ELSEVIER

The Spine Journal 8 (2008) 630–638

THE SPINE JOURNAL

Outcome of untethering for symptomatic spina bifida occulta with lumbosacral spinal cord tethering in 31 patients: analysis of preoperative prognostic factors

Jen-Ho Tseng, MD[a], Meng-Fai Kuo, MD, PhD[a], Yong Kwang Tu, MD, PhD[a], Ming-Yuan Tseng, MD, MPhil, MSc[b,*]

[a]*Division of Neurosurgery, Department of Surgery, National Taiwan University Hospital, Taipei, Taiwan*
[b]*Academic Department of Neurosurgery, Addenbrooke's Hospital, University of Cambridge, Cambridge, United Kingdom*

Received 6 August 2005; accepted 17 November 2005

Abstract

BACKGROUND CONTEXT: The most important goal for treating symptomatic lumbosacral spinal cord tethering is early untethering.

PURPOSE: To investigate preoperative symptoms that may have affected the outcome.

STUDY DESIGN: Patients with or without improvement and with or without favorable outcome after untethering were compared retrospectively by chart and image review.

PATIENT SAMPLE: Thirty-one patients (age between 2 days to 25 years) with spina bifida occulta and symptomatic cord tethering were analyzed. Presenting symptoms (neurological deficits, urological dysfunction, and lower limb deformities) were assessed before and after untethering.

OUTCOME MEASURES: Favorable outcome was defined as complete relief of symptoms or mild symptoms whereby patients are able to look after their own personal care without assistance. Unfavorable outcome was defined as moderate or severe disability whereby patients are unable to attend to their own bodily needs without assistance, are bedridden, or require constant nursing attention.

METHODS: Differences in patient characteristics and presenting symptoms were compared between those with and without clinical improvement and favorable outcome. Multivariate logistic regression was used to identify prognostic factors affecting the outcome.

RESULTS: The average age at surgery was 7.2 years, with a male-to-female ratio of 1.2. The average follow-up time was 4 years. At least one of the following symptoms was present in all patients: neurological deficits (83.9%), urological dysfunction (77.4%), or limb deformities (38.7%). After untethering, all patients had either symptoms stabilized (14 patients, 45.2%) or improved (17 patients, 54.8%), and 14 patients (45.2%) achieved total resolving of symptoms. Logistic regression confirmed that younger age (≤2 years, odds ratio [OR] 22.0, p=.026), lipomas of filum terminale (OR 25.6, p=.042), and a poor anal tone (OR 10.4, p=.061) were positive prognostic factors for the improvement in symptoms. The functional outcome was determined by the age at surgery (OR 0.9 per year since 1 year old, p=.04) and the presence of limb deformities (OR 0.06, p=.017).

CONCLUSIONS: In conclusion, our study suggests that untethering should be performed immediately once the patient shows evidence of symptomatic lumbosacral cord tethering, irrespective of age. Untethering can interrupt progression of symptoms, but sphincter dysfunction and muscle weakness are more likely to improve or resolve. Benefits can be seen in all patients, but young children (before 2 years old) have a higher chance to gain favorable outcome. Retethering is a main concern during follow-up, particularly for the more complicated lipomyelomeningoceles. Investigations using electrophysiologic and urodynamic studies are helpful for early detection of subtle symptomatic cord tethering or retethering. © 2008 Elsevier Inc. All rights reserved.

Keywords: Lipoma; Lumbosacral; Spina bifida occulta; Tethered cord; Untethering

FDA device/drug status: not applicable.
Nothing of value received from a commercial entity related to this manuscript.

* Corresponding author. Academic Department of Neurosurgery, Box 167, Level 4, Block A, Addenbrooke's Hospital, University of Cambridge, Cambridge CB2 2QQ, United Kingdom. Tel.: +44 1223 410634; fax: +44 1223 216926.
E-mail address: myt22@cam.ac.uk (M.-Y. Tseng)

1529-9430/08/$ – see front matter © 2008 Elsevier Inc. All rights reserved.
doi:10.1016/j.spinee.2005.11.005

Introduction

Spina bifida occulta with intraspinal lipoma (lipomyelomeningocele, lipoma of filum terminale) may cause symptomatic spinal cord tethering, including neurological deficits, sphincter dysfunction, and limb deformities [1]. Functional losses secondary to the tethering or the compressive effects of the lipoma are often aggravated during growth spurt [1,2]. Since the introduction of magnetic resonance imaging (MRI), early diagnosis and untethering can be done to reduce the potential lifelong disabilities [1,2]. Other examinations, such as electrophysiologic and urodynamic studies, can also be used for preoperative and postoperative assessment [3,4]. For those without neurological deficits, indications for prophylactic untethering are controversial [4,5].

For symptomatic cord tethering, early untethering and reconstruction of the thecal sac has been advocated [1,2,6]. However, surgical outcome depends upon not only the anatomical complexity, but also the clinical presentation before untethering [1,2]. For lipomas of filum terminale, division of the tight filum terminale is adequate; for lipomyelomeningoceles, the untethering is more complicated and carries higher risks of local complications and postoperative deficits [7]. Furthermore, the likelihood of retethering increases with time [8–10]. By retrospectively reviewing records of symptomatic patients diagnosed with spina bifida occulta and lipomyelomeningocele, our objectives were to determine potential prognostic factors that may be used to predict the surgical outcome, particularly the reversibility of various deficits by untethering.

