Retethering of transected fatty filum terminales

Clinical article

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Object. Untethering of a tethered spinal cord (TSC) by transecting or removing a fatty filum terminale is a relatively simple procedure that can prevent or ameliorate neurological symptoms, and the postoperative prognosis is usually good. Progressive neurological deterioration caused by recurrent tethering has been rarely reported. The authors present their experience in cases in which a sectioned fatty filum terminale has become retethered.

Methods. The authors retrospectively analyzed the surgical results of pediatric patients with fatty filum terminale–TSC treated by transection of the filum. The patients’ charts were reviewed for demographic data, clinical presentation, surgical therapy, and follow-up data.

Results. Of the 225 children who underwent TSC release by sectioning the fatty filum from 1992 to 2005, there were 6 patients (2.7%; 3 males, 3 females) in whom the fatty filum retethered. The mean age at the first diagnosis of TSC was 5.2 years (range 2 months–12.3 years). The mean duration from the first untethering procedure to retethering was 5.4 years. The mean age at the time of retethering was 10.6 years (range 7–17.5 years). Symptoms of retethering were urinary incontinence, low-back pain, difficulty walking, constipation, leg pain, and worsening foot deformity. Patients underwent cystometrography at the time retethering was indicated by increased bladder capacity, large post-void residual volume, decreased bladder capacity, increase in filling pressure, and poor sensation of filling. Magnetic resonance imaging revealed adherence of the rostral stump of the sectioned filum to the midline dorsal surface. All patients underwent the second untethering procedure. Four patients improved neurologically and experienced no retethering thereafter (mean follow-up period 5.5 years). Two patients experienced additional retethering after temporary improvement following the second untethering procedure.

Conclusions. Retethering of the spinal cord is a rare condition occurring after the sectioning of a fatty filum terminale. Awareness of this rare sequela is necessary for appropriate long-term management of TSC caused by a fatty filum terminale. Cystometrography is useful for detecting the lesion and confirming the diagnosis of retethering. (DOI: 10.3171/2010.10.PEDS09550)

Key Words • tethered spinal cord • fatty filum terminale • retethering

Abbreviations used in this paper: MMT = manual muscle test; TSC = tethered spinal cord.

This article contains some figures that are displayed in color online but in black and white in the print edition.
Retethering of the fatty filum terminale

TABLE 1: Clinical findings in the 6 patients with retethering of the fatty filum terminale*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs) at 1st Untethering, Sex</th>
<th>Signs &amp; Symptoms at Diagnosis</th>
<th>Age at Retethering (yrs)</th>
<th>Symptoms at Retethering</th>
<th>Cystometrography Results</th>
<th>FU (yrs)</th>
<th>2nd Retethering</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5, M</td>
<td>difficulty walking</td>
<td>9.0</td>
<td>urinary incontinence, constipation, low-back pain</td>
<td>decreased bladder capacity</td>
<td>0.5</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>4, F</td>
<td>constipation, urinary incontinence, urinary tract infection</td>
<td>7.0</td>
<td>urinary incontinence, low-back pain</td>
<td></td>
<td>2.0</td>
<td>yes</td>
</tr>
<tr>
<td>3</td>
<td>2 mos, F</td>
<td>thoracic congenital vertebral anomalies</td>
<td>7.3</td>
<td>difficulty walking</td>
<td>increased post void residual</td>
<td>9.3</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>12.3, M</td>
<td>low-back pain, rt leg pain, urinary incontinence</td>
<td>17.5</td>
<td>rt leg pain, urinary incontinence</td>
<td>large bladder capacity, poor sensation of filling, increased post-void residual</td>
<td>6.3</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>9, F</td>
<td>difficulty walking</td>
<td>13.0</td>
<td>difficulty walking</td>
<td></td>
<td>3.0</td>
<td>yes</td>
</tr>
<tr>
<td>6</td>
<td>11 mos, M</td>
<td>vertical talus</td>
<td>9.9</td>
<td>low-back pain, worsening foot deformity, difficulty walking</td>
<td>increased filling pressure, increased bladder capacity</td>
<td>6.0</td>
<td></td>
</tr>
</tbody>
</table>

* FU = follow-up.

