Child's Nervous System (2022) 38:387–395
https://doi.org/10.1007/s00381-021-05353-y

ORIGINAL ARTICLE

Occult tethered cord syndrome: a rare, treatable condition

Jeyul Yang[1] · Jae-Kyung Won[2] · Kyung Hyun Kim[3] · Ji Yeoun Lee[3,4] · Seung-Ki Kim[3] · Hyung-Ik Shin[5] · Kwanjin Park[6] · Kyu-Chang Wang[1]

Received: 20 August 2021 / Accepted: 30 August 2021 / Published online: 5 October 2021

Abstract

Purpose Occult tethered cord syndrome (OTCS) is an entity that shows tethered cord syndrome (TCS) with normal spinal MRI findings. The definition and treatment of OTCS have been controversial since first proposal. The purpose of this study was to evaluate the existence, prevalence, histological characteristics, and surgical outcomes of OTCS.

Methods We retrospectively analyzed patients who underwent untethering surgery for OTCS from January 2010 to December 2019. Inclusion criteria were (1) clinical manifestation of TCS; (2) supported by urodynamic study (UDS) or electromyography/nerve conduction study; (3) no structural lesions in the urological tract or spinal cord, and no developmental delay; and (4) postoperative follow-up for > 6 months. Sectioned fila from OTCS patients were histologically compared with those from cases of thickened filum or low-lying conus.

Results Five (four female, one male) of 439 patients (1.1%) who underwent untethering surgeries for occult spinal dysraphism corresponded to OTCS. Mean age at the time of surgery was 16 years (7–22 years). Mean postoperative follow-up duration was 45 months (15–114 months). The main symptom was urinary dysfunction in four patients and leg pain in one. All patients had detrusor-sphincter dyssynergia. Fila from OTCS patients revealed increased fibrous tissue as in TCS patients. Four patients showed postoperative improvement and one with preoperative static course had no improvement.

Conclusions This study suggests that OTCS is a definitely existing entity although rare. OTCS is curable when timely treatment is given. Sudden onset with rapid progression of symptom seems the best indication for surgery.

Keywords Tethered cord · Untethering · Detrusor-sphincter dyssynergia · Filum terminale

✉ Kwanjin Park
urodori@naver.com

✉ Kyu-Chang Wang
kcwang@snu.ac.kr

1 Center for Rare Cancers, Neuro-Oncology Clinic, National Cancer Center, 323 Ilsan-ro, Ilsandong-gu, Goyang, Gyeonggi-do 10408, Korea

2 Department of Pathology, Seoul National University Hospital and Seoul National University College of Medicine, Seoul, Korea

3 Division of Pediatric Neurosurgery, Seoul National University Children's Hospital, Seoul, Korea

4 Department of Anatomy, Seoul National University College of Medicine, Seoul, Korea

5 Department of Rehabilitation Medicine, Seoul National University Hospital and Seoul National University College of Medicine, Seoul, Korea

6 Division of Pediatric Urology, Seoul National University Children's Hospital, 101 Daehak-no, Jongno-gu, Seoul 03080, Korea

Introduction

Occult tethered cord syndrome (OTCS) is described as an entity involving urinary dysfunction in patients with a normal spinal MRI without findings such as cord elongation or filum thickening [15, 27]. However, OTCS may also include non-urological symptoms such as ankle weakness and pain. OTCS is presumed to be caused by abnormal fixation of the conus medullaris from upward movement and subsequent ischemia of neural tissue [27]. Urinary dysfunction is assumed to result from the vulnerability of the conus to traction-induced dysfunction [27]. OTCS is more of a functional abnormality than an anatomical abnormality, and the lack of radiological evidence despite the presence of symptoms makes the treatment decision challenging.

Decisions made on the basis of evident causal relationships are important in the field of medicine. However, in OTCS, the lack of clues on neuroimaging makes surgeons hesitant to perform filum terminale sectioning. For instance,

the opinions of neurosurgeons on the role of surgery in patients with urinary dysfunction and normal MRI findings were divided in a questionnaire study [17]. This debate is still ongoing because previous studies have reported controversial results regarding the surgical outcomes of OTCS.

Several papers have reported positive aspects of surgery [15, 25]. A review of five retrospective, observational, non-controlled studies supported surgical treatment [14]. By contrast, a randomized controlled study reported that there was no difference in outcomes between surgery and medical treatment, and the existence of OTCS itself was questioned [19]. In addition, the recently published guideline from the International Children's Continence Society also made skeptical claims about surgical treatment [23]. The reason for this debate is thought to originate from differences in the definition of OTCS and the indications for and timing of surgery.

Therefore, to address this controversy, we applied strict criteria on our cohort and saw how many patients met the criteria for OTCS. Their clinical and urodynamic presentations are detailed to stress diagnostic ambiguity. Histological comparison of sectioned fila with the positive control revealed the pathological relevance of OTCS to "true" TCS. Patient outcomes suggested that a group of patients had good responses to surgery.

