MANAGEMENT OF TIGHT FILUM TERMINALE SYNDROME
WITH SPECIAL EMPHASIS ON NORMAL LEVEL CONUS MEDULLARIS (NLCM)

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BACKGROUND
Tethered cord syndrome is classically defined as having the tip of the conus medullaris below the body of L2, instead of at the normal L1-2 disc space level. The syndrome presents most commonly with urinary incontinence. In a patient with urinary incontinence and a low-lying conus medullaris, tethered cord syndrome can be diagnosed without difficulty. However, in a patient with urinary incontinence and a hyperreflexive type of neurogenic bladder, in whom the conus medullaris is found to be at the normal level on magnetic resonance imaging, diagnosing a tethered cord can be challenging.

We hypothesized that patients with hyperreflexive neurogenic bladders have a neurologic basis for their bladder dysfunction, probably secondary to tethering by a tight filum terminale, despite having a normally positioned conus medullaris.

METHODS
We investigated 13 patients with tethered cord syndrome with urinary incontinence. All patients underwent urodynamic tests, somatosensory-evoked potentials (SSEPs), and magnetic resonance imaging. All had normal neurologic exams including lower extremity motor, sensory, and reflex functions.

To minimize disturbance of the normal anatomy, we used a flavotomy approach and transected the tight filum terminale to release the tethered conus medullaris.

RESULTS
We show that urodynamic studies appear to be more predictive of a tight filum terminale than SSEPs. However, pathological SSEP results seem to correlate with poor surgical outcomes.

Ninety-three percent of operated patients were continent by the first post-surgical day. However, disappointingly, almost half lost continence within the first month after surgery.

CONCLUSION
In cases of incontinence with a normal level conus medullaris, but urodynamic studies showing a hyperreflexic neurogenic bladder, we believe that sectioning of the filum terminale often improves continence. © 1998 by Elsevier Science Inc.

KEY WORDS
Filum terminale, hyperreflexive bladder, incontinence.

The three major steps in the embryonic development of the central nervous system (CNS) are neurulation, canalization of the tail bud, and dedifferentiation.

In the neurulation process, which is the first step, the neural plate folds in on itself and with the help of some contractile proteins, the sides meet and fusion takes place, forming the neural tube. Maldevelopment in the neurulation process causes various congenital malformations such as myelomeningocele, meningocoele, lipomyelomeningocele, split cord malformations, and dermal sinus tract. The neurulation process begins in the cervical area and reaches the lumbar region by gestational Day 28. As soon as neurulation reaches the caudal end of the neural placode, it stimulates a group of cells at the caudal end of the neural tube to form the tail bud. The normal process of canalization of the tail bud consists of vacuolization, canal formation by joining of vacuoles, and programmed cell death to form the filum terminale and the caudal end of the medulla spinalis below the level of T12. Errors in the process of canalization of the tail bud cause other congenital CNS malformations such as a thick filum terminale, filar lipoma, and terminal myelocystocele.

In the early stages of embryologic development, the vertebral canal and the spinal cord are of equal length. During development of the fetus, the verte-
The vertebral canal grows faster than the neural tissue and consequently the spinal cord becomes shorter, so that the cord seems to be ascending in the spinal canal. In fact, not only the slower growth rate of the spinal cord, but also the programmed cell death during the tail bud process play a role in the ascendance of the spinal cord in the bony canal. By the time of birth, the spinal cord has reached its adult level in the majority of babies. After birth, the vertebral canal and spinal cord seem to grow at the same rate, because no further rise of the conus medullaris is noted in growing infants [12].

Should an error occur in the development process of the CNS, either during neurulation or in canalization of the tail bud, the neural tissue adheres to other tissues surrounding the neural plate at that point, and as the bony elements of the spinal canal grow at a faster rate than the spinal cord, elongation and tethering of the spinal cord occurs by the time of birth.

Pathological filum terminale, which causes tethering of the cord, is thought to be a result of an error in canalization of the tail bud. Because this process takes place after completion of neurulation, the overlying skin which develops during the neurulation process is normal. Although the defects themselves are actually hidden under the normal skin, signs that can alert the physician to errors in canalization of the tail bud are categorized in four groups: neurologic (weakness of the lower limbs, reflex changes), neurocutaneous (localized hypertrichosis on the back, discoloration of the skin), neuro-orthopaedical (club foot and other deformities of the feet), and neuro-urological (incontinence).

In the conventional definition of a tethered cord, a low-lying conus medullaris on radiological investigation (below the L2 vertebral level) is fundamental [6]. The presence of the signs listed above and a low-lying conus medullaris, especially on magnetic resonance imaging (MRI), are essential to the diagnosis of tethered cord. This condition is easily recognized in patients with a low-lying conus medullaris and thick filum terminale. However, patients with urologic complaints (incontinence) who are found to have a hyperreflexic type of neurogenic bladder on urodynamic studies and a normal-level conus medullaris (NLCM) on MRI present diagnostic dilemmas [11]. Hypertonic bladder suggests a neurogenic bladder dysfunction. The hyperreflexic activity and hyperreflexic contractions of the bladder during urodynamic studies prove the neurogenic nature of the bladder dysfunction and differentiate it from a hypertonic bladder of muscular origin.