S-1. The dura mater was incised and the fatty filum was identified by its thickness and pale color. In some patients, neurophysiological intraoperative monitoring was used depending on surgeon’s preference. We monitored free-running and stimulated electromyography activity of the muscles of the lower extremities and external anal sphincter. We stimulated the fatty filum and confirmed that no evoked electromyography activity was observed from the filum. The filum was coagulated using bipolar cautery and sectioned. The samples of 5–7-mm in length were sent for histological confirmation. Postoperatively patients were assessed periodically at outpatient visits. We performed cystometry for bladder function periodically in 4 patients. We also performed the MMT in 4 patients. If the signs and symptoms of the patients suggested the retethering of the fatty filum, MR imaging was repeated. Surgical untethering was performed for retethering.

Results

At our institution, between 1992 and 2005, 225 children underwent release of a TSC accomplished by sectioning of a thickened filum terminale. The mean age at diagnosis was 53.6 months (range 0–209 months), and the mean age at surgery was 56.1 months (range 1–210 months). Indications for the surgery were essentially the presence of a fatty filum. Untethering was performed for symptoms that included bladder dysfunction, constipation, leg/back pain, and scoliosis (144 patients) and for prophylaxis (81 patients). Forty-six patients with a normal-level conus medullaris underwent surgery. Of the 225 children who underwent TSC release by sectioning of a thickened filum terminale, there were 6 patients (2.7%) in whom the fatty filum retethered (3 males and 3 females) (Table 1).

The mean age at the first diagnosis of TSC was 5.2 years (range 2 months–12.3 years). The initial presenting signs and symptoms included urinary incontinence (2 of 6 patients), difficulty walking (2 of 6), urinary tract infections (1 of 6), constipation (1 of 6), low-back pain (1 of 6), leg pain (1 of 6), vertical talus (1 of 6), and thoracic congenital vertebral anomalies (1 of 6).

The mean time between the original untethering procedure and retethering was 5.4 years. The mean age at the diagnosis of retethering was 10.6 years (range 7–17.5 years). Symptoms at retethering were urinary incontinence (3 of 6 patients), low-back pain (3 of 6), difficulty walking (3 of 6), constipation (1 of 6), leg pain (1 of 6), and worsening foot deformity (1 of 6). Four patients underwent cystometry at the time of retethering that revealed increased bladder capacity (2 of 4 patients), large post-void residual volume (2 of 4), decreased bladder capacity (1 of 4), increase in filling pressure (1 of 4), and poor sensation of filling (1 of 4). Three patients had urinary symptoms at the time of retethering, and all 4 patients who underwent cystometry had bladder function abnormalities. Three patients had difficulty walking, and the MMT showed a decrease in muscle strength. In one patient without difficulty walking or lower-extremity weakness, the MMT showed stable result.

As for complications we did not observe any neurological worsening or CSF leakage during the postoperative period. We had 1 pseudomeningocele that required surgical reexploration and dural repair. Four patients improved neurologically and experienced no retethering afterward (mean follow-up period 5.5 years). Two patients experienced secondary retethering. The time between the first untethering and the secondary retethering was 2 and 3 years, respectively. The ages of the 2 patients at the diagnosis of the secondary retethering were 9.3 and 16 years. Symptoms at the second retethering were difficulty walking in one patient and low-back pain and constipation in another. These 2 patients improved neurologically after the untethering procedure, and status was stable in 1 patient at the 4-year follow-up and in the other at the 0.5-year follow-up.
Illustrative Cases

Case 1

This 5-year-old boy presented with difficulty walking. Magnetic resonance imaging revealed an epidermoid tumor at L5–S1 and a thickened fatty filum. He underwent resection of an epidermoid tumor and sectioning of the thickened fatty filum. A primary dural closure was performed.