Methods

We retrospectively analyzed patients who were followed up for more than 6 months after untethering surgery for OTCS at Seoul National University Children's Hospital from January 2010 to December 2019. Medical records, radiological data, urodynamic study (UDS) results, and electromyography (EMG) and nerve conduction study (NCS) data were reviewed. OTCS was defined when (1) clinical manifestations of tethered cord syndrome, such as voiding, defecation, motor or sensory dysfunction or pain, which is not controlled with conservative measures; (2) symptoms supported by UDS or EMG/NCS data; (3) a conus medullaris located in the normal position (between T12-L1 junction and L2-3 junction); and (4) a normal-thickness filum terminale (< 2 mm) were observed [4–6, 28]. The conus medullaris location was chosen because the L3 conus level has an intermediate tethered cord risk [26]. Filum terminale thickness was measured at the thickest location in T2-weighted axial images taken with 1.5-Tesla MRI, and lipoma presence within the filum terminale was not considered among the diagnostic criteria [4–6, 28]. Patients with structural lesions in the urological tract, developmental delay, syringomyelia, or other apparent anatomical tethered cord findings were excluded. Patients with anorectal malformation were included only when their urological presentations were found to be neurogenic after sufficient medical management by the experts concerned.

Our team is rather conservative on indications for surgery in occult spinal dysraphism. Urological manifestations of OTCS were not determined by a single history or abnormal test result. Rather, repeated studies and urodynamic confirmation in the face of repetitive abnormalities were required for OTCS diagnosis. When young children show persistent incontinence or increased postvoid residual urine (PVR), follow-up with repeated examinations is recommended rather than early surgery because many times the symptoms are transient, especially when toilet training is still incomplete. If OTCS is suspected in a patient who has chronic static problems without progression, we do not recommend surgery. Surgery is offered only if the patient has a new onset or progression of symptoms or signs. However, if the patient shows evident detrusor-sphincter dyssynergia in a UDS at follow-up, we perform surgery even if the clinical course is static (patient 5). Hyperreflexia in UDSs in patients with urinary incontinence and normal MRI findings reportedly showed good responses to untethering [12, 13].

Medical records were reviewed for demographic data, main symptoms, main symptom duration, preceding symptoms, onset and progression pattern, concurrent neurological status, coexisting constipation, and associated anomalies. Preceding symptoms were defined as those that may have been caused by a tethered cord but of a different type from the main symptoms. Progression pattern was categorized as rapid, slow, or static. Rapid progression was defined as an aggravation of main symptoms at every visit or UDS. Slow progression was defined as a preoperative aggravation of main symptoms, but not at every outpatient visit or UDS. Static course was defined as no change in severity throughout the preoperative period. The patients were followed up at 3- to 6-month intervals depending on symptom severity.

All patients underwent preoperative UDSs, which were performed and the results of which were interpreted by an expert pediatric urologist (KP). PVR was recorded based on the amount measured by sonography during an outpatient visit or UDS. EMG with an NCS was conducted by pediatric rehabilitation doctors (HIS et al.) when the manual muscle power test was not feasible or its results suggested progression. EMG with an NCS was also performed to rule out neurological disorders other than tethered cord. All patients received an MRI examination before untethering surgery. The decision to perform untethering surgery was made after multidisciplinary evaluation by pediatric neurosurgeons, urologists, rehabilitation medicine doctors, and neurologists. Untethering surgery was performed after one-level laminotomy or hemilaminectomy followed by filum terminale sectioning.

Histological findings of the filum terminale of OTCS were reviewed by an expert neuropathologist (JKW). A comparison between sectioned tissues of the filum terminale of OTCS patients and tissue samples from five definite TCS patients (positive control group) was performed. Definite TCS was defined as clinical manifestations of a tethered cord with abnormal MRI findings such as a thickened filum terminale or low-lying conus associated with UDS or EMG abnormalities. The comparison with definite TCS patients was performed because obtaining normal filum terminales was not feasible. The control group was preferably matched by sex and age to the OTCS group. However, due to the different clinical courses, there was an age difference between the groups. Tissues were stained, and a quantitative comparison based on microscopic analysis of ependymal, adipose, fibrous, and elastin tissues and nerve twigs was made.

Postoperative outcome was evaluated in terms of the main symptom. PVR as a surrogate marker for bladder-emptying function was measured by sonography at the outpatient clinic or during a UDS. The timing of the postoperative UDS depended on the patient's outcome and condition.

For statistical analysis, Student's paired *t*-test was performed to compare the histological findings of the OTCS patient group and the control group. SPSS Statistics version 18 (SPSS Inc., Chicago, IL, USA) was used. All analyses were two-sided, and a p value < 0.05 was considered significant. The present study was approved by the institutional review board of the Seoul National University Hospital (IRB no. 2004–104-1118). Informed consent was waived by the IRB since this study was retrospective.

Results

In total, 439 untethering surgeries were performed for occult spinal dysraphism during the study period. Among them, five patients (1.1%; 4 females and 1 male; mean age at the time of surgery = 16 years) had OTCS. The youngest patient was 7 years old, and all patients were beyond toilet training age. The most common main symptom was difficulty in urination, presenting as straining, urinary dribbling, or urinary incontinence (Table 1). In one patient (patient 1), the main symptom was acute posterior lower-extremity pain. The patient underwent surgery for imperforate anus and had urinary incontinence as a preceding symptom.