Methods

Patients

This study was conducted as a part of in-hospital clinical audits; therefore ethics approval was not required [11,12]. Reviews of medical records and images were approved by the associated units (Departments of Surgery, Medical Records, and Radiology). Records of 51 patients coded with the International Classification of Diseases, Ninth Revision 756.17 (spina bifida occulta) and lumbosacral cord tethering diagnosed during January 1, 1980 to July 31, 2005 were available for analysis. The diagnosis of cord tethering on MRI was made by a qualified neuroradiologist and defined as the position of the conus medullaris below L3 [1,13]. To assess the outcome of untethering for symptomatic patients, five patients who underwent prophylactic untethering, seven patients who had excisions of lumbosacral lipomas for cosmetic purposes, and seven patients who were diagnosed before the MRI era (1990) were excluded. Another patient aged 38 years was excluded as an age outlier. Data of the final 31 patients were eligible for analysis (Table 1). Variables potentially affecting the surgical outcome included patient gender, age at surgery, presence of lipomyelomeningoceles or lipomas of the filum terminale, presenting neurological deficits, urological dysfunction, limb deformities, and use of patches for thecal sac augmentation during surgery. Neurological deficits included anal sphincter dysfunction and weakness or atrophy of the lower limbs. Urological dysfunction included abnormal urodynamic studies, vesicoureteric reflux, urinary incontinence, and complications of chronic urinary retention (hydronephrosis or chronic renal insufficiency). Lower limb deformities included pes cavus, talipes equines, equinovarus or equinovalgus.

These presenting symptoms (neurological, urological, and orthopedic) were continually assessed during follow-up. Favorable outcome was defined as complete relief of symptoms or mild symptoms whereby patients are able to look after their own personal care without assistance. Unfavorable outcome was defined as moderate or severe disability such that the patients are unable to attend to their own bodily needs without assistance, are bedridden, or require constant nursing attention.

Operation procedures

For lipomyelomeningoceles, untethering procedures consist of dissecting and debulking the lipoma upon the lumbodorsal fascia, laminectomy on the lower lumbar spine according to the level involved, and opening the dura for exposing the intradural portion of the lipoma. The junction between the lipoma and the neural placode is dissected and divided for untethering. The lipoma is subtotally excised to allow the neural placode to move freely within the spinal canal, and any tethering arachnoidal adhesions are divided. After the subtotal resection of the lipoma, the pia is closed if possible and the filum terminale is divided to release any potential tethering. For lipomas of filum terminale, untethering is achieved simply by dividing the tight filum terminale. Successful intraoperative untethering is defined as the conus medullaris retracts rostrally for at least one level of lamina. When the cerebrospinal fluid (CSF) space seems inadequate, patches of dural substitute are used. After satisfactory hemostasis, the wound is closed in layers.

During follow-up, comparison is made between patients with and without improvement, with complete and partial symptoms resolved, and favorable and unfavorable outcome. Data are presented as mean values $\pm$ standard deviation (SD), and considered significant when the two-tailed p value falls below .05. Univariate analysis was performed using the chi-square test; the Fisher exact test was used when any number of the cell in the 2*2 table is <5. Multivariate logistic regression was used to identify significant factors affecting the outcome. The model is built by a retrograde subtraction method, ie, all significant variables found in the univariate analysis are entered and the insignificant one is excluded sequentially using the Wald test. The odds ratio (OR) is presented with 95% confidence interval (95% CI), and the adjusted R-squared in each model is expressed as R^2.

Table 1
Summary of clinical characteristics of 31 patients who underwent untethering for symptomatic lumbosacral cord tethering

