Key Points

1. Tethered Cord Syndrome is where the spinal cord is abnormally attached to the bony spine
2. The traction results in bladder/bowel problems and weak legs
3. Traditionally defined as a low-lying conus on MRI
4. Since 1990, some doctors have speculated that TCS may exist without MRI evidence due to a thickened and tight filum terminale
5. Connection to Chiari is unclear, but recently some Chiari experts are diagnosing occult TCS more frequently in Chiari patients
6. Surgery for occult TCS is to cut the filum terminale; indications for this surgery are very controversial
7. Survey of nsg's shows disagreement over whether surgery should be performed without MRI evidence
8. Published surgical results are generally positive, but are poorly designed and represent only a small number of patients
9. Without stronger scientific evidence, it is difficult for patients to evaluate this procedure

Definitions

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

encopresis - involuntary bowel movement

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

incontinence - inability to control urination

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

Controversy Surrounds Occult Tethered Cord Syndrome

September 20, 2006 - 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.

The controversy surrounding surgery for occult tethered cord was highlighted recently in the May, 2006 issue of the Journal of Neurosurgery: Pediatrics, which published several papers on the subject. The papers were based upon a professional society meeting of neurosurgeons in December, 2004 which discussed and debated the subject, and included the results of a survey (Steinbok et al.), and selected surgeons arguing for (Selden) and against (Drake) surgery.

Before diving into the details of these reports, it is worthwhile to talk about why this is important to the Chiari community. While it is clear that tethered cord is in the same neighborhood as Chiari - especially with spina bifida patients - in 2005 a Spanish neurosurgeon, Dr. Royo-Salvador, proposed that, in at least some cases, a tight filum terminale actually *causes* Chiari. In other words, he theorizes that in these cases the mechanical traction anchored at the base of the spinal cord pulls the dura tight and forces the cerebellum to herniate. He went on to publish his results in sectioning the filum terminale of Chiari patients rather than a decompression surgery.

Unfortunately, the strength of his results were limited by methodological weaknesses, it did raise some interesting questions as to the relationship between Chiari and tethered cord. Many CM/SM patients suffer from bladder and bowel problems and weak legs, so are these symptoms due to Chiari, syringomyelia, tethered cord, or a combination of all three?

While it would be easy to dismiss one report, informal communications with patients and others in the Chiari community have shown that some experts are now actively screening Chiari patients for tethered cord due to a tight filum terminale and performing tethered cord surgery in addition to (or instead of) decompression surgery. Since a tight filum terminale is not necessarily discernible on MRI, the diagnoses are being made - just like for non-Chiarians - based upon symptoms and formal urodynamic testing.

As with the larger controversy, the question then becomes for Chiari patients, is occult tethered cord syndrome for real and should surgery for it be considered? One thing the recent publications on the subject make clear is that there are no easy answers.

At the meeting of pediatric neurosurgeons referenced above, a survey was taken based upon 4 hypothetical case studies. In each case, the patient was the same clinically, but the MRI results were different. Specifically,

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

pathogenesis - the origin of a disease and how it develops

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

thoracic - the middle part of the spine, the chest area

Sources

Sources: Metcalfe PD, Luerssen TG, King SJ, Kaefer M, Meldrum KK, Cain MP, Rink RC, Casale AJ. Treatment of the occult tethered spinal cord for neuropathic bladder: results of sectioning the filum terminale.J Urol. 2006 Oct;176(4 Suppl):1826-30.

Steinbok P, Garton HJ, Gupta N. Occult tethered cord syndrome: a survey of practice patterns. J Neurosurg. 2006 May;104(5 Suppl):309-13.

Drake JM. Occult tethered cord syndrome: not an indication for surgery. J Neurosurg. 2006 May;104(5 Suppl):305-8.

Selden NR. Occult tethered cord syndrome: the case for surgery.J Neurosurg. 2006 May;104(5 Suppl):302-4.

the case involved a 6-year old girl suffering from incontinence with only minor neurological signs. The MRI results varied as follows:

- Case 1: Low-lying conus; thick, fatty filum
- Case 2: Normal conus; normal filum
- Case 3: Normal conus; some fat in filum
- Case 4: Normal conus, thoracic syrinx, no Chiari

For each case, the surgeons were asked whether they thought TCS was present and whether it should be operated on. Not surprisingly, there was near unanimous agreement for surgery with Case 1, due to the presence of both symptoms and radiological evidence.

The controversy, however, came to the forefront with the responses to Case 2. The surgeons were nearly evenly split with 29% recommending surgery, 35% against surgery, and 35% saying they were unsure (see Table 1).

Table 1
Nsg Survey Results For Case With Symptoms But No MRI Evidence

Agree	Not Sure	Disagree
29%	35%	35%

Note: Respondents were asked whether they were in favor of surgery.

Highlighting the reliance many surgeons have on objective tests, the addition of a small MRI finding of some fat in the filum tipped the scales towards surgery for Case 3, with 76% agreeing that surgery should be recommended.

The surgeons were split again for Case 4 with a syrinx but not Chiari, with 43% for surgery, 39% against, and 14% unsure.

Beyond highlighting the deep controversy over surgery for occult TCS, it is interesting to note that these results are reminiscent of a similar survey on decompression surgery which also revealed a wide range of opinions in the surgical community. Based upon the survey results, the authors naturally call for rigorous controlled trials to examine this issue further, and as we will see below, the lack of such trials is one of the major problems with this surgery.

