

# The Management of Tethered Cord Syndrome complicating Occult Spinal Dysraphism in Children: Experience from a Spina Bifida Clinic

*Kanishka Das, Attibele Mahadevaiah Shubha*

*Department of Neonatal & Pediatric Surgery, St. John's Medical College, St. John's National Academy of Health Sciences, Bangalore, Karnataka, India.*

**Abstract:** *The management of tethered cord syndrome complicating occult spinal dysraphism in children is a widely debated topic. The pathology is occult and the pathogenesis is dynamic. The management is sometimes controversial, especially for entities like a fatty filum where the natural history is unclear. However, precise imaging and timely surgical intervention in select cases can prevent the occurrence or arrest the progression of neurologic deficits. This article reviews the current understanding of the syndrome and presents a cumulative experience of managing 36 such cases at a multidisciplinary spina bifida clinic.*

## INTRODUCTION

The term "Tethered spinal cord" was first coined by Hoffman<sup>1</sup> in 1976 to refer a low lying conus medullaris with a thickened (> 2 mm diameter) filum; however the term "spina bifida occulta" (Virchow, 1875) and the first successful surgical intervention for a tethered spinal cord (Jones, 1891) have been described almost a century prior to this. Tethered Cord Syndrome (TCS) refers to progressive neurological deterioration in the functions of the lower spinal cord resulting from traction on the conus medullaris<sup>2</sup>. Though generally associated with a low lying conus (the 'classic' type), TCS is also seen in a normally positioned conus<sup>3</sup>. 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. 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 tethered cord syndrome in pediatric occult spinal dysraphism (OSD). The authors also present a cumulative experience of managing 36 cases of OSD with TCS over twelve years at a multidisciplinary spina bifida clinic in India.

## PATHOPHYSIOLOGY

The caudal spinal cord develops from secondary neurulation; subsequent embryologic events include merging with the cranially developing cord (primary neurulation), canalization, regression and

eventual ascent of the conus due to relatively faster growth of the vertebral canal compared to the neural tube. The final position of the normal conus is anywhere between the midlevel of T<sub>12</sub> to the lower portion of L<sub>3</sub>, but generally at the L<sub>1</sub>-L<sub>2</sub> disc interspace. The ascent of the conus from L<sub>2</sub>-L<sub>3</sub> to the average adult level of L<sub>1</sub>-L<sub>2</sub><sup>4</sup>, previously thought to occur by 3 months of age, probably happens as early as the 40th postmenstrual week<sup>5</sup>.

The filum terminale gently anchors the conus to the sacral bony vertebral canal and suspends the entire lower cord in the buoyant cerebrospinal fluid. A taut (short fibrous) or a thick (fat infiltrated) filum is inelastic and the resultant tension on the conus with spinal movements leads to neuronal dysfunction. Similarly when the cord movement is limited by anomalous fixity to a dorsal (e.g. lumbosacral lipoma, dermal sinus tracts, congenital inclusion tumours like dermoids and epidermoids) or ventral structure (e.g. bony diastem, neurenteric cyst), repeated mechanical shocks have been hypothesized to cause neurological deterioration. A diminution in the microcirculation to the stretched cord has been observed. At the cellular level, ischemia, hypoxia and impaired oxidative metabolism lead to abnormal neuronal transmission<sup>6</sup>. The process of tethering is a dynamic culmination of several mechanical and biological factors such as relative linear growth of the spinal cord and its bony canal, non-neoplastic growth of fat in the fatty filum and lumbosacral cord lipomata, progressive accumulation of putty material in congenital inclusion tumours and the ensuing chemical meningitis and exogenous infection tracking along a dermal sinus tract. The lower motor neuron (LMN) symptoms are considered to be an effect of mechanical compression while the upper motor neuron (UMN) symptoms apparently result from ischemia and stretch. The large fibers of the tracts cranial to the site of tethering are the most susceptible to injury<sup>7</sup>. All these are reflected in the diverse presentations of TCS in our series. (Table 1).