The duration of main symptoms varied rather widely, from 3 months to a lifetime. Regarding onset and progression patterns, onset was sudden in three patients (cases 1–3) and insidious in two (cases 4 and 5). Two patients (cases 1 and 2) showed rapid progression, two patients (cases 3 and 4) showed slow progression, and the other patient (case 5) showed no progression. Overall, two patients had sudden onset and rapid progression (cases 1 and 2), one patient had sudden onset and slow progression (case 3), one had insidious onset and slow progression (case 4), and one had insidious onset and static course (case 5). The reason for untethering surgery in one patient with static course (case 5) was because the patient complained of no improvement in the main symptoms with conservative management, evident neurogenic bladder detrusor-sphincter dyssynergia (DSD) was found in a UDS, and surgery risk was low.

Representative urodynamic traces and concordant voiding cystographic findings for all OTCS patients are

Table 1 Clinical manifestation of the five patients with occult tethered cord syndrome

Case No.	Sex/age (years)	Main symptom	Duration of the main symptom (months)	Other preceding symptom (duration)	Onset, progression pattern	Motor/ sensory deficit	Constipation	Associated anomaly
1	F/13	Pain	14	Urinary incontinence (1 year)	Sudden, rapid	−/−	+	Imperforate anus
2	M/22	Voiding difficulty[a], increased PVR	3	None	Sudden, rapid	−/+	+	Dimple
3	F/18	Voiding difficulty	7	None	Sudden, slow	+/+	+	Scoliosis
4	F/7	Voiding difficulty	Since birth	None	Insidious, slow	−/−	−	None
5	F/21	Urinary incontinence	Since birth	None	Insidious, static	−/−	+	None

PVR post-voiding residual

[a]Includes difficulty in initiation

shown in Fig. 1. Cases 3 and 4 showed elevated pressure, indicating low compliance. Cases 1 and 4 demonstrated significant detrusor overactivity during the filling phase. Cases 2–5 revealed typical features of DSD. These are the signs of upper motor neuron damage caused by tethered cord syndrome. Case 1 mainly presented with pain, and her urological problems could be attributed to fecal impaction inherent to her underlying imperforate anus. Her urodynamic trace did not indicate neurogenic bladder despite the presence of detrusor overactivity. Her detrusor overactivity was mild. Her fecal impaction was implicated in dysfunctional voiding, and this made lack of opening at different levels from other cases during the voiding trial.

Patients' preoperative findings are shown in Table 2. All patients had DSD with increased PVR. One patient was already on clean intermittent catheterization (CIC) at admission. Four patients had undergone preoperative EMG/NCS, the results of which were normal in three patients, whereas one patient showed mild dysfunction of the dorsal column-medial lemniscus pathway.

In all patients, the conus medullaris was located at the level of the L1-2 junction, with a mean filum terminale thickness of 1.1 mm (range, 0.7–1.5 mm). Surgical findings revealed one patient with a tight-looking filum terminale, whereas no abnormal findings were found in other patients.

Fila from a total of 10 patients, five patients each from the OTCS and TCS groups, were histologically analyzed

