conus

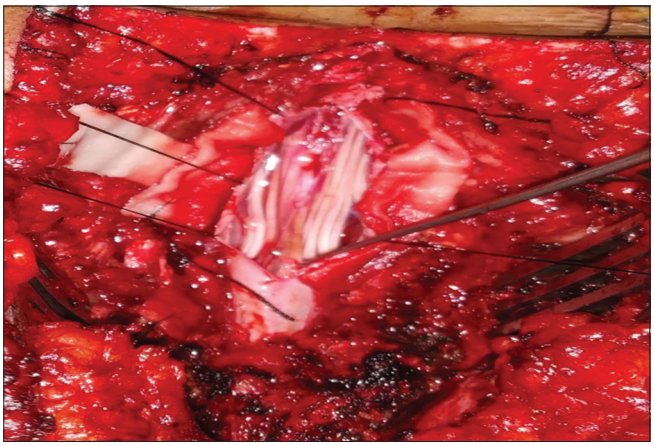

Figure 5: Thickened filum terminale lifted by Dandy nerve hook

Pre-operative clinical assessment

Recruitment of patients

Patients who were referred for surgical evaluation for suspected TCS were identified and approached for study participation. Informed consent was obtained from all participants.

Clinical assessment

A detailed clinical assessment was performed for each patient, including a review of symptoms, medical history, and physical examination. The presence of any neurological deficits was also recorded.

Imaging studies

All patients underwent magnetic resonance imaging (MRI), and in some cases, computed tomography or ultrasound imaging studies to confirm the diagnosis of TCS and evaluate its extent.[11-13]

Pathological examination

If surgical intervention is performed, specimens were collected and examined to determine the extent of spinal cord tethering and the presence of any underlying pathologies.

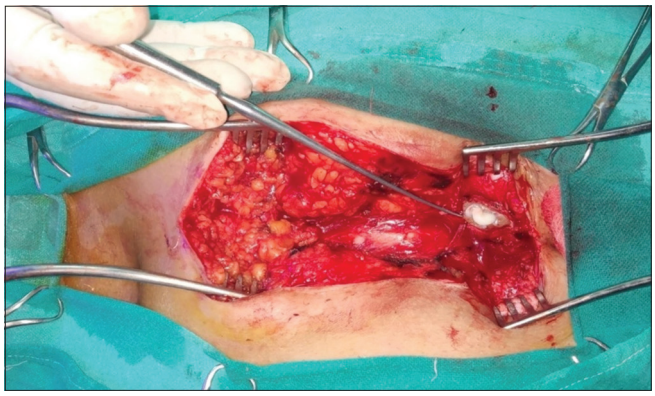

Figure 6: Penfield dissector showing dermal sinus expelling sinus content

Surgical intervention

Patients underwent surgical intervention for TCS as clinically indicated.

Post-operative follow-up

Patients were followed post-operatively to assess surgical outcomes, including changes in symptoms and neurological deficits.

Data collection

Data on patient demographics, clinical presentation, radiographic findings, pathological findings, kyphosis angle, surgical intervention, and post-operative outcomes were collected and recorded in a study database.[14-16]

Data analysis

Descriptive statistics was used to summarize patient demographics, clinical presentation, and radiographic findings.[17-20] The accuracy of different imaging modalities in diagnosing TCS was evaluated using sensitivity, specificity, and diagnostic accuracy. The relationship between pathological findings and clinical outcomes was analyzed using appropriate statistical methods. Surgical outcomes, including changes in symptoms and neurological deficits, were assessed using appropriate statistical methods.

Statistical methods

Statistical analysis was performed using R, a software environment for statistical computing and graphics (version 3.6.1; Vienna, Austria). Numerical variables (nonparametric) were expressed as a median with interquartile range. Categorical variables were expressed as a percentage. Bivariate analysis between categorical variables was performed by the Chi-square test. The normality of numerical variables was analyzed by the Shapiro–Wilk test. Bivariate analysis between categorical and numeric variables was performed using the Wilcoxon rank-sum test

with continuity correction comparing two groups and the Kruskal–Wallis rank-sum test comparing more than two groups. P<0.05 was considered statistically significant.

