

## Tethered spinal cord syndrome: a developmental overview

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### Abstract

Tethered cord syndrome is a stretch-induced functional disorder associated with the fixation (tethering) effect of inelastic tissue on the caudal spinal cord, limiting its movement. This abnormal attachment is associated with progressive stretching and increased tension of the spinal cord as a child ages, potentially resulting in a variety of neurological and other symptoms. Due to the variation of the growth rate of the spinal cord and the spinal column, the progression of neurological signs and symptoms is highly variable. Some infants are presented with tethered cord syndrome at birth (so-called congenital), while others develop the symptomatology in infancy or early childhood. The majority of these cases are mostly developmental, corresponding to the progressive development of excess fibrous connective tissue (fibrosis) in the filum terminale. The filum terminale is a strand of tissue that bridges the spinal cord tip and the tailbone (sacrum). The inelastic structures in children originated from defective closure of the neural tube (the precursor of the spinal cord) during embryonic development, eventually forming a condition known as spina bifida. Because of its functional (physiological) nature, tethered cord syndrome can be reversible if surgically treated in its early stage.

**Keywords:** Tethered cord syndrome, filum terminale

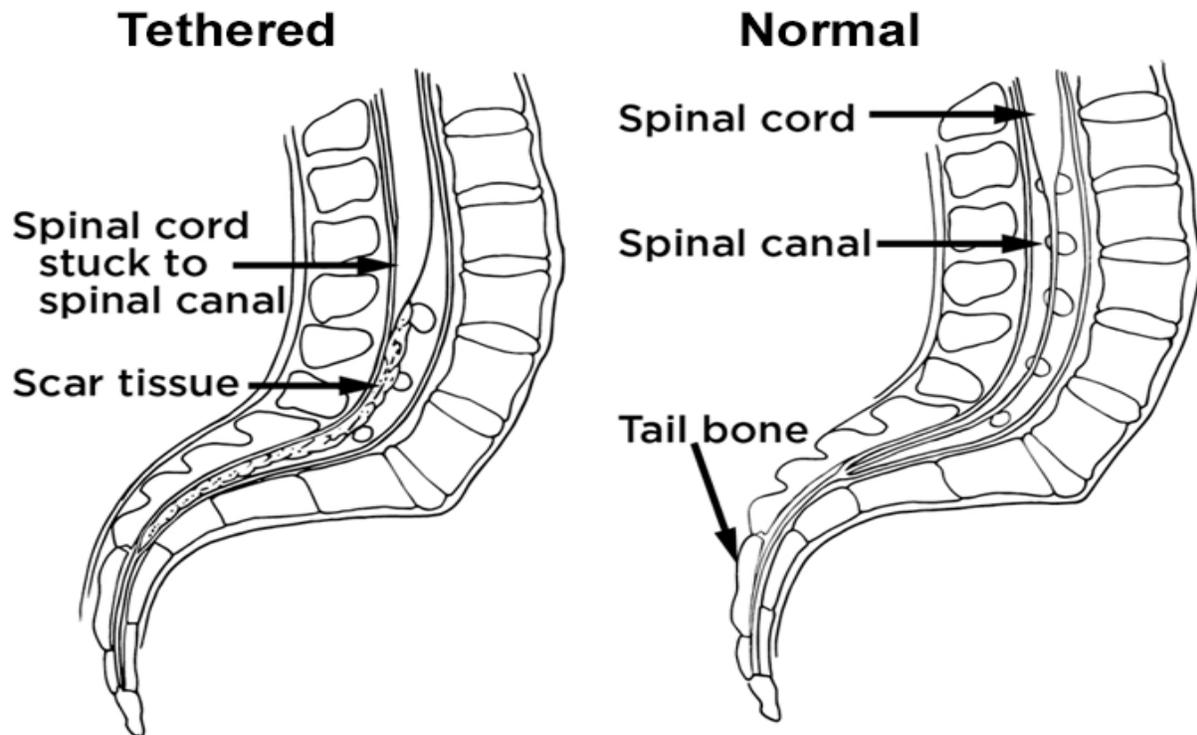
### Introduction

The "cord" in "tethered cord" is the spinal cord<sup>[1]</sup>. The spinal cord is the bundle of nerves that carries messages between the brain and the body<sup>[3]</sup>.