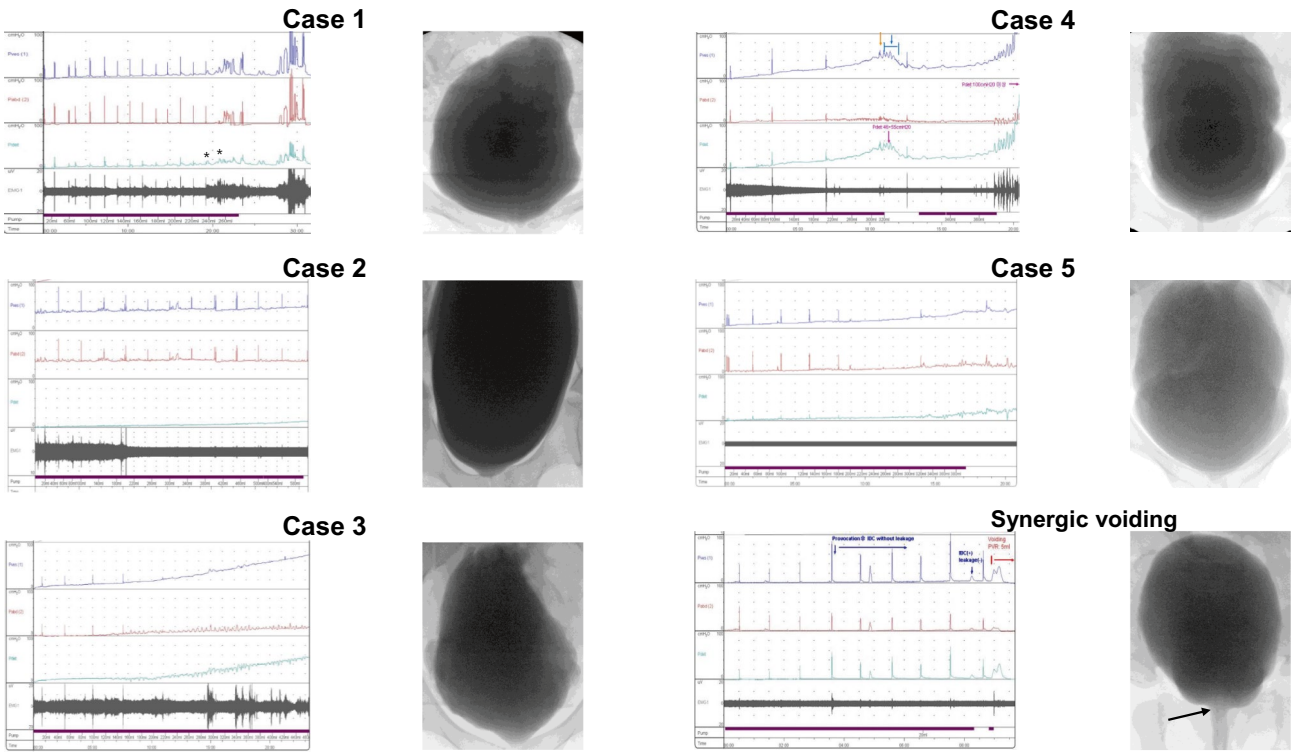

Fig. 1 Representative urodynamic trace and concordant cystography during voiding phase for all cases of OTCS. For each case, preoperative urodynamic trace was seen in left side panel. Vesical, abdominal and detrusor pressor was depicted in 1st , 2nd and 3rd row. During filling phase, bladder should be compliant and detrusor pressure should not significantly increase, but cases 3 and 4 showed elevated pressure indicating low compliance. Case 1, case 4 and even the case for synergic voiding revealed significant detrusor activities during filling phase (asterisk). Case 2 and case 5 were normal during filling phase, however when they tried to void their external sphincter would not open revealing dilated posterior urethra which was characteristic of typical detrusor sphincter dyssynergia (DSD). So they fail to void and were called acontractile neurogenic bladder. Interestingly, similar sphincteric shape was found in case 3 and 4 when detrusor pressure was elevated. This is the sign of upper motor neuron damage caused by tethered cord syndrome. The finding was contrasted to reference case of synergic voiding showing maintenence of low detrusor pressure during filling, opening of sphincter and subsequent urination exerted by elevated detrusor pressure (arrow). This suggests normal reflex arc. Case 1 mainly presented neuroorthopedic symptoms and her urologic problems could be attributed to fecal impaction inherent to her underlying condition of imperforate anus. Thus, her urodynamic trace did not reveal any evidence for neurogenic bladder despite the presence of small detrusor overactivity. Her fecal impaction may be implicated in dysfunctional voiding and this showed incomplete closing action of sphincter during voiding trial. This contrasted to the other cases revealing complete closing action of sphincter

Table 2 Preoperative findings of the five patients with occult tethered cord syndrome

Case No.	PVR (cc)	UDS findings	EMG findings	Level of terminal conus medullaris	Thickness of filum terminale (mm)	Surgical findings
1	360	Normal compliance, normal detrusor contraction, detrusor internal sphincter dyssynergia	Normal	L1/2	0.7	Tight filum terminale
2	800	Underactive detrusor, DSD	Normal	L1/2	1.5	Normal
3	400	Decreased compliance and capacity, DO (−), DSD	Normal	L1/2	1.5	Normal
4	130	Decreased compliance and capacity, DO, DSD	Not done	L1/2	1	Normal
5	CIC	Decreased compliance, acontractile detrusor, competent sphincter, DSD	Mild dysfunction of dorsal column-medial lemniscus pathway	L1/2	0.9	Normal

CIC clean intermittent catheterization, *DO* detrusor overactivity, *DSD* detrusor-sphincter dyssynergia, *EMG* electromyography, *PVR* post-voiding residual, *UDS* urodynamic study

(Table 3). The fibrous tissue increase and elastin decrease in OTCS patients were slightly exacerbated or similar to those in TCS patients. The overall fibrous tissue portion tended to be higher in OTCS patients than in TCS patients ($p = 0.069$). The overall elastin portion showed no significant difference compared with TCS patients ($p = 0.148$). Nerve twigs were all present in OTCS patients.

The mean follow-up duration after untethering surgery was 45 months (range, 15–114 months). There were three patients who showed improvement after untethering surgery (Table 4). One patient had no improvement or aggravation, and one patient showed temporary improvement followed by re-aggravation. The latter patient (patient 3) underwent augmentation cystoplasty 2 years after untethering surgery. There were no acute complications after untethering surgery.

Illustrative cases

Case 2

A 22-year-old male was admitted with reduced bladder sensation and voiding difficulty that began 3 months prior. He had no medical history other than a shallow dimple on the tip of the coccyx, which did not require treatment. Physical examination revealed saddle hypoesthesia, but no motor weakness or foot deformity was noted. No abnormal findings were found on MRI, but DSD was shown in a UDS. The symptoms progressed rapidly, and PVR increased to 800 cc during a 3-month period. Medical treatment was given initially but had no effect. We therefore performed untethering surgery, which revealed a normal-appearing filum terminale without tension. Spontaneous urination was completely recovered, and PVR was normalized 3 years after surgery. Saddle hypoesthesia also disappeared.