Tethering of cord is theoretically possible at two points – one at the site where an extrinsic lesion such as a dermal sinus tract traverses through the dura and another at the caudal attachment of the conus to a taut or fatty filum. Either or both tethers may be present in any given case and they need to be individually addressed. In the 36 cases reviewed here, 31 had an extrinsic tethering lesion and 5 had

**Correspondence:** *Dr. Kanishka Das, Professor and Head, Department of Neonatal and Pediatric Surgery, St. John's Medical College, Bangalore, PIN -560034, India. e-mail : kanishkadas@hotmail.com*

an isolated fatty filum. Only 2/31 had a taut filum in addition to extrinsic mass lesion.

**Table 1:** Clinical profile of 36 children (neonate -13 yrs) with occult spinal dysraphism and tethered cord syndrome.

Clinical feature	n
<b>Cutaneous stigmata (28/36)</b>	
Fatty mass - lumbosacral	20
Hypertrichosis	9
Dermal sinus	4
Deviated gluteal furrow	4
Multiple neurocutaneous markers	7
<b>Neurologic deficits (23/36)</b>	
Incontinence – urinary /fecal	9
Hydroureteronephrosis, UTI	5
Paraparesis	4
Limb deformity/ gait disturbance	3
Foot , trophic ulcers	3
Recurrent fever, meningitis	2

## ANTENATAL DIAGNOSIS

Antenatal detection of spina bifida occulta without a surface protuberance is rare. A low association of elevated biochemical markers (alpha fetoprotein, acetyl cholinesterase) and type-2 Arnold-Chiari malformation also explains the low antenatal detection of OSD. Nevertheless, antenatal diagnosis has been documented even for dermal sinus tracts<sup>8</sup>. About one-fifth of our cases, mostly lumbosacral lipoma, had been diagnosed in the late second trimester on sonography. None had had hydrocephalus or type-2 Arnold-Chiari malformation. In the event of an antenatal diagnosis, we adopted a realistic counseling as to the probable nature of pathology and the likely neurological deficits. Most of the prospective parents chose to continue the pregnancy and we consistently emphasized on an immediate postnatal assessment for further management.

## CLINICAL FEATURES

OSD may be asymptomatic and go unnoticed. They may be incidentally detected during evaluation for an unrelated disease; they may also present as a cosmetic problem or manifest with subtle or gross neurological signs and symptoms. It is fortuitous when the associated cutaneous stigmata draw attention to the underlying spinal anomaly prior to the onset of neurological impairment. These cutaneous marks are often multiple and they occur in 50-70% of all OSD<sup>9</sup> as compared to 3% in healthy neonates<sup>10</sup>. In our series 80% had cutaneous stigmata and 20% had more than one neurocutaneous marker. Despite the theoretical possibility of early diagnosis based on neurocutaneous markers, many of our patients presented quite late with established neurological deficits of varying degree! (fig.1) The manifestations of TCS in OSD include variable degree of pain, sensorimotor deficits, orthopedic deformities and bladder and / or bowel dysfunction. Often a patchy, asymmetric combination of UMN (spasticity, hyper-reflexia) and LMN (atrophy, hypo-reflexia) symptoms referable to the conus and the lumbosacral nerves occurs. Pain is rarely recognizable in infants or young children. Motor deficits are more evident than sensory aberrations. A few children typically present with painless ulcerations of the foot or leg (trophic ulcers)<sup>11</sup> Recurrent meningitis is common in children with an untreated



**Fig. 1:** Multiple neurocutaneous markers in OSD – hypertrichosis and deviated gluteal fold.

discharging dorsal dermal sinus tracts. Progressive orthopedic deformities (foot deformities, limb length discrepancies, scoliosis, gluteal asymmetry, gait disturbances) are usually reported in 75- 90% of patients. Urological abnormalities are generally subtle (frequency, urgency) but may be overt (incontinence, urinary infections) in established disease. Besides urinary infections, such symptoms usually become obvious only beyond infancy<sup>12</sup>.

Evidence of TCS may be clinically absent at presentation. Recognition of TCS on specific investigations may predate clinical manifestations by a variable period ranging from months to years. Evolution of clinical features may be gradual or sudden. The type of neurological deficit (LMN Versus UMN) and the sequence of their appearance are unpredictable. The prevalence of the various symptoms is similar in both the classic (low conus) type and normal conus type of TCS; however neurogenic bladder is more likely with the former.