No.	Sex	Age at surgery	Intraspinal lipomas	Neurological deficits	Urological dysfunction	Orthopedic deformities	Use of dural patches	Follow-up time	Follow-up status	Follow-up conditions
1	F	3 mo	Lipomyelomeningocele	Nil	Nil	Bilateral pes cavus	Yes	6 mo	Stationary	Same as preoperative
2	F	4 days	Lipomyelomeningocele	Poor anal tone	Nil	Nil	Yes	2 yr	Improved	No sphincter dysfunction
3	F	2 mo	Lipomyelomeningocele	Poor anal tone	VU reflux	Nil	No	10 yr	Improved	No sphincter dysfunction and VU reflux
4	M	4 mo	Lipomyelomeningocele	Nil	VU reflux	Nil	Yes	5 yr	Improved	No VU reflux
5	F	8 mo	Lipomyelomeningocele	Poor anal tone	VU reflux	Left talipes equinovalgus	Yes	5 yr	Improved	No sphincter dysfunction, improved dorsiflexion
6	M	19 yr	Lipomyelomeningocele	Left leg weakness and atrophy	VU reflux	Left pes cavus	Yes	9 yr	Stationary	Same as preoperative
7	M	22 yr	Lipomyelomeningocele	Left leg weakness and atrophy	Incontinence with a neurogenic bladder	Bilateral per cavus	No	15 yr	Stationary	Same as preoperative
8	M	19 yr	Lipomyelomeningocele	Spasticity if both legs	Incontinence with a neurogenic bladder	Nil	No	4 yr	Stationary	Same as preoperative
9	M	22 yr	Lipomyelomeningocele	Anal incontinence, saddle anesthesia	Neurogenic bladder, VU reflux	Nil	No	6 yr	Stationary	Same as preoperative
10	M	2 days	Lipomyelomeningocele	Poor anal tone, left leg weakness	Nil	Nil	No	13 yr	Improved	No neurological deficits and sphincter dysfunction
11	F	6 yr	Lipomyelomeningocele	Poor anal tone	VU reflux	Nil	Yes	5 yr	Improved	No sphincter dysfunction
12	M	4 yr	Lipomyelomeningocele	Poor anal tone	Urinary incontinence, flaccid bladder	Right talipes equinovarus	No	1 yr	Improved	No sphincter dysfunction, but persistent pes equinovarus
13	F	6 yr	Lipomyelomeningocele	Left leg weakness and atrophy	Urinary incontinence	Bilateral talipes equinovarus	No	8 yr	Stationary	Same as preoperative
14	M	6 yr	Lipomyelomeningocele	Both leg weakness and atrophy, poor anal tone	Urinary Incontinence	Right talipes equinovarus	No	7 yr	Stationary	Same as preoperative
15	M	5 yr	Lipomyelomeningocele	Nil	VU reflux	Nil	No	9 yr	Stationary	Same as preoperative
16	M	9 yr	Lipomyelomeningocele	Both leg spasticity, poor anal tone	Urinary incontinence, VU reflux with right hydronephrosis	Bilateral pes cavus	Yes	3 mo	Improved	No sphincter dysfunction, less foot deformity
17	M	25 yr	Lipomyelomeningocele	Nil	Urinary incontinence, VU reflux	Nil	No	7 yr	Improved	No sphincter dysfunction
18	F	5 yr	Lipomyelomeningocele	Left leg weakness, poor anal tone	Urinary incontinence, VU reflux	Nil	Yes	6 yr	Stationary	Same as preoperative
19	M	5 yr	Lipomyelomeningocele	Poor anal tone	Urinary incontinence, UV reflux	Nil	Yes	3 mo	Stationary	Same as preoperative
20	M	18 yr	Lipomyelomeningocele	Left leg weakness and paresthesia	Urinary incontinence, renal function impairment	Left pes cavus	No	1 yr	Stationary	Same as preoperative
21	M	15 yr	Lipomyelomeningocele	Left leg weakness and paresthesia	Urinary incontinence	Left pes cavus	No	1 yr	Stationary	Same as preoperative
22	M	1.5 yr	Lipoma of filum terminale	Left leg mycolonic jerks	Nil	Nil	No	3 mo	Improved	No neurological deficits
23	M	4 yr	Lipoma of filum terminale	Left leg spasticity	Nil	Nil	No	4 mo	Improved	No neurological deficits
24	F	21 yr	Lipoma of filum terminale	Left leg paresthesia	Nil	Nil	No	6 mo	Improved	No neurological deficits

25	F	3 yr	Lipoma of filum terminale	Both leg spasticity	Flaccid neurogenic bladder	Right pes equinovarus	No	6 mo	Stationary	Same as preoperative
26	F	4 mo	Lipoma of filum terminale, sacral subcutaneous sinus tract	Poor anal tone	Urinary incontinence	Nil	No	1 yr	Improved	No sphincter dysfunction
27	F	4 yr	Lipoma of filum terminale	Poor anal tone	Nil	Nil	No	1 yr	Improved	No sphincter dysfunction
28	F	9 mo	Lipoma of filum terminale, sacral subcutaneous sinus tract	Poor anal tone	VU reflux, right side hydronephrosis	Nil	No	1 yr	Improved	No sphincter dysfuncation
29	F	9 mo	Lipoma of filum terminale, syrinx, sacral subcutaneous sinus tract	Nil	Flaccid neurogenic bladder	Nil	No	1 yr	Improved	No sphincter dysfunction
30	F	10 mo	Lipomyelomeningocele	Poor anal tone	Urinary incontinence, flaccid neurogenic bladder	Nil	No	1 yr	Improved	No sphincter dysfunction
31	M	9 days	Lipomyelomeningocele	Weakness of dorsiflexion of left foot	VU reflux	Left talipes equines	No	1 yr	Improved	No sphincter dysfunction, but persistent left talipes equinus and weak dorsiflexion

Abbreviations: mo=months; yr=year; VU=vesicoureteric.

Case 1

Patient 29 is a female infant born full-term via vaginal delivery. Her birth weight was 2.6 kg. Immediately a hemispheric, smooth-surfaced, soft, compressible, and well-defined edged lump was identified at the sacral region. During the 10-month follow-up period, no neurological deficits or limb deformities were found; however, the lump was discovered to be enlarging gradually. When she was 9 months old, she began to have frequent but small amounts of urine and stool voiding. Under the impression of symptomatic cord tethering, she was admitted for further assessment.

On examination, this 10-month-old female was comfortable in bed and moved all limbs actively and symmetrically. No limb deformities were demonstrable. A soft lump approximately 5 cm in diameter with the same characteristics described above was observed at the sacral region. A rectal examination revealed poor anal tone. A further urodynamic study, intravenous pyelography, and a voiding cystourethrography showed a neuropathic bladder of the flaccid type with a sustained high pressure in detrusor muscles. Abdominal ultrasonography did not disclose hydronephrosis or dilated ureters. MRI of the lumbosacral spine revealed the terminal placode attached to a fatty lump extending through the spina bifida at the S3–S4 level to the subcutaneous layer. A terminal syrinx was observed at the L4–S2 level (Fig. 1).