In a radiologic study by Tani et al, it was found that the vertebral canal increases its length by 7% in hyperflexion. The last pair of ligamentum dentatum is at the level of T12, which anchors the conus medullaris to the sides of the vertebral canal. The conus medullaris begins below the T12 level and ends at the L1-2 disc space, then continues with filum terminale down to the S2 level where the filum terminale is attached to the anterior sacral canal. Since the length of the vertebral canal increases during hyperflexion, the filum terminale must elongate as well and return to its original length at the end of hyperflexion. This elongation of the filum terminale must be elastic, so that no tethering is conducted to the conus medullaris [10]. If the filum terminale is tight or has lost its elastic properties, the traction effect on the conus medullaris may lead to the development of tethered cord syndrome.

Although a tight and fibrous filum terminale is present at birth, late onset of symptoms can be explained by the cumulative effect of hypoxia on the conus medullaris, which is caused by tethering of the spinal cord. According to Yamada et al, clinical symptoms appear after accumulation of hypoxic damage in the neural tissue [13]. Improvement of blood flow has also been demonstrated after detethering of the spinal cord [9].

**Materials and Methods**

In our series, we had 42 patients with tethered cords attributable to various pathological conditions. Of these, 13 patients whose filum terminales were cut to detether the conus medullaris had NLCM. Our patients were between the ages of 5 and 17; seven were male. Nine patients did not have primary toilet training; the rest were toilet-trained for various periods of time, but then showed deterioration in their bladder functions. All patients had normal motor, sensory, and reflex functions in their lower extremities.

Patients who had complaints of incontinence beyond 5 years of age and failed to respond to anticholinergic treatment as a conservative measure were chosen for further investigation. Urodynamic tests were performed on all patients, with special attention to bladder hyperreflexia [2,3]. In urodynamic tests, compliance and capacity of the bladder, leak point pressure, and uroflowmetry were recorded. Sphincter electromyography was also done at the same time as flowmetry to determine whether there was dyssynergia between detrusor and sphincter muscles. Bladder hyperreflexia was considered to be the major indication for surgical
sectioning of the filum terminale, as reported by Meyrat et al [7].

SSEP examinations were done in all patients and the results were abnormal in some of them. Delay in N22 wave latency, low amplitude, and blockade of conduction were considered to be pathological results.

MRI was done on all patients. All had normal level coni medullari. Although three patients were reported to have a filum terminale thicker than 2 mm, none appeared abnormally thick at surgery.

There are reports of filum terminale sectioning through an opening provided by either limited or total laminectomy of the lower lumbar vertebral arches [5,11]. In our Division of Pediatric Neurosurgery, as an alternative to this approach, we prefer flavotomy without a laminectomy to release the filum terminale. The aim of this approach is to protect the normal anatomical construction in order to avoid further fibrosis and spinal instability. However, in cases of tethered cords secondary to previous myelomeningocele surgery or in cases accompanied by a split cord malformation, large filar lipoma, or terminal myelocystocele, this approach is not suitable because the skin and dural incisions are too small.

The flavotomy approach is begun with a vertical median skin incision between L5 and S1, after which the left paravertebral muscles are dissected subperiosteally. After the ligamentum flavum is dissected from the edge of the L5 lamina under magnification, it is retracted to the cranial side as a flap (Figure 1), and the underlying epidural adipose tissue is pushed away from the surgical field using a cotton ball. A dural incision is made, as small as possible, and the filum terminale is located and cut after bipolar coagulation (Figure 2). The dura is then closed watertight with 7/0 nylon sutures, the epidural fat is replaced, and a flap of ligamentum flavum is laid over the epidural fat.

**Results and Discussion**

To evaluate the success of surgical intervention, we grouped our 13 patients into categories by outcome as follows:

1. Good outcome group: Subjective good (patient states that there is an improvement); objective good (urodynamic studies reveal improvement): SBJ+/OBJ+
2. Fair outcome group: Subjective good (patient states that there is an improvement); objective poor (urodynamic studies reveal unsatisfactory improvement): SBJ+/OBJ±
3. Poor outcome group: Subjective poor (patient states that there is no improvement); objective poor (urodynamic studies reveal no improvement): SBJ−/OBJ−

We had seven patients in the first group (53.8%), two in the second group (15.3%), and four in the third group (30.9%). There was no morbidity or mortality in our series of NLCM patients. These results show that approximately 70% of these patients benefited from the operation; in 30%, the problem still remains to be solved. The main dilemma is still the selection of patients for filum terminale sectioning.

After analyzing the data that we recorded, it was found that surgical outcome was not affected by the duration of symptoms, the existence of a urinary infection, toilet training status, or the age of the patient. Khoury et al took failure to respond to...
conservative treatment as the major indication for filum terminale sectioning [5]. We could not find any correlation between the surgical outcome and response to conservative treatment.