Objective documentation of details of the first and subsequent neurological examinations is important to identify worsening of neurological deficit. At the spina bifida clinic, additional corroboration by the physical medicine or rehabilitation personnel is also preferred. Video recordings of the voiding pattern, stance and gait are useful. A departure from the normal motor milestones provided clues to a developing TCS in some.

Asymptomatic neurocutaneous markers can be classified into high- and low- risk categories. The former mandates further investigations to identify an underlying OSD and TCS. High-risk neurocutaneous markers include atypical midline lumbosacral dimples (larger than 5 mm, located more than 25 mm cranial to the anal verge, directed cranially), hemangiomas, protruding lesions (masses, hairy patch, tails) and multiple cutaneous stigmata. Low risk stigmata include coccygeal pits, simple dimples, discolorations and deviated gluteal fold<sup>13</sup>. There is no consensus on recommending further neuroimaging in low-risk group. Less than 10% of lumbosacral dimples referred to us as neurocutaneous markers merited further investigation.

## INVESTIGATIONS

A high index-of-suspicion and meticulous clinical evaluation leads to various imaging modalities that are employed to confirm and grade the degree of abnormality in OSD-TCS. Plain Radiography, commonly used earlier, is seldom employed today as it identifies only the gross vertebral defects and provides no information about the spinal cord. A progressive scoliosis is reliably visualized on plain radiography. We often incidentally detected L<sub>5</sub> - S<sub>1</sub> bifid spine during evaluation for an unrelated pathology but any further investigation was individualized. (fig 2)

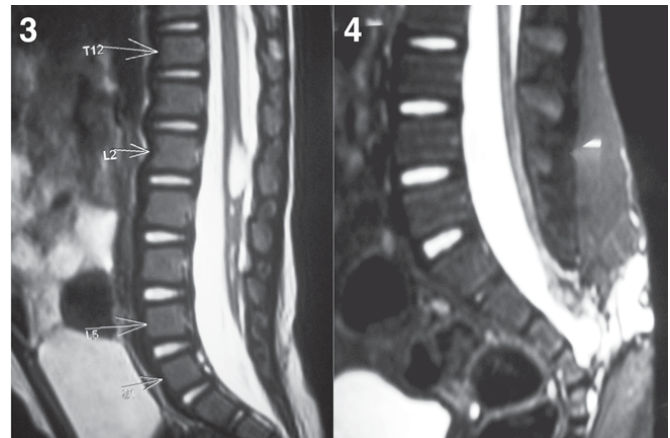


**Fig 2:** X-ray lumbosacral spine showing incidentally detected S<sub>1</sub> spina bifida.

Ultrasonography is a useful tool in the neonate and young infants for evaluation of OSD and TCS<sup>14</sup>. Although ultrasonography is entirely operator dependent, the acoustic window in the lumbar spine is adequate for a detailed evaluation of the meninges, the spinal cord and its movements and the filum terminale. Identifying the level of the conus medullaris is simpler than identifying fat or assessing the thickness of the terminal filum. The acoustic window is usually lost by 4 to 5 months of age after which images can be difficult to interpret. The pulsations of the normal spinal cord with every heart beat are dampened in a tethered cord. The obvious advantages of neonatal spinal USG are the ability to obtain a dynamic view, without the sedation and radiation exposure<sup>9,14</sup>. Ultrasound is a sensitive test for OSD and in experienced hands shows good correlation with magnetic resonance imaging (MRI). We are steadily gaining experience with ultrasonographic assessment of infantile spinal cord and use it to complement MRI. MRI is usually postponed in asymptomatic neonates till 6-12 months of age because of logistic reasons. At all ages, renal USG with standard measurements (e.g. antero-posterior pelvic diameter; ureteric dimensions, bladder wall thickness) helps in detecting a neurogenic bladder and voiding disturbances.