Case 3

An 18-year-old female presented with sudden-onset voiding difficulty and enuresis accompanied by flank and back pain. Acute pyelonephritis was diagnosed by laboratory examinations. MRI, UDS, and EMG/NCS were performed to determine the cause. DSD was shown in the UDS, but MRI and EMG/NCS findings were normal. Untethering surgery was performed after excluding peripheral neuropathy, and the symptoms improved. PVR decreased from 400 to 150 cc, and bladder sensation recovered. CIC after self-voiding was recommended postoperatively. Twenty months after surgery, she started to be followed up at another hospital due to accessibility. Unfortunately, the patient's compliance was poor and she did not catheterize as recommended. Two years after the untethering surgery, the patient developed azotemia and metabolic acidosis. Kidney ultrasonography revealed an aggravation of diffuse calyceal dilatation and extreme cortical thinning, and obstructive uropathy was shown in a UDS. She underwent bilateral ureter dissection and bladder augmentation at another hospital and is currently on CIC without additional azotemia.

Discussion

The definition and treatment of OTCS have been controversial since first being proposed [24]. The concept of OTCS has even been challenged recently [19]. The diagnostic criteria, such as the conus medullaris level, presence of syringomyelia, or thickened filum terminale (> 2 mm in diameter), vary among studies [1, 18]. In our study, a low-lying conus medullaris (lower than the L2-3 junction), syringomyelia, and a thickened filum terminale were

Table 3 Comparison of histopathology between thickened filum terminale and occult tethered cord syndrome

Case No.	Age (years)	Sex	Ependymal tissue	Nerve	Adipose tissue	Vessel	Fibrous area*	Masson trichrome (collagen)†	Elastin (elastic fiber)	Reticulin†
Thickened filum terminale										
1	5	F	Y	Y	Y (5%)	Y	70%	75%	5% (thin, dispersed)	75%
2	7	M	N	Y	Y (50%)	Y	40%	60%	< 1% (thin, dispersed)	20%
3	8	F	Y	N	Y (50%)	Y	40%	50%	< 1% (thin, dispersed, short)	40%
4	9	F	N	Y	Y (70%)	Y	60%	80%	10% (not coiled, straight, thin)	80%
5	11	M	Y	N	Y (70%)	Y	30%	50%	0% (nearly absent)	80%
Occult tethered cord syndrome										
1	13	F	Y	Y	Y (10%)	Y	80%	70%	0% (nearly absent)	70%
2	22	M	Y	Y	N	Y	80%	60%	< 1% (thin, dispersed)	60%
3	18	F	Y	Y	N	Y	45%	50%	0% (nearly absent)	50%
4	7	F	Y	Y	N	Y	90%	70%	0% (nearly absent)	70%
5	21	F	Y	Y	N	Y	60%	90%	< 1% (thin, dispersed)	90%

Y yes, *N* no

*Percentage from total area

†Percentage from fibrous tissue

excluded to see whether patients with "strictly" normal MRI could develop tethered cord syndrome and to clarify the indications for surgery.

Our study, which applied strict diagnostic criteria, showed that OTCS is definitely an existing entity, although the incidence is low. Patients beyond toilet training age with no abnormal MRI findings despite the presence of symptoms that were supported by UDS findings clearly existed in our cohort. However, these patients were rare, even taking into consideration that our institution is a tertiary hospital with a pediatric neurological center, and constituted 1.1% (0.9% if patient 5, who showed no change after surgery, was excluded) of our occult spinal dysraphism cases during the study period. This finding is in line with previous results in terms of OTCS rarity [22].

Common findings of the patients were urinary symptoms. In all but one patient (case 1), urinary problems attributable to OTCS were found and these problems showed a urodynamic pattern suggestive of neurogenic bladder, such as decreased compliance, a lack of bladder contractility, detrusor overactivity, and DSD. The latter two are suggestive of upper motor neuron signs when TCS is involved. Moreover, preoperative UDSs were performed at ages older than 7 years. Regarding the clinical manifestations and ages of the patients, the reliability of UDS is considered acceptable. Patient 1 presented with no urological problems but pain. Although the patient had urinary incontinence, it was not of neurogenic origin but was due to dysfunctional voiding caused by constipation, which was associated with imperforate anus. Her urinary incontinence did not respond to untethering, although the pain improved rapidly after surgery. As the patient grew, her urinary incontinence gradually improved.

In contrast to the similarity of the preoperative findings, surgical outcomes varied among patients. Two patients (patients 1 and 2) fully recovered their self-voiding function, whereas one patient (patient 3) showed temporary improvement and then deteriorated. Because the latter patient moved to another hospital, we could not confirm whether the decision for augmentation cystoplasty was made appropriately. The patient may have improved without cystoplasty if she had catheterized as recommended and was well managed by conservative treatment as in her initial clinical course immediately after untethering surgery. Our experience revealed that urological function improvement after untethering may be a slow process and take up to 3 to 4 years [8]. We question whether augmentation cystoplasty was a premature decision.