Arguing in favor of surgery for occult TCS was Dr. Nathan Selden, a pediatric neurosurgeon from Oregon Health & Science University, who presented the results from a medical literature review on surgical outcomes. Using search terms such as tethered cord, filum terminale, and voiding dysfunction, Selden searched the literature from 1964 - 2004 for studies which reported surgical outcomes on children with no MRI evidence for, but symptoms of, tethered cord due to a tight filum terminale.

In all, Selden found 7 such studies (including his own experience with 6 patients) representing a total of 161 patients. In support of the pro-surgery position, overall 87% of patients improved after surgery, with improvement referring to bladder problems and based primarily on informal patient reports. For those studies which involved formal urodynamic testing, a significantly lower 63% of patients demonstrated objective improvement. It should also be pointed out that there were no serious complications and only 3 minor complications were reported for the entire group of patients.

Table 2
Overall Surgical Results From Literature Review

# Studies	Total # Patients	% Improved
7	161	87%

Note: Improvement refers to patient reports of bladder/bowel functioning

While these results seem impressive, it is important to keep in mind - as Selden readily admits - that the referenced publications are considered weak scientifically due to methodological problems. In addition to the fact that only 161 patients are represented overall, all the studies were retrospective (meaning the cases were examined after the fact), there was no standard, objective measure of outcome, and perhaps most importantly there were no control groups.

A control group is a scientific device which helps validate the treatment being assessed. A control group would generally receive a standard treatment which the new treatment can then be compared to; or in some cases a

sham treatment. There can be a powerful placebo effect with surgery, and well respected studies (published in the New England Journal of Medicine) have shown that patients with arthritic knees experience an improvement of symptoms even after a mock surgery. A more rigorous study of the filum terminale surgery might randomly assign patients to one of three groups: medication, filum surgery, and fake surgery. That way the results from the groups could be compared directly.

The lack of strong evidence that the surgery is effective was just one of the arguments made by Dr. James Drake, of the University of Toronto, in his paper against surgery for occult tethered cord. Drake also points out that there is an unclear definition of the syndrome, the pathogenesis and natural history are not known, and there are no objective clinical tests.

In terms of definition, Drake highlights that different clinicians use different definitions to determine when occult tethered cord is present. In other words, even setting aside the imaging, there is not one set of symptoms that doctors agree constitute occult TCS, and without such a definition it is difficult to even begin to evaluate the syndrome objectively. One doctor may be very accurate in using his judgment to identify good surgical candidates, but another might not.

In terms of the natural history of occult TCS, Drake argues that some assume, mistakenly, that all cases will progressively get worse, thus necessitating surgery. Yet, according to Drake, there is no evidence that this is the case, and there are even some indications the opposite may be true.

Similarly, no evidence has been published on how a tight filum terminale is supposed to translate into symptoms. While there are theories that the tension disrupts blood flow, the theoretical basis for the occult tethered cord has not yet been developed.

In terms of diagnosing based on symptoms, Drake stresses that the symptoms associated with occult tethered cord are actually very common. Specifically, urinary continence among school aged children is very prevalent, with some reports showing that more than 16% of children suffer from some level of incontinence (and this may continue into adulthood). In addition the neurological signs and symptoms that sometimes accompany the tethered cord can actually have many different causes. Thus, absent an objective clinical test, or at least a more rigorous symptomatic definition, it is very difficult to diagnose occult TCS.

Table 3
Surgical Results Metcalfe Study (36 Patients)

	% Improved
Patient Report Urinary	72
Patient Report Constipation	92
Urodynamic Testing	57

Finally, Drake brings to his argument the underwhelming evidence (as discussed above) to support the effectiveness of the surgery. Thus, he concludes, absent clear scientific evidence, there is the basis for subjecting patients to even limited surgical risk.

While there have been several publications since this debate, they continue to lack the scientific methodology necessary to say conclusively that surgery should be used for occult TCS. A report by Metcalfe et al. in the October 2006 issue of the Journal of Urology is one such study.

The authors report on their experience with 36 children who were referred for sectioning of the filum due to severe bladder and bowel problems. While their results are impressive, with 92% of patients reporting an improvement in constipation and 72% an improvement in urinary problems, the study was retrospective in nature with no clear inclusion criteria, and no control group was used. In addition, it is interesting to note that when objective urodynamic testing was used as the outcome measure, the success rate dropped to 57%. In addition, even the authors point out that they only referred children with severe problems, and those children represented a tiny fraction (.04%) of the total cases they evaluated.

Does this mean that occult tethered cord is not for real? No. Does it negate the idea that some Chiari patients will benefit from sectioning the filum? No. It just means there is no evidence either way and it is important for patients to understand this.

Perhaps stronger magnets will allow MRIs to show a subtle difference in the filum, or even somehow measure its tension; perhaps a randomized, controlled clinical trial will prove conclusively that surgery based on symptoms is worthwhile, but until then this procedure is a lot like many of the alternative treatments discussed in this publication. Namely that individuals may report good results, but the theory behind the treatment is unproven and the evidence of its effectiveness is not conclusive.

However, it's also important to keep in mind that this is not unusual when it comes neurosurgery. The resources simply do not exist to conduct large-scale randomized, controlled trials for every type of procedure. Therefore, surgeons tend to develop techniques based upon their experiences with their own patient groups and from the

reports of other, respected surgeons.

Unfortunately, this leaves patients in a difficult position when facing the dilemma of whether to have surgery; however this is a position all too familiar to Chiari patients, and one that is likely to remain for a long time to come.