

Teethered Cord Syndrome: A Retrospective Clinical Study at a Tertiary Care Hospital

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ABSTRACT

Background: Progressive neurological, urological, and orthopedic dysfunction due to congenital fixation or tethering of the spinal cord is known as Teethered Cord Syndrome (TCS). The clinical constellation of signs and symptoms associated with TCS may include dermatologic, urological, gastrointestinal, neurological, and orthopedic findings. It is essential to identify this entity at a very early stage by various cutaneous stigmata so that damage caused by stretching of cord can be prevented thereby avoiding various constellation of neurologic and an orthopedic problem as the child grows. Although the incidental radiographic finding of asymptomatic teethered cord is becoming increasingly common, more often some combination of clinical signs and symptoms results in patients coming to the attention of neurosurgeons. Therefore this study focuses primarily on the clinical entity of teethered cord syndrome with discussion of its embryology, history, pathophysiology, diagnosis and treatment.

Methods and Materials: All patients both children and adults, who presented in Department of Neurosurgery at B. J. Medical College and Civil Hospital Ahmedabad, during 2.5 years period from July 2012 to December 2014 with signs and symptoms of TCS were included in our study. In cases with swelling over back and where there was no radiological evidence of tethering and no clinical features of tethering of cord were excluded from study. MRC grading was used to assess power. Plain roentograms, Magnetic resonance imaging and CT Scans were used to assess the nature of tethering and associated pathologies. Ultrasonography was used to assess urological abnormality.

Results: In our series 45% of patients who had residual weakness were restored to normal power at the end of 12 month follow-up and 60% of cases were restored to normal day to day activities at the end of 2 year follow-up. Among 10 patients who had partial improvement in bladder continence, 60% regained normal bladder sensations and were maintained on intermittent self-catheterization at the end of 2 year follow-

up. Patients with orthopedic abnormality like scoliosis and foot deformity underwent corrective orthopedic surgeries and restored to normal daily activities at the end of 2 year follow-up. All patients underwent stringent physiotherapy post-operatively and assessed regularly at subsequent follow-up visit which greatly improved the life style of these patients.

Conclusion: The main problem in the diagnosis of the syndrome of teethered cord syndrome in children at the present stage is that few parents and clinicians are aware of the importance of certain skin manifestations, neurological, orthopedic, urological disorders that may indicate spinal dizrafizm. Early detection of disease and timely surgical intervention help to halt or stabilize the progression of neurologic deficit.

Patients suspected of having TCS must be referred and treated by the age of 2 years, or soon after diagnosis, as they are likely to develop progressive neurological deficits if untreated. Normal radiology in the presence of clinical features of cord tethering should not exclude the diagnosis of TCS.

Keywords: Teethered Cord Syndrome, Neurological Deficits, Surgical Intervention.

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INTRODUCTION

The term "filum terminale syndrome" was first used by Garceau in 1953 in describing three patients.¹ Two decades later, in 1976, Hoffman and colleagues coined the term "teethered spinal cord"

to describe the symptoms of their patients with an low lying conus medullaris and a thickened (> 2 mm diameter) filum terminale;² however the term "spina bifida occulta" (Virchow, 1875) and the

first successful surgical intervention for a teethered spinal cord (Jones, 1891) have been described almost a century prior to this.³ Teethered Cord Syndrome (TCS) refers to progressive neurological deterioration in the functions of the lower spinal cord resulting from traction on the conus medullaris.³ Though generally associated with a low lying conus (the "classic" type), TCS is also seen in a normally positioned conus.⁴

Now, TCS is known to be associated with diverse etiologies including spinal dysraphism (aperta or occulta), vertebral and orthopedic abnormalities (scoliosis, limb deformities), caudal regression, tail fold anomalies and anorectal malformations. Amongst occult spinal dysraphism, TCS is common in lumbosacral lipoma, split cord malformations, diastematomyelia, dermal sinus tracts, congenital inclusion tumors complex and fibrous adhesions.

