Key Points

1. Occult tethered cord and TCS surgery are controversial topics in their relationship to Chiari
2. Large study from TCI used symptoms to identify TCS among patients with Chiari and low lying tonsils
3. Found that 63% of patients with low lying tonsils had indications of TCS
4. Found that those identified as TCS did NOT have small posterior fossas, as opposed to classic Chiari
5. TCS patients did tend to have elongated brainstems and wider foramen magnums
6. Performed section of the filum terminale in more than 300 patients
7. Overall 93% of children improved and 83% of adults improved
8. Every patient that improved with surgery showed elongated brainstem on MRI.
9. Not clear if a tight filum can cause cerebellar tonsils to herniate

Definitions

brainstem - base of the brain which connects to the spinal cord and controls basic functions such as breathing and heart rate

conus - cone shaped area at the lower end of the spinal cord

filum terminale - fibrous thread that connects the lower end of the spinal cord to the bony spinal column

foramen magnum - opening in the base of the skull through which the brain and spine connect

lumbar - one of the sections of the spine, the lower back region

morphometric - in this article, refers to measuring dimensions of the skull and brain

TCI Results On Chiari & Tethered Cord Surgery

July 31st, 2009 -- Tethered Cord Syndrome (TCS) is a condition where the spinal cord tissue attaches abnormally to the bones of the spine. The resulting tension causes symptoms such as bladder and bowel incontinence and weakness of the legs. Sometimes skin abnormalities develop over the attachment point, and can be a clue to the underlying problem. TCS is usually treated surgically by "freeing" the cord. Success of the surgery is mixed with many patients experiencing relief from some symptoms but not others.

TCS can be due to a number of different factors, such as spina bifida or fatty deposits, but recently much attention has been paid to the role that the filum terminale plays in tethered cord. The filum terminale is a fibrous thread which connects the very bottom of the spinal cord to the coccyx bone. If the filum terminale is unusually thick, or tight, it can essentially place the spinal cord in traction and pull it down. For children, as their spinal cords grow, they will be pulling up on an anchor that is too strong.

Because the cord is mechanically pulled down, TCS can often be seen on an MRI, which shows that the conus - a lower section of the spinal cord - is abnormally low relative to the bony vertebra. Specifically, the conus is usually located at the L1/L2 level, and MRI evidence that it is lower than this is a strong indication of tethered cord. If a cord is tethered due to the filum terminale, the surgery to correct it is fairly simple; the filum terminale is cut, or sectioned, and the tension on the cord is released.

While the traditional diagnosis of TCS relies on imaging evidence, beginning in 1990, some physicians began to speculate that a subset of patients might be suffering from tethered cords which do not show up on MRI's. Referred to as occult tethered cord, the theory is that even though the conus is at the normal level, the filum terminale is abnormally fatty, thick, or tight, and thus puts the cord under tension. These physicians began to section the filum terminale based on symptoms - such as intractable urinary incontinence - rather than MRI results.

Naturally, since the surgeons were basing their decisions mostly on their own judgment, controversy began to grow over this practice. Conservative surgeons pointed out that there was no clear evidence that these patients had tethered cords and that the risks of surgery were not warranted. More aggressive surgeons pointed to their own track record of success in improving patients' symptoms with the relatively simple surgery.

In addition to confusion surrounding whether occult tethered cord is a true clinical condition, the relationship between occult tethered cord and Chiari is not clear. Given the downward pull of a tight filum, some have proposed that tethered cord syndrome can lead to Chiari. However this view is not widely accepted. Indeed, there is evidence to support both sides of the argument. Ellenbogen documented, via MRI, a child with a clearly fatty and tight filum, who then developed Chiari over time. Although this is compelling, an MRI alone does not mean that the tight filum actually caused the tonsils to herniate. On the flip side, Tubbs found, through a cadaver study, that tension applied to the bottom of the spinal cord dissipates very quickly as you move up the spine, and thus is unlikely to affect the brain. However, Tubbs also found, in a different study, that an unusually high percentage of people with lipomyelomeningocele also have Chiari.

Recently, the controversy over tethered cord and Chiari spilled into the national media and even the legal system, with a number of high profile lawsuits against the Chiari Institute (TCI) and their use of tethered cord surgery. While this publication, and Conquer Chiari, takes no position in terms of the relative merits of any of these claims, TCI recently published their preliminary findings on tethered cord and Chiari in the journal, Surgical Neurology. This provides an opportunity to look at the data they have collected outside of the media/legal spotlight, in a dispassionate manner.