Before a baby is born, the spinal cord is normally the same length as the bones that surround it. These bones are called the spinal column. As the baby grows, the spinal column gets longer than the spinal cord<sup>[1]</sup>. This means the spinal cord has to be able to move freely inside the spinal column. But in

some babies, the bottom end (tail) of the spinal cord is "tethered" or tied down to the bottom end of the spinal column. This is called tethered cord<sup>[4]</sup>.

Tethered cord means the spinal cord cannot move inside the spinal column. As the child grows taller, the spinal cord is stretched. If the nerves are stretched, they may not work properly, and this can cause problems for child<sup>[1]</sup>.



### Definition of Tethered Spinal Cord Syndrome –

Tethered spinal cord syndrome is a neurological disorder caused by tissue attachments that limit the movement of the spinal cord within the spinal column. These attachments cause an abnormal stretching of the spinal cord. This syndrome is closely associated with spina bifida<sup>[4]</sup>.

TCS has been defined as a spectrum of congenital anomalies resulting in an abnormally low position of the conus medullaris that may lead to neurological, musculoskeletal, urological, or gastrointestinal abnormalities.<sup>[2,4]</sup>

### Incidence –

The true incidence of OSD and primary TCS is not known. Unlike open neural tube defects, closed defects such as the TCS are usually diagnosed with the onset of symptoms or found incidentally during workup of unrelated problems<sup>[4]</sup>. The incidence of open neural tube defects has

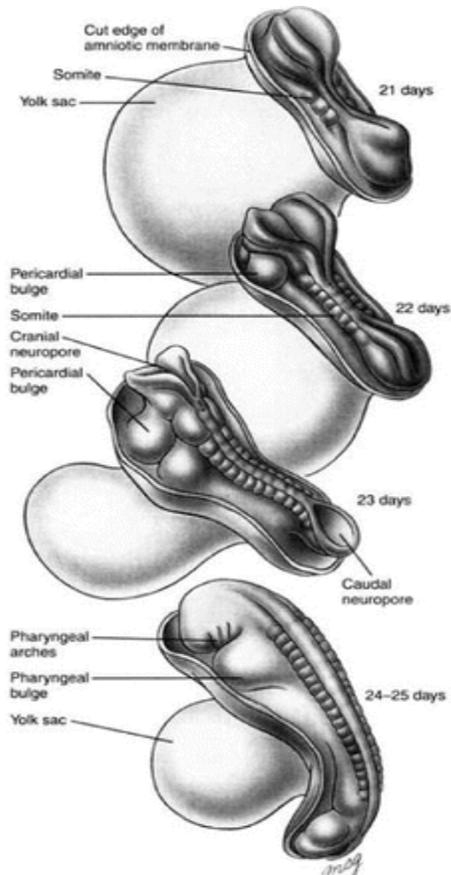
declined dramatically since the introduction of foliate supplementation<sup>[5]</sup>.