MRI is the modality of choice for an accurate anatomic delineation of the spinal pathology and diagnosis of TCS. It has replaced computed tomography, even in neonates<sup>15</sup>. In symptomatic children, including neonates, we obtained MRI at presentation. Sagittal T1- and T2- weighted images illustrate the level of the conus, while T1-weighted axial images delineate the fat in the terminal filum and facilitate measuring its diameter. In addition, the anatomy and relations of the other forms of OSD are imaged in great detail. (Fig.3, 4). All radiological abnormalities detected on the MRI do not necessarily need surgical intervention. For example, incidental syringomyelia noted in the MRI (Fig 3) requires simple surveillance if it is neurologically stable after cord de-tethering. As with ultrasonography in the young infant, dynamic or phase contrast MRI can evaluate cord motion<sup>16</sup>; but is not popular as it requires anesthesia or sedation.

Simple clinical observations such as observed voiding and reviewing voiding diaries were useful in monitoring urological complications of TCS. Ultrasonographic findings such as hydronephrosis, thickened bladder wall and significant post-void residue prompted further investigations like voiding cystourethrogram (VCUG) and



**Fig. 3 & 4:** Sagittal T2 - MRI of spinal bifida occulta. (3) showing a low lying conus medullaris (L4-5), dorsally displaced cord and a thick taut filum terminale. Also seen is a syrinx at L2. (4) showing a low lying (L4-5), dorsally displaced conus medullaris tethered to a heterogeneous caudal fatty mass. At exploration, the caudal mass was a conus dermoid and the filum was enmeshed in a lipomatous mass that extended extradurally into the subcutaneous plane

urodynamic studies (UDS). The earliest features of subclinical neurovesical dysfunction are detrusor hyper-reflexia and detrusor-sphincter dys-synergia resulting in vesicoureteral reflux and hydronephrosis<sup>17</sup>. Though it was technically demanding and required strict standardization, especially in the neonate, UDS yielded objective data to support surgical intervention in asymptomatic children with TCS. It provided an accurate assessment of improvement in neurovesical dysfunction after detethering.

Other advanced preoperative and intraoperative studies that further characterize the neurological and urological status are electromyography (rectal versus urethral; continuous versus evoked) and somatosensory evoked potential monitoring of the posterior tibial nerve. These are currently experimental and are not routinely used in clinical settings.

## TREATMENT

It is evident that the diagnosis of TCS is both anatomical and pathophysiological and hence a decision of surgical intervention is based on cumulative information from both clinical evaluation and investigative modalities. Decision making is relatively easier in the symptomatic patients with neurologic deficits than in asymptomatic patients. Some entities of the latter group, whose natural history is now well documented, when managed with watchful expectancy inevitably evolve into full blown TCS in due course of time. They include lumbosacral lipomas, dermal sinus tracts, congenital inclusion tumors such as dermoids and epidermoids, diastematomyelia and neurenteric intraspinal cysts. Most of the contemporary experts would concur that these need early surgical correction<sup>18</sup>.

The residual group includes asymptomatic patients with low conus, a fatty filum, a thick filum or a combination of these. Their management is mired in controversies that have been discussed extensively in a recent review<sup>18</sup>. Both conservative and surgical options are propounded in their management. Many believe that sectioning of the filum reverses clinical and urodynamic deficits in a subset of the symptomatic tight or fatty filum terminale; since the procedure has an acceptable minimal morbidity in trained hands,

similar treatment would be preemptive in the asymptomatic cases. However, the diminution in the detrusor hyperreflexia after sectioning of the filum is confounded by a known spontaneous rate of cure with this entity. Like many others, we adopted a middle path approach and maintain close clinical and investigative follow up at our Spina Bifida Clinic. The parents are counseled in detail about this strategy and an informed consent is obtained. At the earliest evidence of clinical or subclinical deficit, we advise de-tethering. There were 5 / 36 children (Age range 2 months-13 years) who were neurologically normal over an average follow-up of 3.5 years (Range 2-11 years). Two children who were originally assigned to conservative management were subsequently withdrawn from the protocol and were operated within a month of onset of neurological deficit; both of them showed complete recovery in the subsequent 6 months.