TCI has a patient base unlike any other in the Chiari community and for this study looked at more than 3,000 children and adults seen between 2002 - 2007. They classified each person as having Chiari based on the strict definition of at least 5mm of herniation, or as having low lying tonsils (LLT) if the herniation was less than 5 mm.

To identify tethered cord syndrome, the researchers looked for classic TCS symptoms, such as bowel/bladder issues, low back pain etc., and also laid out a set of tests, of which at least 5 had to be positive to qualify as TCS. Examples of such tests included the presence of neurogenic bladder, symptom relief on a toe walk test, and symptom aggravation on a heel walk test. In addition, detailed morphometric studies were performed, looking at both skull and brain dimensions.

Using the established criteria, they found that a relatively small number of the Chiari group (14%) had indications of TCS, but a majority of the low lying tonsil group (63%) qualified as having TCS. It is important to keep in mind that it is difficult to apply these findings to the general Chiari population because TCI tends to attract more difficult cases. In fact, of the entire study population, nearly half had had at least one previous failed surgery.

occult - a disease or problem that is not readily apparent; in other words can not be seen on images

posterior fossa -region in the back of the skull where the cerebellum is situated

section - to cut

spina bifida - birth defect where the neural tube does not close properly

tethered cord syndrome (TCS) - condition where the spinal cord is improperly attached, or tethered, to the spine

spinal cord - bundle of nerve fibers that runs from the base of the brain all the way down the back, through the bony spine

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

cerebellum - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

Chiari malformation I - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

decompression surgery - general term used for any of several surgical techniques employed to create more space around a Chiari malformation and to relieve compression

syringomyelia - condition where a fluid filled cyst forms in the spinal cord

Source

Source: Association of Chiari malformation type I and tethered cord syndrome: preliminary results of sectioning filum terminale. Milhorat TH, Bolognese PA, Nishikawa M, Francomano CA, McDonnell NB, Roonprapunt C, Kula RW. Surg Neurol. 2009 Jul;72(1):20-35

Regardless, in addition, they found significant anatomical differences between those classified as having TCS and those with traditional Chiari. Specifically, to no one's surprise, those with traditional Chiari were found to have small posterior fossas compared to healthy controls. However, this was not true for the TCS group, meaning that their skulls were similar in size to healthy controls. The researchers did find, however, that the TCS group, on average, had elongated brainstems, wider foramen magnums, and downward displacement of the hindbrain.

Out of the TCS group, 74 children and 244 adults underwent sectioning of the filum terminale with generally favorable results (Table 2). Although only a small group had complete resolution of their symptoms, overall 93% of children and 83% of adults did improve after surgery. Follow-up imaging showed that in many cases the conus moved up after surgery, indicating the spine was being pulled down. In addition, they found that the brainstem shortened and the cerebellar tonsils moved up. Interestingly, every patient with a positive surgical outcome had an elongated brainstem prior to surgery.

The authors readily admit that it is not clear why if the filum is putting traction on the spine sufficient to pull down the brain that the bottom part of the spine, the conus, would be in a normal position. But they do point out that since the conus consistently moved up after surgery, that it is likely it was under traction.

The authors further point out that the best way to treat patients with Chiari and indications of TCS is still to be determined. Based on their experiences to date, they only section the filum if there is evidence of brainstem elongation and other indications the brain is being pulled down. Further, if there is a large herniation, they first perform a posterior fossa decompression before considering TCS surgery.

It is important to note that the TCI publication contains a good deal more data, for example involving syrinxes and scoliosis. Due to space limitations, it is recommended that the interested reader go directly to the publication (see Source) for more details.

The occult TCS controversy, especially as it relates to Chiari, is likely to continue for some time. One way to address the issue head on would be to develop a non-invasive technique to measure the tension of the filum in these types of patients to see if they are indeed pulling down on the spine.

Table 1: Selected Symptoms and Signs of 318 Patients Undergoing Section of the Filum Terminale

	Children (%)	Adults (%)
Total	12	88
Related to TH		
Suboccipital Headache	82	90
Posterior Neck Pain	78	81
Dizziness	60	67
Nausea, Vomiting	49	48
Related to TCS		
Low Back Pain	77	90
Leg Pain	82	82
Urinary Problems	93	73
Leg numbness, weakness	70	72
Muscular Atrophy	19	25
Bowel Problems	64	68
Pelvic Numbness	38	69

Note: TH = tonsillar herniation; TCS = tethered cord syndrome

Table 2: Surgical Ouctome (Section of the Filum, 318 Total)

	Children (%)	Adults (%)
Resolved	36	18
Improved	57	65
Unchanged	7	16
Worse	0	1

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