The patient developed normally, without any functional limitations. His bowel and bladder control were unimpeded.

Four years after the primary tethered cord release, the patient experienced an increase in the frequency of urinary incontinence and he also had several episodes of stool incontinence. He had occasional back pain when he bent or walked long distances. Cystometrography revealed that bladder capacity decreased from 279 ml at present.

Magnetic resonance imaging revealed that the conus medullaris terminated at the lower L-2 level (Fig. 1A). The rostral stump of the cut filum appeared adherent to the midline dorsal dural surface near the level of discontinuity (Fig. 1B). There was no evidence of a recurrent epidermal inclusion cyst.

Through a new L-4 level laminectomy, the filum was noted to be densely adherent to the midline dorsal dura. Through the old L-5 laminectomy, endoscopy demonstrated that the rostral stump of the filum was densely adherent to the midline posterior dura (Fig. 1C). The fatty filum was sectioned at the L-4 level. At 4-month follow-up, cystometrography revealed improvement and the patient had urinary bladder continence. Postoperative MR imaging revealed the conus to be at the same segmental level, with rostral stump of the filum apart from the dura.

Case 2

This 4-year-old girl presented with a history of constipation since infancy, episodes of urinary dribbling accidents, and urinary tract infections. She also had pains in her knees and legs bilaterally. Magnetic resonance imaging revealed a slightly low-lying conus at the bottom of L-2 with a fatty filum. The filum was sectioned, and primary dural repair was performed. The patient developed normally and she had no longer suffered urinary incontinence or constipation.

Five years after the primary TSC release, the patient returned with complaints of urinary urgency, incontinence, and intermittent low-back pain. Magnetic resonance imaging showed a persistent fatty filum, which terminated at the previous L5–S1 laminectomy site. We confirmed the previous sectioning of the filum by a segmented absence of fatty filum (Fig. 2A–D). The proximal end of the sectioned filum was attached to the dorsal dural sac in the midline.

We extended the previous L5–S1 incision superiorly, and a laminectomy of the lower half of L-4 was performed. The proximal end of the residual fatty filum was stuck to the dorsal surface of the dura (Fig. 2E), which was released and sectioned. Postoperatively the patient’s urinary incontinence resolved and her low-back pain improved. However, 2 years after the second procedure, she began to suffer low-back pain and constipation. Magnetic resonance imaging revealed more prominent fatty tissue, which appeared to adhere to the dorsal dura at the L-4 level. A laminectomy of the upper portion of L-4 was performed. An arachnoidal adhesion to the filum was observed and dissected away from it. At the 3-month follow-up the back pain and constipation had improved.

Discussion

The surgical treatment of a TSC caused by a fatty filum terminale is a relatively straightforward procedure that usually achieves good results and minimal morbidity. During the release of a fatty filum, the pia mater of the conus medullaris remains intact, the circumference of the common dural tube is unaltered by primary dural closure, and there is usually sufficient space between the spinal cord and dorsal dural sac. Thus, retethering of the cord has been considered to be very rare. In our series, we found that 6 (2.7%) of the 225 patients who underwent fatty filum sectioning had recurrent tethering. The ages at the retethering (mean age 10.6 years) were typical for those of patients with tethered cord syndrome and were seen during the growth spurt, as previously described. The symptoms at recurrent TSC involving a fatty filum were similar to those of original tethering. They included urinary incontinence, low-back pain, difficulty walking, constipation, leg pain, and worsening foot deformity.