Urinary incontinence and increased PVR and DSD were improved but still present in one patient (case 4) who had a relatively short follow-up. Another patient (case 5) had no improvement. Common characteristics of patients who fully and permanently benefited from untethering surgery were the sudden onset and progression of the main symptoms.

Table 4 Postoperative outcomes of the five patients with occult tethered cord syndrome

Case No.	Improvement of the main symptom	CIC	Urological outcome (interval from operation in months)	Follow-up duration (months)
1	Improved pain	No	PO repeated UTI with increased PVR → gradually normalized PVR for PO 2 years	114
2	Normalized voiding and PVR	No	UDS (2): normal storage phase, underactive detrusor → normalized voiding and PVR → no additional UDS	41
3	Temporarily improved voiding difficulty, but bladder augmentation (PO 2 years)	Refused, then augmentation cystoplasty	UDS (6): improved → lost follow-up → azotemia, UDS (29, at other hospital) DO in storage phase, detrusor underactivity in voiding phase → augmentation cystoplasty	30
4	Improved urinary incontinence	Yes	Improved urinary incontinence UDS: normal storage, improved compliance and DO (15)	15
5	No change of urinary incontinence	Yes	UDS (6): no change of decreased compliance, acontractile detrusor and DSD → no change of urinary incontinence	25

DO detrusor overactivity, *DSD* detrusor-sphincter dyssynergia, *PO* postoperative (post-untethering), *PVR* post-voiding residual, *UDS* urodynamic study

Symptoms caused by OTCS seem to occur with age, although the age at the surgery did not affect the outcomes. However, the occurrence and progression pattern of tethering seemed important. The patients with improvement (including the patient with temporal improvement) experienced a sudden onset of urinary symptoms, and accordingly, they could pinpoint the date of symptom occurrence or progression. In addition, symptoms completely improved in patients who experienced rapid progression. By contrast, the patient with no clinical improvement had an insidious onset and static preoperative course. Another common finding of patients with no improvement was that their urinary symptoms were primary and static, defined as not having a previous dry period of more than 6 months since birth [2], as shown in case 5.

Primary urinary incontinence is reported to be caused by (1) neurological problems such as a tethered cord since birth, (2) an inability to reach urinary maturation due to developmental delay, or (3) the presence of constipation [9, 11]. For patients with developmental delay or constipation, it is difficult to define whether untethering contributed to the alleviation of the symptoms or whether the symptoms improved over a natural course as time passed. In our study, patients with developmental delay were excluded and constipation was actively managed before the decision to perform surgery. Most OTCS studies have included both primary and secondary urinary incontinence despite it being more logical to include only secondary urinary incontinence or to separate primary and secondary urinary incontinence to elucidate the outcomes of surgery in children. Hence, caution is needed when interpreting surgical outcomes in these studies.

Fibrous tissue was ubiquitously found in fila and involved in cord tethering in an analysis of definite TCS patients' fila [21]. In addition, OTCS filum terminale histology was reviewed by Hendson et al. [7] by comparing the fila of patients with those of pediatric cadavers. The authors reported that OTCS fila showed loose fibrous connective tissue and evenly dispersed elastic fibers. Forty-eight percent of patients showed decreased elastic fibers. Other studies have also reported increased fibrous tissue and reduced elastin fibers in the filum terminale of TCS patients [16, 20, 21, 27]. As shown by Yamada et al. [27] through a stretch test of the filum terminale, reduced viscoelasticity owing to decreased elastin and increased fibrous tissue in the filum terminale is thought to be the cause of OTCS. The pathoembryogenesis of how such a result appears during medullary cord regression still needs further research. Interestingly, a nerve twig, which is reported to be involved in abnormal urodynamics [21], was found in all fila of our OTCS patients. We do not know the reason for urological dysfunction in patients with nerve twigs in fila. In summary, our findings are in line with previous studies showing a near absence of elastin fibers and a high fibrous tissue percentage.

Numerous studies have reported on the positive effects of untethering for OTCS [1, 3, 14, 15]. One study that reviewed previous articles reported that overall, 78.3% of 289 patients improved after surgery [22]. In contrast, Steinbok et al. [19] recently published a randomized controlled pilot study, reporting a slight but non-significant improvement (6% vs 4%) in the urodynamic score. However, the study was limited to patients who had refractory bladder and bowel dysfunction for approximately 5 years, which is comparatively

longer than our study group. Hence, the negative result in a randomized controlled study of chronic refractory cases is understandable considering that prompt intervention improved patients with sudden onset and progression in our study. Moreover, since the studies did not separate primary and secondary urinary incontinence, the results may have underestimated the surgical outcomes.

In this study, the case of patient 3 is interesting. The patient recovered during the time she was being followed up at our institution. The reasons for her symptom aggravation may be multifactorial; one of the factors is thought to be her low compliance with CIC. The patient did not perform CIC as scheduled, which may have resulted in symptom re-aggravation. The lesson from the case is that postoperative urological care is as important as surgery, especially for those who have not fully recovered, because conserving kidney function is one of the primary goals of untethering surgery.