### Embryology -

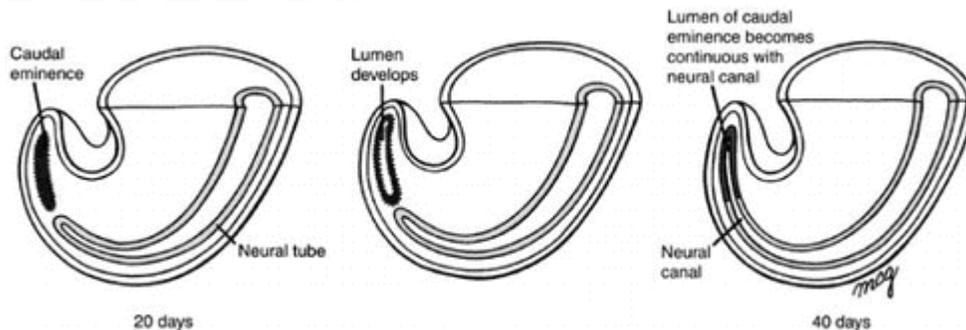
Knowledge of neural embryology is essential to understanding TCS because it is commonly associated with a variety of disorders that are a result of abnormal development of the nervous system. A brief review of caudal nervous system embryology aids in understanding the anatomical basis of TCS (Figs. 1–2 ). The neural tube forms during the process of neurulation, which occurs during Days 18–28 of gestation. Initially, the ectoderm overlying the notochord proliferates to form the neural plate, which subsequently involutes to form the neural folds and then closes to form the neural tube<sup>[6,10]</sup> The process of neural tube closure begins by Day 22–23 and extends cephalad and caudad with the posterior neuropore closing last by Day 25–27.<sup>[10]</sup> Following neurulation, the distal neural tube undergoes canalization. Distal to the posterior neuropore,

undifferentiated cells from the primitive streak form the caudal cell mass. The distal neural tube forms from fused vacuoles that developed from the caudal cell mass. This structure, in turn, develops into the conus medullaris, cauda equina, and filum

terminale. During the end of the canalization period (Days 43–48), the ventriculus terminalis forms at the terminal end of the neural tube near the coccyx, marking the site of the future conus medullaris<sup>[10]</sup>



**Figure 1: Artist's illustration of primary neurulation. The lateral edges of the neural folds meet in the midline and fuse.**



**Figure 2: Artist's illustration of formation of the neural tube inferior to the second sacral level by secondary neurulation. Mesoderm invading this region during gastrulation condenses into a solid rod called the caudal eminence, which later develops a lumen.**

### Pathogenesis -

The spinal cord includes the bundle of nerves that controls leg movement and sensation as well as bladder function. The spinal cord typically divides into small nerve roots at the L2 vertebral body. During development of the spinal cord, tissue and fat, or other body elements that do not belong near the spinal cord can become attached to the spinal cord. Sometimes the tissue prevents the normal development of the spinal cord so that there are problems with urination and leg weakness<sup>[1]</sup>. In most cases, there are no problems at birth. As the body grows, however, the spinal cord then becomes stretched and damaged by the abdominal attachment. This condition is called a tethered spinal cord<sup>[5]</sup>.

### Causes -

The Following situations and conditions can cause a tethered spinal cord:

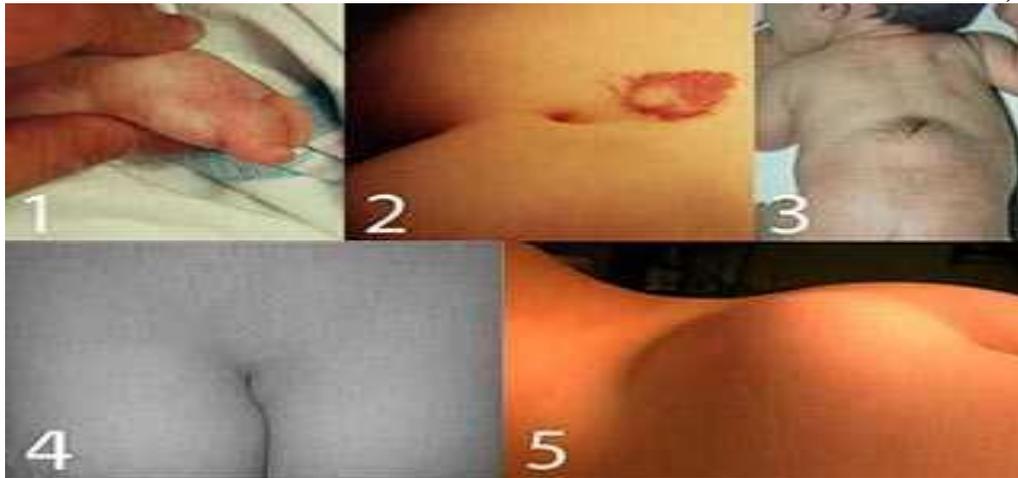
- Dermal sinus tract – scaly channel of tissue along the spinal cord that is prone to infection and other complications<sup>[2]</sup>
- Diastematomyelia – split spinal cord<sup>[2]</sup>
- Lipoma – benign (non-cancerous), fatty growth<sup>[3]</sup>
- Lipomyelomeningocele – birth defect that causes a fatty mass under the skin of a child's back, which may pull on the spinal cord<sup>[4]</sup>. Occult spinal dysraphism is often discovered by cutaneous manifestations such as hypertrichosis, capillary hemangioma, dermal sinus tract, subcutaneous lipoma, or an asymmetrical gluteal cleft. Other manifestations can include leg length discrepancy, foot asymmetry/deformity, scoliosis, neurogenic bladder, frequent urinary tract