Yamada et al.⁴ in 1981 broadened the stretch-induced functional disorder to include patients with other anomalies, such as myelomeningocele, lipoma, lipomeningomyelocele, diastematomyelia, meningocele, and dermoid sinus. Recently, Yamada and Won⁵ described three categories of TCS. The first was lumbosacral cord anchored by an inelastic filum, and the second category included caudal as well as many sacral myelomeningoceles. The third category was divided into two groups; the first was paraplegia associated with lipomeningomyelocele and myelomeningocele, apparently with no functional lumbosacral neurons. No neurologic benefit is expected from surgery in this group. The second group included asymptomatic patients with an elongated cord and a thick filum. These patients require close observation for subtle symptoms, particularly incontinence, which if untreated might quickly become irreversible. The neurological sequelae involve a combination of upper- and lower- motor neuron dysfunction that are clinically quiescent or manifest variably in the lower limbs, the bowel and the bladder. The optimal investigations and treatment of TCS in children is controversial and protocols vary from conservative approach to the overtly aggressive intervention. This article briefly outlines the pathophysiology, clinical presentation, investigations and treatment of teethered cord syndrome.

AIMS

1. To study various clinical presentations of teethered cord syndrome.

2. To assess neurological status of patients presenting with teethered cord syndrome.
3. To assess radiological abnormalities present in patients with teethered cord syndrome.
4. To co-relate clinical manifestations with underlying radiological abnormalities.
5. To study the role of surgical intervention in management of these cases.

METHODS AND MATERIALS

It was a retrospective study. 80 patients both children and adults, who presented and were managed in Department of Neurosurgery at B. J. Medical College and Civil Hospital Ahmedabad, during 2.5 years period from July 2012 to December 2014 with signs and symptoms of TCS were included in our study.

In cases with swelling over back and where there was no radiological evidence of tethering and no clinical features of tethering of cord were excluded from study. MRC grading was used to assess power. Plain roentograms, Magnetic resonance imaging and CT Scans were used to assess the nature of tethering and associated pathologies. Ultrasonography was used to assess urological abnormality. The frequency and presentation of each type of spinal dysraphism and the natural history of untreated late presenting cases were studied for the progression of signs and symptoms. The current study focuses on TCS by age incidence, sex distribution, varied clinical presentation and cutaneous stigmata, neurological manifestation, radiological features, operative procedures and post-operative outcome.

RESULTS

In our study majority of our patients were in the age group of <2 years which constituted 62.5% of cases 10% of cases were >18yrs. Earliest age of presentation in our series was 1 month and 40 years was oldest. There was female preponderance with incidence in females being 52.5% and males being 47.5%.

Among the varied cutaneous stigmata, 52.5% of cases presented with lipomeningocele, 17.5% with meningomyelocele, 10% with tuft of hair (hypertrichosis), 13.75% with recurrent ulcerations of foot, 10% each with foot deformities and dermal, 6.25% with scoliosis, 6.25% with skin dimpling, 5% with atretic meningocele, 5% of cases presented with re-tethering after previous surgery for meningomyelocele and 1.25% presented with naevus.

Table 1: Local Examination Findings

Local examination findings	Total no. of patients	Percentage
Lipomeningocele	42	52.5
Meningomyelocele	14	17.5
Foot ulcerations	11	13.75
Dermal sinus	8	10
Tuft of hair	8	10
Back pain	8	10
Foot deformities	8	10
Dimpling of skin of back	5	6.25
Scoliosis	5	6.25
Recurrent /re-tethering	4	5
Atretic meningocele	4	5
Dimpling /puckering of skin of back	5	6.25
Naevus	1	1.25



Figure 1: Atretic meningocele with dermal sinus



Figure 2: Dorsolumbar nevus



Figure 3: Trophic Ulcer over foot



Figure 4: Foot deformity with resorption of Toes

Radiologically identified abnormality being dorsal lipoma in 20%, terminal lipoma in 21.25%, transitional lipoma in 15%, combined lipoma in 5%, thickened with low lying conus in 30%, diastematomyelia in 20%, diplomyelia in 15%, syringomyelia in 17.5%, intraspinal dermoid in 7.5%, agenesis of lower lumbo-sacral segments in 6.25%, associated hydrocephalus in 22%.

Depending on site of these spinal cord abnormalities 2.5% were in cervical level, 6.25% in dorsal level, 11.25% in dorso-lumbar level, 43.75% in lumbar level, 28.75% in lumbo-sacral level and 7.5% in sacral level.