The fundamental goals of surgical intervention in OSD with TCS (Table 2) are to stabilize or improve neurological deficits in symptomatic patients and to prevent future deficits in asymptomatic patients. It aims at restoring the normal mobility of spinal cord by eliminating the tethering structures and resuspending the freed conus and spinal nerves in the cerebrospinal fluid. We operated the symptomatic and the high-risk categories immediately after a precise diagnosis. The surgical procedure was individualized according to the etiology and specific pathology. For example, in an intramedullary dermoid with a tethering lipoma, it included evacuation of the contents of the dermoid (Fig 5), gentle curettage of the epithelial lining from within, division and debulking of a tethering lipoma and duraplasty in a complex tethered cord. As in any surgery for spina bifida, the accent was on neural preservation and a lax duraplasty around the repositioned cord and nerves. After the detethering of the low conus from an extrinsic tethering element, we reassessed the laxity of the conus or filum, the return of cord pulsations and pulsations of the arterioles on the surface of the cord. If these showed a gross improvement over the pre-detethering state, we did not proceed to divide the filum terminale. In the rest, we proceeded to do so through the same durotomy or through an additional dural exposure at lower level. (Fig 6) Some authors would add this step in all detethering procedures for TCS in OSD. Where the anatomy of the otherwise easily identifiable filum (Fig.6) is unclear, intraoperative electrophysiological studies and nerve stimulation can be used to identify the filum for precise detethering. This time consuming exercise was rarely necessary.

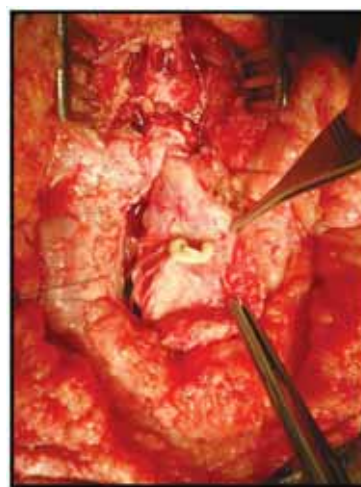
## OUTCOME

The results of carefully selected surgical detethering in both symptomatic and asymptomatic TCS in OSD were gratifying. Neurologic deficits were arrested but any recovery of preoperative deficits was unpredictable. Reported rates of recovery of sensory, motor and neurovesical dysfunction vary widely and reflect non uniform assessment and report<sup>18-20</sup>. A short-duration radicular pain or spasticity is often completely relieved while an established neurovesical dysfunction or orthopedic deformity at presentation is usually irreversible; indeed there is a spectrum in between. In our cohort, only 4/23 with deficits showed a tangible recovery after surgery, two with recent sensorimotor deficits and two others with early neurovesical dysfunction.

In conclusion, the management of the tethered cord syndrome in neonates and children with occult spinal dysraphism involves a multidisciplinary comprehensive approach with antenatal counseling, postnatal evaluation and vigilant follow-up at a spina bifida clinic. Judicious investigations and their interpretation followed by timely,

**Table 2:** Management of tethered cord syndrome in 36 children with occult spinal dysraphism.

Treatment modality	N
<b>Operative procedures</b>	<b>31</b>
Laminectomy , detethering of cord	31
Excision/ debulking of of cord lipoma / dermoid /epidermoid	11
Excision of bony/ fibrous diastem	5
Division of filum terminale	2
<b>Conservative follow up</b>	<b>5</b>



**Fig. 5:** Operative photograph of the conus dermoid cyst (Fig.4) with putty material extruding dorsally. Note the closely related spinal nerves.



**Fig 6:** Operative photograph of the sacral spinal canal after detethering of the filum imaged in Fig.3. Note the retracted filar ends (with sutures) and the ventrally exiting lowest sacral nerves.

selective surgical management in trained hands facilitate neural preservation in a majority and facilitate recovery in a few children.