RESULTS

Age distribution (Table 1)

6-12 months is the most common age at presentation

Gender-wise distribution (Table 2)

In this study 63.3% were male and 36.6% were female

Cutaneous stigmata (Table 3)

Cutaneous stigmata were present in around 70% (n=21) of the patients in this study, of which lipoma was found in 46% followed by dermal sinus in 17%.

Type of spinal dysraphism associated with tethered cord (Table 4)

36.6% of the patient with Thethered cord syndrome presented after being previously operated for Meningomyelocele

Associated anomalies found on MRI (Table 5)

In MRI study Syringomyelia(30%) is the most common radiological finding followed by Scoliosis/kyphoscoliosis (6.6%)

MRI findings of the position of the conus (Table 6)

L3(53.3%) vertebral level is the most common position of conus followed by L2(6.6%) level

Type of operation done (Table 7)

Excision of lipoma with the division of flum terminale is the most common operative procedure which was performed

Follow-up of signs and symptoms (Table 8)

Refer to the table below

Table 1: Age distribution

Age	Number of patients	Percentage
0–6 months	6	20
6–12 months	11	36.6
1–5 years	7	23.3
5–14 years	4	13.3
>14 years	2	6.6
Total	30	100

Table 2: Gender-wise distribution

Gender	Number of patients	Percentage
Male	19	63.3
Female	11	36.6
Total	30	100

Table 3: Cutaneous stigmata

Cutaneous stigmata	Number of patients	Percentage
Lipoma	9	42.8
Dermal sinus	4	19
Cervical meningomyelocele	2	9.5
Lumbar meningomyelocele	3	14.2
Tuft of hair	1	4.7
Skin dimple	1	4.7
Lumbar meningocele	1	4.7
Total	21	100

Table 4: Type of spinal dysraphism associated with tethered cord

Type of spinal dysraphism	Number of Patients	Percent
Operated case of MMC	11	36.6
Dermal sinus	5	16.6
Cervical meningomyelocele	2	6.6
Lumbar meningomyelocele	5	16.6
Thickened filum terminal	1	3.3
Diastematomyelia	1	3.3
Dorsal meningomyelocele (Figure 3)	3	10
Conus dermoid (Figure 4)	1	3.3
Lumbarmeningocele	1	3.3
Total	30	100

DISCUSSION

Age and gender distribution

The present study found that most patients are aged 1–10 years, differing slightly from previous studies. It also showed a male predominance with a male-to-female ratio of 1.72:1, consistent with earlier findings. Kafle et al., reported a similar ratio of 1.77:[22] in a study of 97 patients. In contrast, Iskandar et al., found 12 men and 22 women in their prospective study.[23]

Types of associated spinal dysraphism with cutaneous stigmata

Our study found that 42.8% of patients had cutaneous stigmata like lipoma and 19% had a dermal sinus (Figure 6). Ailawadhi et al., observed spine curvature abnormalities in 61.7% of 34 patients. In their study, 29.4% had cutaneous stigmata: 2.9% had a sacral dimple (Figure 1), 2.9% had a subcutaneous lipoma, and 14.7% had a tuft of hair (Figure 2). In addition, parchment skin, dermal sinus, and skin appendage were each seen in one patient. Urological complaints were present in 32.3% of patients, limb weakness in 55.8%, numbness in 17.6%, leg and foot deformity in 32.3%, and back pain in 20.5%.[24]

In the present study, lipomyelomeningocele was found in 9 (42.8%) patients, 4 (19%) patients had dermal

sinus, cervical meningomyelocele 2 (9.5%), lumbar meningomyelocele 3 (14.2%), thickened filum terminale were found in 1 (3.3%) of patients, and 1 (3.3%) patient had diastematomyelia. Rajpal et al., 2007 in their prospective study on lipomyelomeningocele (25 patients), tight filum terminal (22 patients), SCMs (15 patients), syringomyelia (7 patients), dermoid cysts (two patients), and meningocele (one patient).[25]

Position of conus

In the study by Ailawadhi et al., the conus was observed to be low lying at the L3 level in 58.8% of patients. In the present study, out of 30 patients, 53.3% had the conus at the L3 level, 13.3% at L4, 6.6% at L2, 3.3% at L5, and 23.3% had a normal conus position.