After the diagnosis of symptomatic tethered cord, procedures of untethering were performed as described previously, but dural patches were not used. The spinal cord was moving freely within the CSF space and retracted for at least one lamina high. The wound was closed uneventfully. Four days after the operation, a bulging mass with fluid thrills was found on the operation site of the sacral area. Reopening the wound revealed subcutaneous collection of CSF and a small defect on the dura, which was repaired by a primary closure. The following clinical course was smooth, and she was discharged 1 week after the second operation. During the 1-year follow-up, her activities were normal. Repeated urological examinations indicated that the abnormal urodynamics had completely resolved.

Case 2

Patient 31 is a male infant of heterozygotic twins born by a mother who conceived by in vitro fertilization. The prenatal clinic examination did not reveal fetal deformities. After 37 weeks of gestation, he was delivered through vagina with a birth weight of 2.4 kg. Immediately a hemispheric, smooth-surfaced, soft, compressible lump with well-defined edges at the sacral region was found. On examination, this newborn infant lay comfortable in bed. His left ankle showed talipes equines. A further urodynamic study demonstrated a high pressure in the detrusor muscles and dyssynergia of the detrusor and urinary sphincter. A lumbosacral MRI revealed a spina bifida at the L4–S1 level and a lipoma extending from the subcutaneous tissue to the neural placode. No associated syrinx was seen (Fig. 2). Under the diagnosis of

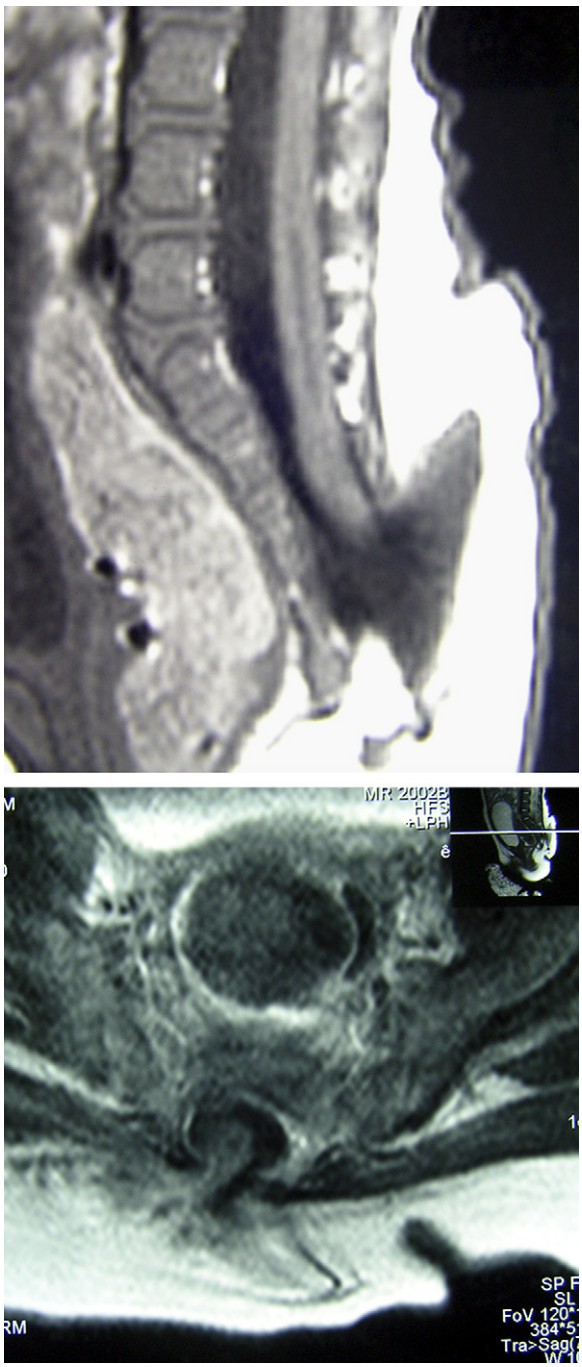

Fig. 1. T1-weighted lumbosacral spine magnetic resonance imaging of Patient 29 (Table 1) before the untethering (top: a longitudinal view; bottom: an axial view). There is a lipoma (high signal intensity) extending from the subcutaneous layer through the spina bifida into the intraspinal cerebrospinal fluid space (low signal intensity) and attaching to the neural placode (intermediate intensity) at the S3–S4 level. A syrinx can be seen above the placode at L4–S2 level.

symptomatic cord tethering, this male underwent untethering surgery, as described previously, 9 days after birth. Postoperative recovery was uneventful, and he was discharged 2 weeks after the operation.