50% of cases presented with weakness of lower limbs and among them 2.5% had grade 0, 1.25% had grade 1, 7.5% had grade 2, 20% had grade 3 and 18.75% had

grade 4. 1.25% had hypertonia in lower limbs, 16.25% had hypotonia and 71.25% had normal tone. In 25% of cases reflexes were brisk and in 25% of cases reflexes were absent and 50% cases had normal reflexes. Sensory disturbances were present in 25% of cases and among them 55% had definite sensory level and 45% had tingling and numbness. 30% of cases presented with bladder incontinence and 17.5

% of cases had associated bowel incontinence. And Among patients with bladder incontinence 60% of them had significant post-void residual urine on ultrasound and 40% of them empty bladder.

Among orthopedic problems 13.3% of cases had foot deformities in the form of equinus deformity, forefoot varus, club foot deformity and resorption of distal ends of toes. Recurrent ulcerations were present in 10% of cases and majorities were associated with foot

deformities. In 6.25% of cases kyphoscoliosis was present and among them 60% had diastematomyelia and 40% had diplomyelia and these were most commonly found with tethering of cord in dorsal and dorsolumbar region. Limb length discrepancy along with thinning of limbs was present in 10% of cases. These set of patients had abnormal gait and backache which worsened on doing strenuous activity. These problems were observed most commonly in children between 10-18 yrs which corresponds to age of rapid growth.

Clinico-radiological co-relation showed that among patients who had swelling over lower back 60% had lipoma, 1.67% had meningomyelocele, 3.33% had dermoid cyst, 3.33% had sacral agenesis, 13.33% had diastematomyelia and 3.33% had diplomyelia.

Among patients with tuft of hair, diastematomyelia was present in 50% of cases, diplomyelia in 25%, dermoid in 25%. Among patient with dermal sinus 50% had spinal dermoid, 25% had diplomyelia. Among patients with dimpling of skin 20% had diastematomyelia and diplomyelia in 80% of cases.

56.25% of cases remained same as pre-op. Of 31.25% cases that showed improvement in power as compared to pre-op, 20% had full recovery to normal power and 80% had partial improvement in power after surgery. 10% had post-operative deterioration of power. Among them 80% had grade 5 power pre-operatively and 20% had pre-op weakness,

Among patients with bladder incontinence 50% had same pre-op status, partial improvement in bladder control was observed in 50%. 17.5% had Bowel incontinence and among them 35.71

remained same as pre-op and 50% had partial improvement and 14.29% had complete recovery. In patients with recurrent ulcerations there was improvement in healing of ulcers in 75% of cases. Patients with foot deformities, limb length discrepancy and kyphoscoliosis underwent orthopedic corrections after detethering procedure. These patients showed significant improvement in gait and reduction of backache at the end of 1 year follow up. 20% post-operative complications.

Among them 50% had CSF leak, 37.5 had wound infection and dehiscence and 12.5% had post-operative hydrocephalus. 37.5% of CSF leak and 50 % of wound infections were managed

conservatively. 62.5% with CSF leak and 50% with wound infection underwent re-exploration. Follow-up was carried out at regular intervals which showed 45% of patients who had residual weakness were restored to normal power at the end of 1 year and total 60% of cases were restored to normal power at 2 year follow up. Among 10 patients who had partial improvement in bladder incontinence, 60% regained normal continence and maintained on intermittent self-catheterization at the end of 2 year follow-up. Patients with orthopedic problems like scoliosis and foot deformity were rehabilitated to near normal activity after corrective orthopedic surgery and subsequent physiotherapy.