The case 2 is also interesting and confounding. The case raises the question of how untethering surgery improved the symptoms despite a normal-looking filum terminale during surgery. One hypothesis is the patient position during surgery, as we maintained lumbar lordosis to avoid spinal cord traction because spinal cord stretching caused by flexion during surgery may induce ischemic injury [27]. Moreover, since we did not perform a stretch test on the filum terminale as Yamada et al. [27] did, filum terminale elasticity could not be evaluated and considered normal. Therefore, we currently do not know how untethering surgery improved the symptoms, and further research is needed.

Case 5 had no improvement after untethering surgery, and no tightly tethered filum terminale was observed during surgery. The patient had an insidious onset of symptoms with a static preoperative course, which was different from the patients who had permanent or temporal improvement. Surgical decisions for patients with suspected OTCS with insidious symptom onset and static preoperative course are difficult. Nevertheless, we cannot totally exclude the possibility of improvement in those patients because of presence of typical DSD. Therefore, untethering surgery should be considered in patients in whom OTCS is suspected until further investigations on more patients reveal that patients with sudden symptom onset and progression are the only ones who can benefit from surgery, as there is a low complication rate with filum terminale untethering surgery [10].

There are limitations in this study. This was a retrospective study with a small number of patients. Due to the low prevalence of OTCS, it is challenging to enroll a large number of patients from a single institution. Nevertheless, the patients' symptoms and signs were objectively confirmed by UDSs, EMGs, and NCSs, and the patients were not young children, in whom clinical manifestations and UDS findings are less reliable. In addition, histological comparison was performed with TCS patients, not with healthy controls. However, our results showed a high portion of fibrotic tissue, as shown in the definite TCS patients, who are already known to have more filum fibrotic tissue than healthy controls. A more systematic review of highly selected patients is mandatory.

Conclusion

Based on our results, OTCS is definitely an existing entity, although exquisitely rare. Nonetheless, OTCS is curable when an accurate diagnosis is made and appropriate treatment is given. Fibrous tissue seems to be one of the reasons for cord tethering despite normal imaging findings. A sudden onset of urinary dysfunction or pain with progression, along with DSD proven by UDSs, is the best indication for untethering surgery in OTCS patients. Surgery should be performed promptly to avoid missing the proper time window. Moreover, postoperative care should be continuously provided to avoid urinary symptom re-aggravation because neurogenic bladder improvement is not a rapid process and may not be complete.

Author contribution All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by Jeyul Yang, Jae-KyungWon, Kwanjin Park, and Kyu-Chang Wang. The first draft of the manuscript was written by Jeyul Yang, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

Availability of data and materials The datasets generated during and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval The present study was approved by the institutional review board of the Seoul National University Hospital (IRB no. 2004–104-1118). Informed consent was waived by the IRB since this study was retrospective.

Conflict of interest The authors declare no conflicts of interest.

References

1. Albright AL (2005) Occult tight filum terminale syndrome: results of surgical untethering. Pediatr Neurosurg 41:58. https://doi.org/10.1159/000084869
2. Austin PF, Bauer SB, Bower W, Chase J, Franco I, Hoebeke P, Rittig S, Walle JV, von Gontard A, Wright A, Yang SS, Nevéus T (2016) The standardization of terminology of lower urinary tract function in children and adolescents: update report from the standardization committee of the International Children's Continence