During the 3-year follow-up, repeated urodynamic studies indicated that the sphincter dysfunction had completely

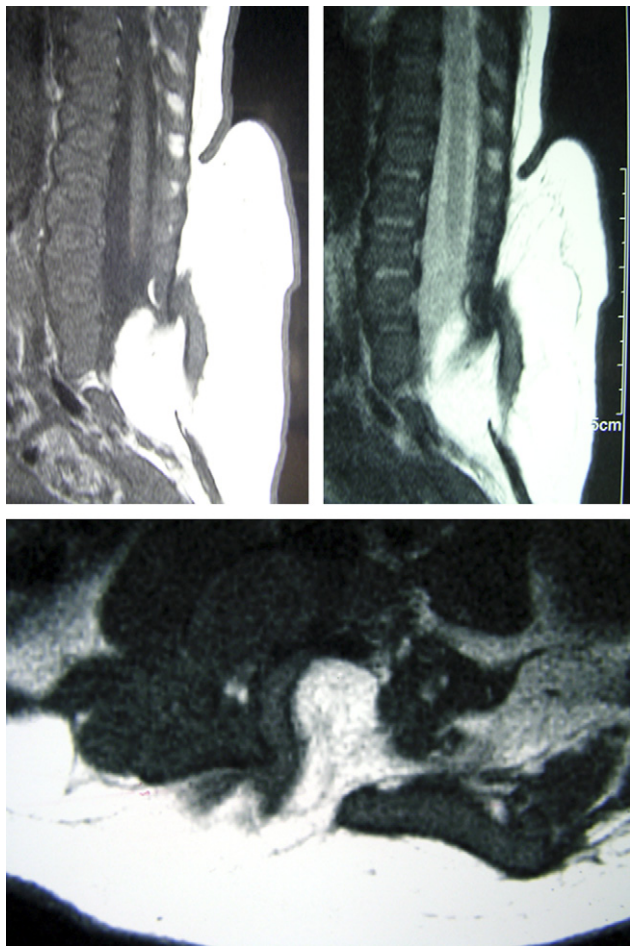

Fig. 2. Lumbosacral spine magnetic resonance imaging of Patient 31 before the first untethering (top left: T1-weighted longitudinal view; top right: T2-weighted longitudinal view; bottom: T2-weighted axial view). There is a lipoma (high signal intensity) extending from the subcutaneous lump through the spina bifida into the intraspinal space and attaching to the neural placode (intermediate intensity) at the L4–S1 level.

resolved, but he still had weak dorsiflexion of the left foot and could only walk with an ankle-foot orthosis. Repeated lumbosacral spine MRI revealed that the neural placode was at a low position and tethered with the residual lipoma to the dura at L4–S1 level. No associated syrinx was evident (Fig. 3).

A repeated untethering surgery was performed, in which adhesions among the placode, the residual lipoma, and the dura were released. However, the left side sacral nerve roots were found shorter than the right ones, which might have contributed to the cord tethering effects. Postoperative recovery was smooth, and a further 1-year follow-up showed that the neurological deficits remained unchanged.

Results

Clinical data

Table 1 summarizes clinical characteristics of the 31 patients. Tables 2 and 3 summarize univariate analysis

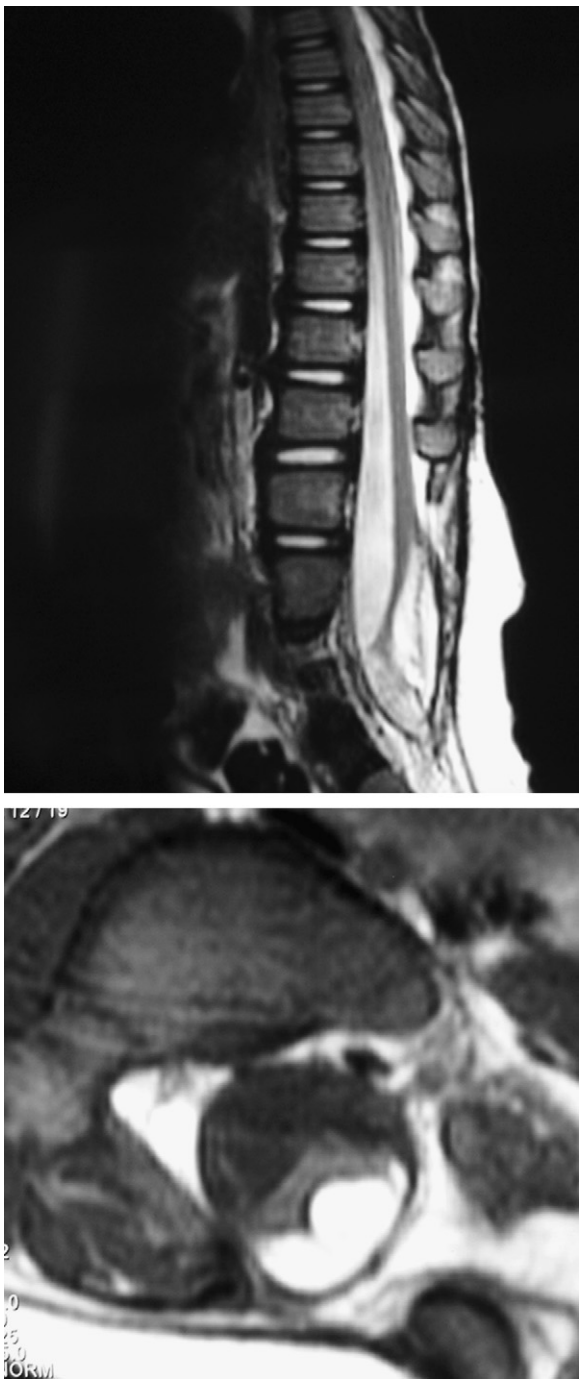

Fig. 3. Lumbosacral spine magnetic resonance imaging of Patient 31 before the second untethering (top left: T1-weighted longitudinal view; top right: T2-weighted longitudinal view; bottom: T2-weighted axial view). The lumbosacral thecal sac is enlarged, compared with that in Figure 2. However, an absence of cerebrospinal fluid space can be seen at the left posterolateral aspect of the neural placode (intermediate intensity) and the residual lipoma (high intensity) at the L4–S1 level.