Three patients (50.0%) had urinary symptoms at the time of retethering, and all 4 patients who underwent cystometry showed bladder function abnormalities. The prevalence of neurogenic bladder dysfunction in the TSC syndrome is high. Sometimes abnormality on cystograms can be the only sign of retethering, without any neurological deficits, in the case of lipomyelomeningocele. In our series one patient had no apparent urinary symptoms, but cystometry revealed an increased...
post-void residual volume. In 51 of 225 patients who underwent fatty filum sectioning we performed postoperative cystometry. We performed the first postoperative study at 2–6 months after the surgery. We repeated it when a patient developed bladder symptoms or approximately on a 6-month to yearly basis. Seven patients had abnormal studies that were unchanged since the preoperative study. Seven patients had abnormal studies that improved over preoperative studies. In 31 patients the studies showed normal findings. Six patients had abnormal studies, the findings of which deteriorated compared with previous one; in 4 of these patients there was retethering of a fatty filum. The other 2 patients underwent cystometry at 2 months after surgery, and subsequent studies showed improvement. It might be that cystometrography can show the abnormal result and not be so reliable shortly after surgery. Overall, cystometrography is considered to be effective in detecting and confirming recurrent tethering of a sectioned fatty filum. It can be particularly useful in the increased linear growth phase of adolescence for detecting retethering early enough to allow reversal and prevention of symptom progression.

A low-lying conus medullaris, the standard radiographic finding of a primary tethered cord, is not diagnostic of recurrent tethering. A more reliable indicator of recurrent tethering is the obliteration of the normal subarachnoid space between the dural tube and neural elements. In both our series and a previous case report, the filum was retethered to the dorsal sac of the dura at the midline. Magnetic resonance imaging characteristically revealed the filum stacked to the dorsal dura at the midline and subarachnoid space was obliterated. There are 31 children who did not experience retethering and had postoperative MR imaging. Of these 31 patients, MR imaging demonstrated the filum attached to the dorsal dura with obliteration of the subarachnoid space in 5 patients (16.1%). In 26 patients (83.9%), the proximal filum was not attached to the dorsal dura, and there was subarachnoid space surrounding the filum. Therefore, we could say that MR imaging findings of the filum attached to the dorsal dura with obliteration of the subarachnoid space are characteristic of a recurrent tethering; however, it is not strictly specific for it.

It is generally important to prevent a recurrent tethered cord to keep the neural elements within patent subarachnoid space circumferentially. In the case of sectioning a fatty filum, keeping the sufficient subarachnoid space around the cord is usually accomplished by primary dural closure. Some authors advocate using inert material such as a Gore-Tex membrane between the dura and neural elements or expanding the canal with an inert fixed shape-holding material in the case of lipomeningocele or myelomeningocele. However, these are not considered to be practical for the cases involving a fatty filum. The other commonly recognized surgical principles in preventing retethering are minimizing bleeding into the subarachnoiditis and avoiding injury and coagulation of the pial surface to decrease the adherence of pia to surrounding tissues. These should be taken into consideration also in the case of a fatty filum. The CSF leak and postoperative meningitis contribute to the retethering; however, none of our patients had these complications after the initial surgery.

We demonstrated the efficacy of surgical untethering for recurrent tethering of the spinal cord by a fatty filum. All the patients improved neurologically after the procedure. Four patients (66.7%) remained stable afterward (mean follow-up period 5.5 years). We suggest the surgical untethering is the first-line treatment for retethering of a sectioned fatty filum. Basically we extended a skin incision rostrally from the previous one. An additional laminectomy was performed, and intact dura was exposed and cut. The more rostral part of a fatty filum was exposed, released from the surrounding tissues, and sectioned.

Conclusions

Retethering of the spinal cord is a rare condition after the sectioning of a fatty filum terminale. However, awareness of this rare sequela is necessary for appropriate long-term management. We advocate surgical untethering as the first-line treatment for this condition. Cystometrogra-
phy is considered to be useful to demonstrate the underlying neurogenic bladder due to retethering.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Tomita, Ogiwara, Alden, Bowman. Acquisition of data: Ogiwara, Lyszczarz, Alden, Bowman. Analysis and interpretation of data: Ogiwara, Lyszczarz, Alden, Bowman. Drafting the article: Ogiwara, Lyszczarz. Critically revising the article: Tomita, Alden, Bowman, McLone. Reviewed final version of the manuscript and approved it for submission: all authors.

References


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