Society. Neurourol Urodyn 35:471–481. https://doi.org/10.1002/nau.22751
3. Bao N, Chen ZH, Gu S, Chen QM, Jin HM, Shi CR (2007) Tight filum terminale syndrome in children: analysis based on positioning of the conus and absence or presence of lumbosacral lipoma. Childs Nerv Syst 23:1129–1134. https://doi.org/10.1007/s00381-007-0376-8
4. Brown E, Matthes JC, Bazan CI, Jinkins JR (1994) Prevalence of incidental intraspinal lipoma of the lumbosacral spine as determined by MRI. Spine 19:833–836. https://doi.org/10.1097/00007632-199404000-00018
5. Bulsara KR, Zomorodi AR, Enterline DS, George TM (2004) The value of magnetic resonance imaging in the evaluation of fatty filum terminale. Neurosurgery 54:375–380. https://doi.org/10.1227/01.NEU.0000103451.63301.0B
6. Cools MJ, Al-Holou WN, Stetler WR, Wilson TJ, Muraszko KM, Ibrahim M, Marca FL, Garton HJL, Maher CO (2014) Filum terminale lipomas: imaging prevalence, natural history, and conus position. J Neurosurg Pediatr 13:559–567. https://doi.org/10.3171/2014.2.peds13528
7. Hendson G, Dunham C, Steinbok P (2016) Histopathology of the filum terminale in children with and without tethered cord syndrome with attention to the elastic tissue within the filum. Childs Nerv Syst 32:1683–1692. https://doi.org/10.1007/s00381-016-3123-1
8. Lee JY, Kim KH, Park K, Wang K-C (2020) Retethering: a neurosurgical viewpoint. J Korean Neurosurg Soc 63:346–357. https://doi.org/10.3340/jkns.2020.0039
9. Loening-Baucke V (1997) Urinary incontinence and urinary tract infection and their resolution with treatment of chronic constipation of childhood. Pediatrics 100:228–232. https://doi.org/10.1542/peds.100.2.228
10. Ogiwara H, Lyszczarz A, Alden TD, Bowman RM, McLone DG, Tomita T (2011) Retethering of transected fatty filum terminales. J Neurosurg Pediatr 7:42–46. https://doi.org/10.3171/2010.10.peds09550
11. Schultz-Lampel D, Steuber C, Hoyer PF, Bachmann CJ, Marschall-Kehrel D, Bachmann H (2011) Urinary incontinence in children. Dtsch Arztebl Int 108:613–620. https://doi.org/10.3238/arztebl.2011.0613
12. Selçuki M, Coşkun K (1998) Management of tight filum terminale syndrome: with special emphasis on normal level conus medullaris (NLCM). Surg Neurol 50:318–322. https://doi.org/10.1016/S0090-3019(97)00377-7
13. Selcuki M, Ünlü A, Uğur HÇ, Soygür T, Arıkan N, Selcuki D (2000) Patients with urinary incontinence often benefit from surgical detethering of tight filum terminale. Childs Nerv Syst 16:150–154. https://doi.org/10.1007/s003810050482
14. Selden NR (2006) Occult tethered cord syndrome: the case for surgery. J Neurosurg Pediatr 104:302–304. https://doi.org/10.3171/ped.2006.104.5.302
15. Selden NR (2007) Minimal tethered cord syndrome: what's necessary to justify a new surgical indication? Neurosurg Focus 23:1–5. https://doi.org/10.3171/FOC-07/08/E1
16. Selden NR, Nixon RR, Skoog SR, Lashley DB (2006) Minimal tethered cord syndrome associated with thickening of the terminal filum. J Neurosurg Pediatr 105:214–218. https://doi.org/10.3171/ped.2006.105.3.214
17. Steinbok P, Garton HJ, Gupta N (2006) Occult tethered cord syndrome: a survey of practice patterns. J Neurosurg Pediatr 104:309–313. https://doi.org/10.3171/ped.2006.104.5.309
18. Steinbok P, Kariyattil R, MacNeily AE (2007) Comparison of section of filum terminale and non-neurosurgical management for urinary incontinence in patients with normal conus position and possible occult tethered cord syndrome. Neurosurgery 61:550–556. https://doi.org/10.1227/01.neu.0000290902.07943.e6
19. Steinbok P, MacNeily AE, Hengel AR, Afshar K, Landgraf JM, Hader W, Pugh J (2016) Filum section for urinary incontinence in children with occult tethered cord syndrome: a randomized, controlled pilot study. J Urol 195:1183–1188. https://doi.org/10.1016/j.juro.2015.09.082
20. Tehli O, Hodaj I, Kural C, Solmaz I, Onguru O, Izci Y (2011) A comparative study of histopathological analysis of filum terminale in patients with tethered cord syndrome and in normal human fetuses. Pediatr Neurosurg 47:412–416. https://doi.org/10.1159/000338981
21. Thompson EM, Strong MJ, Warren G, Woltjer RL, Selden NR (2014) Clinical significance of imaging and histological characteristics of filum terminale in tethered cord syndrome. J Neurosurg Pediatr 13:255–259. https://doi.org/10.3171/2013.12.peds13370
22. Tu A, Steinbok P (2013) Occult tethered cord syndrome: a review. Childs Nerv Syst 29:1635–1640. https://doi.org/10.1007/s00381-013-2129-1
23. Tuite GF, Thompson DN, Austin PF, Bauer SB (2018) Evaluation and management of tethered cord syndrome in occult spinal dysraphism: recommendations from the International Children's Continence Society. Neurourol Urodyn 37:890–903. https://doi.org/10.1002/nau.23382
24. Warder DE, Oakes WJ (1993) Tethered cord syndrome and the conus in a normal position. Neurosurgery 33:374–378. https://doi.org/10.1227/00006123-199309000-00004
25. Wehby MC, O'Hollaren PS, Abtin K, Hume JL, Richards BJ (2004) Occult tight filum terminale syndrome: results of surgical untethering. Pediatr Neurosurg 40:51–57. https://doi.org/10.1159/000078908
26. Wilson DA, Prince JR (1989) John Caffey award. MR imaging determination of the location of the normal conus medullaris throughout childhood. Am J Roentgenol 152:1029–1032. https://doi.org/10.2214/ajr.152.5.1029
27. Yamada S, Won DJ, Pezeshkpour G, Yamada BS, Yamada SM, Siddiqi J, Zouros A, Colohan AR (2007) Pathophysiology of tethered cord syndrome and similar complex disorders. Neurosurg Focus 23:1–10. https://doi.org/10.3171/FOC-07/08/E6
28. Yundt KD, Park TS, Kaufman BA (1997) Normal diameter of filum terminate in children: in vivo measurement. Pediatr Neurosurg 27:257–259. https://doi.org/10.1159/000121263