Table 5: Associated anomalies found on MRI

Associated anomaly	Seen In	Percentage
Syringomyelia	9	30
Scoliosis/kyphoscoliosis	2	6.6
Dural ectasia	16	53.3
Hydrocephalous (pre-operative)	5	27
Hydrocephalous (post-operative)	1	3.3
CV junction anomaly	1	3.3
ACM	9	30
Diastematomyelia	1	3.3

Table 6: MRI findings of the position of the conus

Position of conus	Number of patients	Percentage
L3	16	53.3
L2	2	6.6
L4	4	13.3
L5	1	3.3
Normalconus position	7	23.3
Total	30	100

Intraoperative findings

In this study, 36.6% of patients had lipoma with tight filum terminale, followed by adhesion in 20% of cases, adhesion with tight filum terminale in 16.6%, and other combinations such as dermal sinus with adhesion with tight filum terminale, myelomeningocele with thickened filum terminale (Figure 5), and others. A study by Lee et al. in 2006 also reported similar findings, with lipoma and tight terminal filum being the most common, followed by tight filum terminale alone, myelomeningocele, and various combinations of abnormalities involving adhesions, SCM, dermoid tumor, and others.

Outcome

In the present study, a majority of patients showed improvement in urinary symptoms (75%), followed by improvements in bowel symptoms (57%), pain symptoms (80%), motor deficit (75%), and sensory deficit (83.3%). Huttmann et al.,[26] observed that in their study of 56 adult TCS patients, the pain relief rate was 86%, with lower limb spasticity remission at 71%, and remission rates for bladder dysfunction and movement dysfunction at 44% and 35%, respectively. Another study by Lee et al., in 2006 found improvement in urinary symptoms in 50% of patients, motor weakness in 64%, back pain symptoms in 83%, and sensory symptoms in 50%.[27]

Table 7: Type of operation done

Type of spinal dysraphism	Operation	Number	Percentage
Lipomyelomeningocele	Excision of lipoma with the division of flumterminale	9	30
Thickened filum terminal	Excision of filum terminale	2	6.6
Lumbar meningomyelocele	Excision of meningomyelocele	5	16.6
Cervical meningomyelocele	Excision of meningomyelocele with the division of adhesion	2	6.6
o/c/o MMC	Adhesion release with excision of filum	11	33
Dermal sinus	Excision of the sinus with the division of adhesion	5	16.6
Diastematomyelia	Laminectomy with excision of the bony spur with excision of filum	1	3.3
Dorsal meningomyelocele	Excision of meningomyelocele with the division of adhesion	3	10
Conus dermoid	Laminectomy with near total excision with excision of filum terminale	1	3.3
Lumbar meningocele	Excision with repair	1	3.3

Table 8: Follow-up of signs and symptoms

Sign and symptoms	Improved (%)	Stable (%)	Worse	Recurrence	Unknown (%)	Total
Urinary dysfunction	9 (75)	2 (6.6)	0	0	1 (8.3)	12
Bowel dysfunction	4 (57.1)	2 (14)	0	0	1 (9)	7
Pain	4 (80)	0	0	0	1 (20)	5
Motor deficit	6 (75)	1 (12.5)	0	0	1 (12.5)	8
Sensory deficit	5 (83.3)	0	0	0	1 (16.6)	6
Sexual dysfunction	0	0	0	0	1 (100)	1

CONCLUSION

Spinal dysraphism is a benign condition presenting mostly in the early stage of life; thorough evaluation is necessary followed by meticulous surgery along with complete detethering to give the patient an optimal chance to live a better social life in the future. Almost all cases of spinal dysraphism involve some degree of tethering of the spinal cord or nerves.[21] Even if symptoms are not present initially, there is a high risk of neurological deterioration later in life, so it is better to do adhesiolysis/detethering during the primary surgery itself and avoid further surgeries in the future. The aim of the treatment should not be merely cosmetic; rather, it should be a releasing surgery for the cord and roots. The spinal canal is opened rostrally and caudally to completely detether the cord from surrounding structures.

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Authors' Contribution:
HDP - Definition of intellectual content, literature survey, prepared the first draft of manuscript, implementation of study protocol, data collection, data analysis, manuscript preparation and submission of article; **AnS -** Concept, design, clinical protocol, manuscript preparation, editing, and manuscript revision, design of the study, statistical analysis and interpretation, review manuscript; **KA -** Review manuscript; **SS -** Literature survey and preparation of figures; **AvS -** Coordination and manuscript revision.

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