infections, upper and lower motor neuron signs, asymmetrical weakness, gait difficulty, spasticity, and back or leg pain.<sup>[10,11]</sup>

- Myelomeningocele (spina bifida) – Patients with MMC are born with a tethered cord and at birth, or shortly thereafter, undergo a repair and closure. Tethered cord syndrome occurs in as many as 2.8–32% of patients with MMC as a result of retethering as the vertebral column grows and lengthens<sup>[4]</sup>
- Spine surgery<sup>[4]</sup>
- Spine trauma<sup>[4]</sup>
- Tumor<sup>[4]</sup>

### Signs and Symptoms –

- A crooked Toe<sup>[3]</sup>( Photo - 1)
- Lesion on the lower back<sup>[3]</sup>
- Fatty tumor or deep dimple on the lower back ( Photo-2)<sup>[3]</sup>
- Skin discoloration on the lower back<sup>[3]</sup>
- Hairy patch on the lower back( Photo- 3)<sup>[3]</sup>
- Back pain, worsened by activity and relieved with rest<sup>[3]</sup>
- Leg pain, especially in the back of legs<sup>[3]</sup>
- Leg numbness or tingling<sup>[3]</sup>
- Changes in leg strength<sup>[5]</sup>
- A crooked crease between the buttocks. (Photo 4)<sup>[5]</sup>
- A lump of the lower back. (Photo 5)<sup>[5]</sup>
- Deterioration in gait
- Progressive or repeated muscle contractions<sup>[5]</sup>
- Leg deformities<sup>[5]</sup>
- Spine tenderness<sup>[5]</sup>
- Scoliosis (curvature of the spine)<sup>[5]</sup>
- Bowel and bladder problems<sup>[5]</sup>



### Diagnosis

If a tethered cord is suspected, one or more tests may be necessary to confirm the diagnosis.

- **MRI:** A diagnostic test that produces three-dimensional images of body structures using powerful magnets and computer technology; can show the spinal cord, nerve roots and surrounding areas, as well as enlargement, degeneration and displacement<sup>[2]</sup>
- **Myelogram:** An x-ray of the spinal canal following injection of a contrast material into the thecae sac; can show pressure on the spinal cord or nerves due to tethered spinal cord<sup>[6]</sup>
- **CT or CAT scan:** A diagnostic image created after a computer reads x-rays; may be used after a myelogram to show how the dye flows around the spinal cord and nerves<sup>[5]</sup>
- **Ultrasound:** A water-soluble gel is placed on the skin where the transducer (a handheld device that directs the high-frequency sound waves to the spine) is to be placed. The gel helps transmit the sound to your skin surface. The ultrasound is turned on and images of the spinal cord moving in the theca sac are obtained.<sup>[5]</sup>

### Surgery

Treatment for a tethered spinal cord usually is surgery to free the spinal cord.

