Tethered cord syndrome

General information

Abnormally low conus medullaris. Usually associated with a short, thickened filum terminale, or with an intradural lipoma (other lesions, e.g. lipoma extending through dura, or diastematomyelia, are considered as separate entities). Most common in myelomeningocele (MM). Diagnosis must be made clinically in meningomyelocele, as almost all of these patients will have tethering radiographically.

Tethered cord syndrome (TCS), is a neurological disorder characterized by the lower settlement of the conus medullaris.

It is a congenital spinal disease which is caused by split cord malformation, meningomyelocele, and spinal tumors.

There is some overlapping of the naming of these conditions and some of them are cases of closed spina bifida. The various forms include such conditions as: tight filum terminale, lipomeningomyelocele, split cord malformations, dermal sinus tracts , dermoids, and cystoceles.

In general, what all of these conditions have in common is a pulling on the spinal cord at the base of the spinal canal. As children grow, their spinal cords do not grow as quickly as there spinal columns so relatively the spinal cord must be able to freely ascend on the inside of the spinal column during growth. If various abnormal structures are holding onto the spinal cord from below it stretches the spinal cord and this can lead to progressive loss of function.

In general, most pediatric neurosurgeons recommend these conditions be operated on to pr otect the growing spinal cord. For the child that has reached adult height with minimal if any symptoms, some neurosurgeons would advocate careful observation only. The operation is tailored to the cause of the tethering. In general the spinal column is o pened from behind to expose the extent of the sites of tethering of the spinal cord. Often neurosurgeons will have neurophysiologists monitor spinal cord and nerve function during the delicate operation to minimize risk to these structures.

The great majority of children tolerate the surgery well and most improve or at least stabilize with regards to their level of function. There is a potential for the spinal cord to retether as the child gets older and for this reason it is im portant that they be carefully monitored.

The association of urinary, cutaneous, and skeletal abnormalities with spinal dysraphism was recognized in the 20th century. Early in the 20th century, some physicians began to suspect that traction on the conus medullaris caused myelodysplasia-related symptoms and that prophylactic surgical management could prevent the occurrence of clinical manifestations. It was not, however, until later in the 20th century that the term “tethered spinal cord” and the modern management of TCS were introduced. This gradual advancement in understanding at a time before the development of modern imaging modalities illustrates how, over the centuries, anatomists, pathologists, neurologists, and surgeons used clinical examination, a high level of suspicion, and interest in the subtle and overt clinical appearances of spinal dysraphism and TCS to advance understanding of

pathophysiology, clinical appearance, and treatment of this entity. With the availability of modern imaging, spinal dysraphism can now be diagnosed and treated as early as the intrauterine stage.

Classification

The first category includes lumbosacral cord anchored by an inelastic filum.

The second category includes caudal myelomeningoceles and many sacral myelomeningoceles.

The third category is divided into 2 groups.

The first group includes patients with paraplegia and lipomyelomeningocele and myelomeningocele who apparently have no functional lumbosacral neurons. No neurologic benefit is expected from surgery in this group.

The second group includes asymptomatic patients with an elongated cord and a thick filum. They need close observation for onset of subtle symptoms, particularly incontinence, which if untreated becomes quickly irreversible [1)].

see also Tethered Cord Syndrome in Adulthood.

Etiology

The cause of a tethered cord is not always known.

It is sometimes found with spina bifida.

Tethered cord happens when something catches hold of the spinal cord and does not let it move freely. Usually, one of these things catches the cord:

A tight filum terminale. Normally it is stretchy, but sometimes it is tight and tethers the cord.

A scar

A piece of bone can catch a part of the spinal cord.

Fat: Sometimes, fat grows in and around the spinal cord and catches hold of it.

Pathophysiology

An understanding of the underlying pathophysiology of tethered cord syndrome (TCS) and modern management strategies have only developed within the past few decades.

Current understanding of tethered cord syndrome first began with the understanding and management of spina bifida; this later led to the gradual recognition of spina bifida occulta and the symptoms associated with tethering of the filum terminale.

Clinical features

Gait disorder, leg weakness and atrophy, urinary disturbance, foot deformities.

Usually children will complain of pain, if they are old enough or may show some signs of discomfort. As things progress they fail to gain or lose function of the legs, bowel or bladder. Luckily most of the conditions are picked up early due to unusual signs in the middle of their lower backs. These include fatty masses, areas of increased pigmentation, dimples or large collections of hair. When noticed these skin signs should prompt an investigation which usually includes an MRI scan. During infancy an ultrasound may be adequate to identify one of these conditions. Not to confuse things, it should be understood that if there is a problem in one site of the spinal cord, then there may be other problems such as syrinxes. For this reason it is a good idea to image the entire spinal cord and potentially the brain prior to treatment

Diagnosis

CISS sequence for imaging TCS can help enhance the overall surgical outcome of the patients ensuring completeness of the surgery. CISS should be routinely performed in the work-up of patients with tethered cord syndrome [2].

Treatment

Tethered cord syndrome treatment

Case series

Tethered cord syndrome case series.

Case reports

Cranial dermoid tumor (CDT) is a congenital benign tumor which is generally located on the midline of the cranium. Even though Tethered cord syndrome (TCS), is highly associated with spinal dermoid cyst, the relationship of CDT and TCS is unusual. Ercan et al. presented a case with an unusual symptom of CDT, motion-dependent pain, and an uncommon togetherness with TCS [3].

1)
Yamada S, Won DJ. What is the true tethered cord syndrome? Childs Nerv Syst. 2007 Apr;23(4):371-5. Epub 2007 Jan 17. PubMed PMID: 17226037.
2)
Sankhe S, Dang G, Mathur S, Muzumdar D. Utility of CISS imaging in the management of tethered cord syndrome [published online ahead of print, 2020 Aug 15]. Childs Nerv Syst. 2020;10.1007/s00381-020-04789-y. doi:10.1007/s00381-020-04789-y
3)

Ercan S, Guzel E, Guzel A. Unusual Clinical Presentation and Association of Cranial Dermoid Tumor and Tethered Cord Syndrome. Pediatr Neurosurg. 2018 Nov 8:1-5. doi: 10.1159/000494087. [Epub ahead of print] PubMed PMID: 30408795.