according to follow-up status and outcome. There were 17 male and 14 female patients with a male-to-female ratio of 1.2. The average age at surgery was 7.2±8.1 years (age between 2 days to 25 years). Twelve patients (38.7%) were 2 years old or younger. All patients had cutaneous stigmata at the lumbosacral region, including subcutaneous lipomas (87%), skin dimples (10%), tufts of hair (5%), or skin tags (5%). Diagnoses of intraspinal lipomas included 23 (74.2%) lipomyelomeningoceles and 8 (25.8%) lipomas of filum terminale. One patient (3.2%) had an associated terminal syrinx. No other associated abnormalities such as diastematomyelia, Chiari malformation, or Rubinstein-Taybi syndrome were found among all these patients. Before the untethering, all patients manifested symptoms in at least one of the three categories: neurological deficits in 26 (83.9%), urological dysfunction in 24 (77.4%), and orthopedic deformities of the lower limbs in 12 (38.7%). The neurological deficits included a poor anal tone in 15 patients (48.4%) and leg weakness in 16 (51.6%). Urological dysfunction included urinary incontinence in 13 patients (41.9%) and vesicoureteric reflux in 13 (41.9%). Orthopedic deformities included pes cavus in 6 patients (19.4%), talipes equinovarus in 4 (12.9%), talipes equinovalgus in 1 (3.2%), and talipes equines in 1 (3.2%).

All of these patients achieved satisfactory untethering intraoperatively, and dural patches were used in 10 patients (32.2%). The mean follow-up period was 4.0±4.1 years (from 3 months to 15 years). During the follow-up, all patients had either symptoms unchanged (14 patients, 45.2%) or improved (17 patients, 54.8%). In 14 patients (45.2%) symptoms totally resolved. The main symptoms that remained after untethering were leg weakness (12 patients, 38.7%) and orthopedic deformities (12 patients, 38.7%). One patient (3.2%) had retethering on MRI and had a second untethering surgery 3 years after the first one, despite resolved sphincter dysfunction. It was found that the adhesions among the neural placode, the residual lipoma, and the dura caused the retethering. After the second untethering, the neurological deficits remained unchanged. Local complications such as minor CSF leaks occurred in one patient (3.2%), which was managed by primary closure of the dura.

Univariate analysis

For achieving symptoms improved after untethering, younger age (≤2 years, p=.001), lipoma of filum terminale (p=.031), and poor anal tone before the operation (p=.045) were positive factors, whereas leg weakness (p=.006) and orthopedic deformities (p=.008) before the operation were negative factors. Other negative factors of borderline significance included male gender (p=.092) and urological symptoms (p=.062).

For achieving symptoms totally resolved, again younger age (≤2 years, p=.001) and lipoma of filum terminale (p=.005) were positive factors, whereas leg weakness (p=.020), urological dysfunction (p=.014), particularly urinary incontinence (p=.036), and orthopedic deformities (p<.001) before the operation were negative factors. Male gender was a negative factor of borderline significance (p=.052).

Table 2
Summary of follow-up status of symptoms after untethering in 31 patients with symptomatic lumbosacral cord tethering

Patient characteristics	Total (n=31)	Improved (n=17)	Stationary (n=14)	p (χ^2)	Totally resolved (n=14)	Not totally resolved (n=17)	p (χ^2)
Male	17 (54.8%)	7	10	.092	5	12	.052
Female	14 (45.2%)	10	4		9	5	
Age at surgery							
≤2 years	12 (38.7%)	11	1	.001	10	2	.001
>2 years	19 (61.3%)	6	13		4	15	
Lipomyelomeningocele	23 (74.2%)	10	13	.031	7	16	.005
Lipoma of filum terminale	8 (25.8%)	7	1		7	1	
Symptoms at diagnosis							
Neurological deficits	26 (83.9%)	14	12	.800	11	15	.467
Poor anal tone	15 (48.4%)	11	4	.045	8	7	.376
Leg weakness	16 (51.6%)	5	11	.006	4	12	.020
Urological symptoms	24 (77.4%)	11	13	.062	8	16	.014
Urinary incontinence	13 (41.9%)	5	8	.119	3	10	.036
Vesicoureteric reflux	13 (41.9%)	7	6	.925	5	8	.524
Orthopedic deformities	12 (38.7%)	2	9	.008	0	12	<.001
Thecal sac augmentation using dural patches	10 (32.3%)	5	5	.709	3	7	.242

However, for achieving favorable outcome, younger age (age ≤2 years, p=.005) and urinary symptoms (p=.054), particularly urinary incontinence (p=.010) before operation, were positive prognostic factors, whereas leg weakness (p=.008) and orthopedic deformities (p=.004) before the operation were negative factors.