## REFERENCES

- 1.) Hoffman HJ, Hendrick EB, Humphreys RP. The tethered spinal cord: its protean manifestations, diagnosis and surgical correction. *Childs Brain* 1976; 2: 145 - 155
- 2.) Hendrick EB, Hoffman H, Humphreys RP. The tethered spinal cord. *Clin Neurosurg* 1983; 30: 457 - 463.
- 3.) Warder DE, Oakes WJ. Tethered cord syndrome: the low-lying and normally positioned conus. *Neurosurgery* 1994; 34: 597 - 600.
- 4.) Barson AJ. The vertebral level of termination of the spinal cord during normal and abnormal development. *J Anat* 1970; 106: 489 - 497.
- 5.) Wolf S, Schneble F, Troger J. The conus medullaris: time of ascendance to normal level. *Pediatr Radiol* 1992; 22: 590 - 592.
- 6.) Yamada S, Knerium DS, Mandybur GM, Schultz RL, Yamada BS. Pathophysiology of tethered cord syndrome and other complex factors. *Neurol Res* 2004; 26: 722 - 726.
- 7.) Cornette L, Verpoorten C, Lagae L, et al. Tethered cord syndrome in occult spinal dysraphism: timing and outcome of surgical release. *Neurology* 1998; 50: 1761 - 1765.
- 8.) Hamill N, Grant JA, Myers SA. Congenital Dermal Sinus. *J Ultrasound Med* 2008; 27: 799 - 802.
- 9.) Bui CJ, Tubbs RS, Oakes WJ. Tethered cord syndrome in children: a review. *Neurosurg Focus* 2007; 23 (2): E2.
- 10.) Powell KR, Cherry JD, Hougden TJ, Blinderman EE, Dunn MC. A prospective search for congenital dermal abnormalities of the craniospinal axis. *J Pediatr* 1975; 87: 744 - 750.
- 11.) Michelson DJ, Aswhal S. Tethered cord syndrome in childhood: diagnostic features and relationship to congenital anomalies. *Neurol Res* 2004; 26: 745 - 753.
- 12.) Iskandar BJ, Oakes WJ. Anomalies of the spine and spinal cord. In: McLone DG (Ed) *Pediatric Neurosurgery: The Surgery of the Developing Nervous System*, Ed 4. Philadelphia: WB Saunders, 2001, pp 307 - 324.
- 13.) Kriss VM, Desai NS. Occult Spinal Dysraphism in Neonates: Assessment of High-Risk Cutaneous Stigmata on Sonography. *Am J Roentgenol* 1998; 12 : 171
- 14.) Ponger P, Ben-Sira L, Beni-Adani L, Steinbok P, Constantini S. International survey on the management of skin stigmata and suspected tethered cord. *Childs Nerv Syst* 2010; 26: 1719 - 1725
- 15.) Schenk J, Herweh C, Gunther P, Rohrschneider W, Zieger B, Troger J. Imaging of congenital anomalies and variations of the caudal spine and back in neonates and small infants. *Eur J Radiol* 2006; 56: 3 - 14
- 16.) McCullough DC, Levy LM, DiChiro G, Johnson DL. Toward the prediction of neurological injury from tethered cord: investigation of cord motion with magnetic resonance. *Pediatr Neurosurg* 1991; 16: 3 - 7.
- 17.) Palmer LS, Richards I, Kaplan WE. Subclinical changes in bladder function in children presenting with nonurological symptoms of tethered cord syndrome. *J Urol* 1998; 159: 231 - 234.
- 18.) Finn MA, Walker ML. Spinal lipomas: clinical spectrum, embryology, and treatment. *Neurosurg Focus* 2007; 23 (2): E10
- 19.) Huttman S, Krauss J, Collmann H, Sorensen N, Roosen K. Surgical management of tethered spinal cord in adults: report of 54 cases. *J Neurosurg* 2001; 95 (2 Suppl): 173 - 178
- 20.) Ostling LR, Bierbrauer KS, Kuntz C. Outcome, reoperation, and complications in 99 consecutive children operated for tight or fatty filum. *World Neurosurg* 2012; 77: 187 - 191.

Emcure<sup>®</sup>

The Expert's Choice...

Matches European Pharmacopoeia's  
Erythropoietin Biological Reference Product

# Cold Chain Maintained at Every Step....