Table 2: Radiological Findings

Radiological findings	Total no. of patients	Percentage
Dorsal lipoma	16	20
Terminal lipoma	17	21.25
Transitional lipoma	12	15
Mixed variety lipoma	4	5
Thickened filum with low lying cord	24	30
Diastometamyelia (scm type1)	16	20
Diplomyelia (scm type 2)	12	15
Syringomyelia	14	17.5
Intramedullary dermoid	6	7.5
Sacral agenesis	5	6.25
Scoliosis	5	6.25
Hydrocephalus	18	22.5

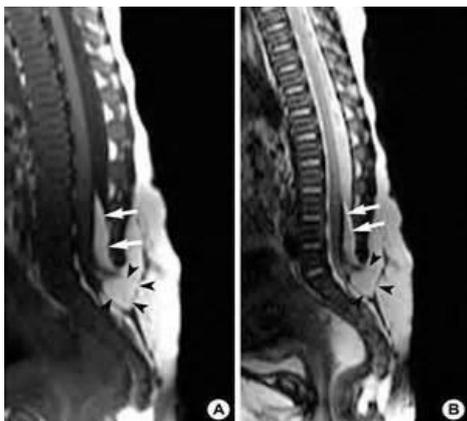


Figure 5: Lipoma with teethered cord – radiological appearance

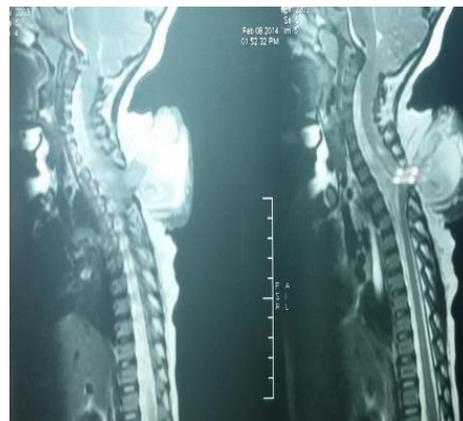


Figure 6: Dorsal meningocele – radiological appearance

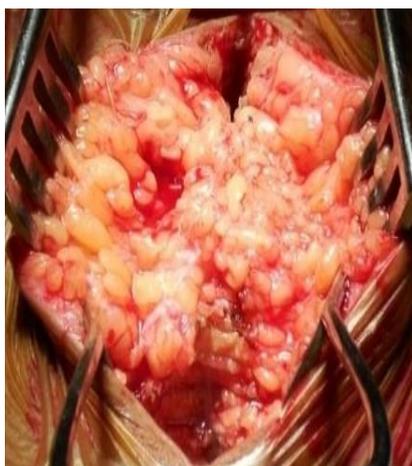


Figure 7: Diffuse lipoma (Intra-op)



Figure 8: Intradural extension of lipoma (intra-op)



Figure 9: Excision of Intradural dermoid cyst:

DISCUSSION

The term “filum terminale syndrome” was first used by Garceau in 1953 in describing three patients.¹ Two decades later, in 1976, Hoffman and colleagues coined the term “teethered spinal cord” to describe the symptoms of their patients with an elongated spinal cord and a thick filum terminale.² Yamada et al.⁴ in 1981 broadened the stretch-induced functional disorder to include patients with other anomalies, such as myelomeningocele, lipoma, lipomeningomyelocele, diastematomyelia, meningocele, and dermoid sinus.⁵

Recently, Yamada and Won described three categories of TCS.^{4,5} The first was lumbosacral cord anchored by an inelastic filum, and the second category included caudal as well as many sacral myelomeningoceles. The third category was divided into two groups; the first was paraplegia associated with lipomeningomyelocele and myelomeningocele, apparently with no functional lumbosacral neurons. No neurologic benefit is expected from surgery in this group. The second group included asymptomatic patients with an elongated cord and a thick filum. These patients require close observation for subtle symptoms, particularly incontinence, which if untreated might quickly become irreversible. The most frequent spinal dysraphism associated with TCS in our study was lipomeningomyelocele (52.5%), followed by secondary to myelomeningocele, which constituted 17.5% of cases and was more prevalent than the 3–15% reported in the literature.^{6,7}

The presenting symptoms in this series were similar to those described by Herman et al.⁸ weakness, deterioration in gait, scoliosis, orthopedic deformities, and urinary incontinence. It has also been recommended that any infant with a mid-line lumbar cutaneous abnormality, such as a hemangioma, lipoma, hair patch or dimple, be evaluated to rule out tethering of the spinal cord.⁹

The slow rate of clinical deterioration usually contributes to the delayed recognition of TCS in patients with myelomeningocele. The preexisting neurological impairments also make subtle neurological changes difficult to detect. Detailed neurological, orthopedic and urological (urodynamic studies) examination and a heightened awareness of the possibility of teethered cord will lead to earlier detection.