#### •-NEUROMONITORING

This lets them keep watch on the nerves and muscles of the lower part of your child's body. It helps neurosurgeons avoid the risk of further damage to your child's nerves.<sup>[3]</sup>

#### •-LAMINECTOMY

To free the spinal cord, a surgery called a laminectomy. They remove one or more parts of bones in the spine (vertebrae). This lets them reach the spinal cord, the spinal nerve roots and the thecal sac around the spinal cord.<sup>[2]</sup>

Then free the spinal cord by gently cutting, or teasing, it away from the scar tissue or fat. Neurosurgeons use a microscope to help them see the area during the surgery.<sup>[5]</sup>

After the spinal cord is free, neurosurgeons sometimes apply a patch to the covering of the spinal cord (dura mater). This limits the chances cerebrospinal fluid (CSF) will leak.<sup>[5]</sup>

#### Results of Untethering Surgery –

The results of surgical untethering in symptomatic TCS patients are generally favorable, but the extent of improvement varies depending on the preoperative symptoms and deficits.

- Lee et al.<sup>27</sup> reported that pain improved in approximately 80% of patients, neurological improvement or stabilization occurred in 90%<sup>[11]</sup>.

- Guerra et al.<sup>[11]</sup> reported similar results, with 48% improvement seen in pediatric patients with abnormal urodynamics.<sup>[13]</sup> Other studies have yielded higher improvement rates for urological dysfunction, with an average of 87% seen in seven studies having a total of 161 patients.<sup>[14]</sup>

### After the operation Nursing Care

After the operation, your child will spend about two to four hours in the Post Anesthetic Care Unit (PACU). Then child is shifted to the Neurosurgical Unit.

- Child will have a bandage on their back. The nurse will check the bandage often. The nurse will also check your child's temperature, heart rate, blood pressure, breathing and leg movements<sup>[1]</sup>.
- Child will have an intravenous (IV) tube. It allows fluids and medicines to be given directly into your child's bloodstream<sup>[3]</sup>.
- For the first two to three days after the operation, your child needs to lie flat in bed. This is to prevent leakage of fluid from around the spinal cord<sup>[5]</sup>.
- The nurse will give position change ( side to side ) to child , about every two to four hours. This will help prevent sores that may develop from lying in bed. It will also help prevent any chest problems after surgery<sup>[4]</sup>..
- **-FUTURE SURGERIES**

Once a child has had surgery to repair the spinal cord or has had the spinal cord freed up (detethered), there is a chance the cord will attach again as the child grows. Some children need more surgeries; this is more common in children who have myelomeningoceles or lipomyelomeningoceles<sup>[6]</sup>.

### Prevention

- Research has shown that women who take folic acid supplements or add natural folic acid to their diets can decrease risk of

giving birth to a baby with spina bifida. Avoiding spinal injuries or trauma also can prevent tethered cord syndrome<sup>[3]</sup>.

- Children who have a tethered cord but no symptoms should have surgery as soon as symptoms appear, so parents and Health care providers should closely monitor these children<sup>[5]</sup>.
- Some problems with the bladder related to a tethered cord cannot be prevented. Parents can watch for symptoms of neurogenic bladder and help their children to follow all instructions from Health Care Providers to manage problems with controlling urination, including home instructions following surgery<sup>[5]</sup>.

### Conclusions

The challenge that neurosurgeons faces 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 the correct diagnosis is made and timely treatment implemented, one is rewarded with a neurologically normal child with a potential for a healthy life<sup>[3]</sup> We hope that the present review and the experience of our institution shed more light on this topic, but the reader must be aware that there is still much controversy and uncertainty regarding this topic. Therefore, there is a need to continue critically looking at this disease process to obtain better data through randomized prospective studies. The relative safety of the procedure should not be a reason to arbitrarily advise surgery<sup>[12]</sup>

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