Results of SSEP examinations are said to be helpful in selecting patients for surgery [1,8]. However, we could not find any correlation between SSEP results and surgical outcomes. Our data showed that SSEPs provide more helpful information about the neurophysiological condition of the conus medullaris during the follow-up period after detethering. We do not think that pathological SSEP results can be used as an indication for surgical intervention. Our data suggest that pathological SSEP results may be predictive of poor outcome. In the SBJ–/OBJ– group, three of four patients had pathological SSEPs, whereas only one patient of seven in the SBJ+/OBJ+ group had normal SSEPs.

Johnson and Levy stated that the conus medullaris and spinal cord have axial movements that are synchronous with heartbeat, and postulated that a cardiac-paced cine MRI examination might show cessation of movement of the conus medullaris, which may be a clue to a tight filum terminale [4]. They selected their patients according to cine MRI criteria, and reported that they had a considerable number of patients who did not benefit from the surgical procedure, although cine MRI had revealed a tethered cord. They concluded that further evaluation of this imaging technique is needed to determine strict criteria for diagnosis of a tethered cord. In our series, MRI showed three patients to have fila terminale thicker than 2 mm; however, we did not see any pathologically thick fila at operation. Two of these patients were in the good outcome group; the third was in the poor outcome group. We could not find any correlation between the MRI findings and patient outcome.

The follow-up period in our series is up to 16 months. All patients but one declared that their incontinence was improved on the day after surgery, a 92% good outcome in the early period. Deterioration in bladder function, however, occurred during the first postoperative month in some patients, decreasing the rate of success to 53%. Late deterioration after initial success was very disappointing. The hyperreflexive activity in the bladder muscle layer reminded us of the hyperactivity syndromes of the cranial nerves (i.e., hemifacial spasm, trigeminal neuralgia, etc.), and we hypothesized that the manipulation of the filum terminale might have an effect on the nerves going to the bladder, which in turn caused cessation of this hyperreflexivity for a while, as sometimes occurs in hemifacial spasm. We have tried to record the spontaneous activity of the conus medullaris during these hyperreflexive contractions, but the invasive nature of the examination prevented us from doing so.

After the first postoperative month, no further deterioration was noted among our patients. In the group of good outcomes patients, the overall success rate was 53%, which is still not satisfactory. The patients in the fair and poor outcome groups required urological help—either medical or surgical treatment—after a period of time.

Conclusions

1. Sectioning of the filum terminale in the treatment of incontinence with a hyperreflexive bladder is not the treatment of choice. However, if selected properly, patients will benefit from the section of the filum terminale. This may be accepted by urologists as a supplementary measure in the treatment of incontinence.

2. Pathological SSEP results seem to be predictive of poor outcome after surgery. Surgical intervention before SSEPs become abnormal would be more helpful.

3. According to our information, the indications for surgery are (a) the patient’s chief complaint is incontinence and (b) urodynamic studies reveal a hypertonic bladder dysfunction with hyperreflexive contractions.

4. MRI is meaningful if it reveals a low-lying conus medullaris; however, a normal level conus medullaris does not mean that there is no tethering.

5. If surgery is indicated, the flavotomy approach to the filum terminale is the least harmful procedure.

6. Further studies are needed to understand the role of the conus medullaris in hyperreflexic bladder dysfunction and the neurophysiologic alterations in the conus medullaris caused by tethering.

References


COMMENTARY
The authors present a very interesting series of 13 patients between the ages of 5 and 17 who presented to their institution with incontinence. This was believed to have a neurogenic basis because of urodynamic studies. Despite the conus medullaris lying in the normal position in all 13 of these patients, and being of apparently normal caliber on MRI scan in 10, they had their filum severed. Approximately 70% of the patients had at least subjective benefit from this procedure. No comment is made on the presence or absence of spina bifida occulta on plain X-ray in these patients.

Interest in this approach to a select group of children with urinary incontinence was presumably piqued by an article by Khoury et al published in the Journal of Urology in 1990 [1]. A number of children with occult spinal dysraphism and apparently neurogenic bladders had their filum terminale divided. Of note is the fact that approximately 70% of these patients improved. An MRI was not performed on the patients in that study; however, a CT myelogram was performed and was read as normal in 87%.

Our experience with these patients at our institution has been similar, as is the reported experience [2]. There is evidence that this approach works!

If, indeed, this is a potential treatment for children with incontinence which has not been considered before, then we must ask “What has happened to these children in the past?” It is quite obviously not a new disease. Is there a large population of adults who are incontinent who could have benefited from this type of procedure as children? The prevalence of incontinence in the adult population is estimated to be approximately 1%, so this may be the case.

Why exactly this procedure works needs to be addressed. The authors’ proposed mechanism is pure conjecture. The importance, if any, of occult spinal dysraphism as raised by the Toronto study, needs to be addressed. One cannot ignore these results.

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REFERENCES