Therefore, patients should be followed-up by a multidisciplinary team consisting of specialists from neurosurgery, orthopedic surgery and urology. It is a misconception that myelomeningocele is a static lesion and those active problems are confined to childhood. The inherent complications require early detection and intervention and justify a follow up protocol for life in susceptible patients if additional handicaps are to be prevented.¹⁰ Surprisingly, in 18–54% of TCS patients the conus medullaris is at the normal level.¹¹⁻¹⁴

The diagnosis of TCS can be a normal level, especially in the absence of the typical clinical features of the syndrome. Therefore, a patient who has the signs and symptoms of teethered cord in the presence of normal radiology should be referred to a specialized center to confirm or exclude the diagnosis. Despite its frequent asymptomatic course, the diagnosis of a congenital lumbosacral lipoma, and more general of a closed spinal dysraphism, implies a periodic, multidisciplinary follow- up for life.¹⁵

Yamada and Lonser¹⁶ listed magnetic resonance imaging clues to aid the diagnosing TCS:

1. Thick filum terminale (>2 mm in diameter) or the presence of structures that suggest the lack of viscoelasticity (e.g. a fibroadipose filum terminale, obliteration of the subarachnoid space suggesting adhesion around the caudal spinal cord or nerve roots, a dermoid or epidermoid mass, myelomeningocele, or lipomeningomyelocele);

2. An elongated spinal cord;

Posterior displacement of the conus medullaris with the filum pressing against the thecal lining at or near the L5 lamina seen on sagittal and axial sections, which is a constant finding. A capacious sacral subarachnoid space is not uncommon. In patients who were asymptomatic during childhood, but developed symptoms of TCS after adolescence, the only imaging finding may be a posteriorly displaced conus medullaris and filum terminale.¹⁶ A patient’s lower urinary tract should also be evaluated. Urodynamic studies are sensitive indicators of lower tract compromise when interpreted properly. When available, this would be the preferred examination. Follow-up urodynamic studies are often the first indicator of re-tethering effects, perhaps before the patient is aware of any changes in function and certainly before any changes can be found in the neurological system.¹⁷ The possibility of re-tethering after the initial operation has generated much attention. Various suggestions and techniques for minimizing the incidence of re-tethering have been proposed, but none has proved to be completely effective or superior.¹⁷ Certain high-risk patients should be evaluated prospectively for TCS. Nogueira et al.¹⁸ recommended that all patients with VACTERL syndrome (Vertebral abnormality, Anal imperforation, Cardiac malformation, TracheoEsophageal fistula, Renal abnormalities, Limb deformities, OMIM No. 192350) should be evaluated prospectively for TCS. Mitsuka et al.¹⁹ recommended that if a twin or sibling of an affected person has appropriate symptoms with stigmata of spinal dysraphism, he or she should undergo investigations early.

CONCLUSION

The challenge that faces neurosurgeons does not lie in the technical aspects of sectioning the terminal filum but in correctly identifying which patients have TCS, which patients are at risk for TCS and future neurological deterioration, and which of these patients would benefit from surgical intervention.

If this problem is identified early and investigated properly using rado-imaging techniques, definitive neurosurgical intervention can be planned which would halt the disease process and correct neurological abnormality.

Multi-disciplinary team approach comprising of neurosurgeons, neurophysicians, orthopedicians, urologists, radiologist and physiotherapists is required for proper management of these cases. Timing and need for surgical intervention should be individualized based on clinical features, age of presentation and radiological abnormalities. Symptomatic onset or aggravation of preexisting patient’s complaints could be anticipated for the majority of TCS in school-aged children, adolescents, and young adults. The surgical outcome is excellent for the resolution of pain and sensory motor deficits, but disappointing for bladder dysfunction. Early diagnosis and adequate surgical release might be the keys to a successful outcome in school-aged children, adolescents, and young adults with TCS.

Rehabilitation of these patients by regular neurological examination during follow-up, corrective orthopedic surgeries and appropriate regular physiotherapy would restore them to normal independent day today activities.

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