Logistic regression confirmed that younger age (≤2 years, OR 22.0, 95% CI 1.5 to 330.3, p=.026), lipomas of filum terminale (OR 25.6, 95% CI 1.1 to 583.1, p=.042), and poor anal tone (OR 10.4, 95% CI 0.9 to 119.5, p=.061) were positive prognostic factors for the improvement in symptoms (R^2=0.45). For complete resolution of symptoms, younger age (≤2 years, OR 20.0, 95% CI 2.4 to 169.0, p=.006) and lipomas of filum terminale (OR 17.5, 95% CI 1.2 to 244.6, p=.034) were the two decisive factors (R^2=0.43). However, the final functional outcome was determined by the age at surgery (OR 0.9 per year since 1 year old, 95% CI 0.8 to 1.0, p=.04) and the presence of limb deformities (OR 0.06, 95% CI 0.01 to 0.6, p=.017, R^2=0.36). No effects on clinical outcome from the use of dural patches were evident.

Discussion

Spina bifida occulta combined with lipomyelomeningoceles or lipomas of filum terminale may produce cord ischemia by traction, compression, or direct transmission of external force, which frequently become symptomatic during growth spurt [1,2]. Early untethering has been confirmed effective for preventing further deterioration [6]. Age is obviously the most important prognostic factor, because residual neurological functions are more likely

Table 3
Summary of outcome status after untethering in 31 patients with symptomatic lumbosacral cord tethering

Patient characteristics	Total (n=31)	Favorable outcome (n=22)	Unfavorable outcome (n=9)	p (χ^2)
Male	17 (54.8%)	10	7	.101
Female	14 (45.2%)	12	2	
Age at surgery				
≤ 2 years	12 (38.7%)	12	0	.005
> 2 years	19 (61.3%)	10	9	
Lipomyelomeningocele	23 (74.2%)	15	8	.232
Lipoma of filum terminale	8 (25.8%)	7	1	
Symptoms at diagnosis				
Neurological deficits	26 (83.9%)	17	9	.118
Poor anal tone	15 (48.4%)	13	2	.062
Leg weakness	16 (51.6%)	8	8	.008
Urological symptoms	24 (77.4%)	15	9	.054
Urinary incontinence	13 (41.9%)	6	7	.010
Vesicoureteric reflux	13 (41.9%)	10	3	.535
Orthopedic deformities	12 (38.7%)	5	7	.004
Dural augmentation using dural patches	10 (32.3%)	8	2	.445

to be preserved in younger children [10]. The purpose of untethering is to restore mobility of the distal spinal cord in order to ameliorate cord ischemia by a subtotal resection of the lipoma, a complete resection of fibrolipomatous stalk joining the intraspinal and extraspinal portions of the lipoma, or division of the tight filum terminale [6].

For lipomas of filum terminale, the untethering can be done simply by dividing the tight filum terminale [2,14], and the results are excellent [7]; in contrast, for lipomyelomeningoceles, the untethering procedures are much more complicated and the effects diminish with the time [7]. Up to 20% of patients who undergo untethering procedures for lipomyelomeningoceles experience local complications (CSF leaks, wound infection), and in one report, 3.9% of these patients manifested neurological deterioration [7]. Minor CSF leakage is the most common local complication; this is because a generously reconstructed thecal sac allows CSF to access the dural closure [6]. These local complications are not difficult to manage, and none of them are associated with retethering [5]. The postoperative neurological deterioration is transient for most patients, though it is associated with the complexity of lipomyelomeningoceles, adhesions, and nerve root deformities [7]. Nevertheless, for symptomatic patients, effective untethering has been shown to reverse sphincter dysfunction [1,4,6,7,10,14] and improve muscle strength [6,7]. Although limb deformities usually remain unchanged after untethering [7,9], associated deformities, such as degeneration of the hip or knee joint, and compensatory scoliosis, have been shown to ameliorate gradually [2,6]. These limb deformities may be associated with abnormal electrophysiology within the spinal cord [15] or abnormal innervation of the lower limb [1,15], and therefore, may be unlikely to be responsive to untethering [1].

In addition to the functional outcome, retethering is one of the main issues that needs to be addressed after untethering [5,8–10,16]. The incidence may be as high as 7% and is usually associated with scarring related to previous procedures [5]. For lipomas of filum terminale, retethering from the sectioned stump is rare [16]; for lipomyelomeningocele, the accumulated incidence of retethering increases over time [3]. To reduce the incidence of retethering, thecal sac augmentation with dural substitutes has been recommended [1,10,17]. However, the effect on reducing retethering is unknown [5,8]. Furthermore, the retethering has been considered to be associated with other anomalies, such as myelodysplasia [7] or tethering by short sacral nerve roots [18].

The timing and indications of untethering for asymptomatic patients are still controversial, particularly for lipomyelomeningoceles, because the precise natural history of them is still unclear [3,7]. Several studies suggest that the untethering should be delayed until symptoms occur, because a low-localized conus on images does not necessarily imply functional impairment [19]. Furthermore, the incidence and pattern of neurological deterioration after prophylactic surgery for lipomyelomeningoceles seems similar to those without operations [3]. Therefore, it has been suggested that for asymptomatic patients, an expectant approach may be adopted [3,20] or surgery may be deferred until 1 year to allow technically easier removal [1]. Nevertheless, patients without apparent neurological deficits may have abnormal electrophysiologic or urodynamic studies [1]. In addition, in infants, urodynamic studies are more difficult to interpret and deterioration can be missed [3]. Moreover, several particular activities may predispose the onset of symptoms in patients whose cord motion is marginally compromised [21,22]. Therefore, prophylactic untethering has been recommended for lipomas of filum terminale where nearly no surgical complications can be seen [2]. The seven patients who had prophylactic untethering within the same cohort of this study all had a favorable outcome and remain symptom-free.

In conclusion, this study suggests that untethering should be performed immediately once the patient manifests evidence of symptomatic lumbosacral cord tethering, irrespective of age. Untethering can interrupt progression of symptoms, but sphincter dysfunction and muscle weakness are more likely to improve or resolve. Benefits can be seen in all patients, but younger children (before 2 years old) have a greater likelihood of attaining a favorable outcome. Retethering is a main concern during follow-up, particularly for the more complicated lipomyelomeningoceles. Investigations using electrophysiologic and urodynamic studies are helpful for early detection of subtle symptomatic cord tethering or retethering.

References

[1] Harrison MJ, Mitnick RJ, Rosenblum BR, Rothman AS. Leptomyelolipoma: analysis of 20 cases. J Neurosurg 1990;73:360–7.
[2] La Marca F, Grant JA, Tomita T, McLone DG. Spinal lipomas in children: outcome of 270 procedures. Pediatr Neurosurg 1997;26:8–16.
[3] Kulkarni AV, Pierre-Kahn A, Zerah M. Conservative management of asymptomatic spinal lipomas of the conus. Neurosurgery 2004;54:868–73.
[4] Balkan E, Kilic N, Avsar I, Boyaci S, Aksoy K, Dogruyol H. Urodynamic findings in the tethered spinal cord: the effect of tethered cord division on lower urinary tract functions. Eur J Pediatr Surg 2001;11:116–9.
[5] Byrne RW, Hayes EA, George TM, McLone DG. Operative resection of 100 spinal lipomas in infants less than 1 year of age. Pediatr Neurosurg 1995;23:182–7.
[6] Herman JM, McLone DG, Storrs BB, Dauser RC. Analysis of 153 patients with myelomeningocele or spinal lipoma reoperated upon for a tethered cord. Pediatr Neurosurg 1993;9:243–9.
[7] Pierre-Kahn A, Zerah M, Renier D, et al. Congenital lumbosacral lipomas. Childs Nerv Syst 1997;13:298–335.
[8] Colak A, Pollack IF, Albright AL. Recurrent tethering: a common long-term problem after lipomyelomeningocele repair. Pediatr Neurosurg 1998;29:184–90.
[9] Schoenmakers MA, Gooskens RH, Gulmans VA, et al. Long-term outcome of neurosurgical untethering on neurosegmental motor and ambulation levels. Dev Med Child Neurol 2003;45:551–5.
[10] Kang JK, Lee KS, Jeun SS, Lee IW, Kim MC. Role of surgery for maintaining urological function and prevention of retethering in the treatment of lipomeningomyelocele: experience recorded in 75 lipomeningomyelocele patients. Childs Nerv Syst 2003;19:23–9.

[11] Royal College of Physicians. Independent ethical review of studies involving personal medical records. Report of a working group to the Royal College of Physicians Committee on Ethical Issues in Medicine. J R Coll Physicians Lond 1994;28:439–43.
[12] Brewster DH, Bain MR, Chalmers JW, Gould A, Dewar JA, George WD. Informed consent: explicit consent is not needed for studies using medical records. BMJ 1998;317:948.
[13] Barson AJ. The vertebral level of termination of the spinal cord during normal and abnormal development. J Anat 1970;106:489–97.
[14] Wehby MC, O'Hollaren PS, Abtin K, Hume JL, Richards BJ. Occult tight filum terminale syndrome: results of surgical untethering. Pediatr Neurosurg 2004;40:51–7.
[15] Feldbrin Z, Gilai AN, Ezra E, Khermosh O, Kramer U, Wientroub S. Muscle imbalance in the aetiology of idiopathic club foot: an electromyographic study. J Bone Joint Surg Br 1995;77:596–601.
[16] Souweidane MM, Drake JM. Retethering of sectioned fibrolipomatous filum terminales: report of two cases. Neurosurgery 1998;42:1390–3.
[17] Aliredjo RP, de Vries J, Menovsky T, Grotenhuis JA, Merx J. The use of Gore-Tex membrane for adhesion prevention in tethered spinal cord surgery: technical case reports. Neurosurgery 1999;44:674–7.
[18] Barolat G, Schaefer D, Zeme S. Recurrent spinal cord tethering by sacral nerve root following lipomyelomeningocele surgery. J Neurosurg 1991;75:143–5.
[19] Oi S, Yamada H, Matsumoto S. Tethered cord syndrome versus low-placed conus medullaris in an over-distended spinal cord following initial repair for myelodysplasia. Childs Nerv Syst 1990;6: 264–9.
[20] Tuuha SE, Aziz D, Drake J, Wales P, Kim PC. Is surgery necessary for asymptomatic tethered cord in anorectal malformation patients? J Pediatr Surg 2004;39:773–7.
[21] Iskandar BJ, Fulmer BB, Hadley MN, Oakes WJ. Congenital tethered spinal cord syndrome in adults. J Neurosurg 1998;88:958–61.
[22] Pang D, Wilberger JE. Tethered cord syndrome in adults. J Neurosurg 1982;57:32–47.