Tethered Cord Syndrome in Adulthood

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Abstract

Objective/Background: The standard treatment for tethered cord syndrome (operative vs nonoperative management) that presents in adulthood remains controversial. A comparative study of tethered cord syndrome in adulthood is needed.

Design: A retrospective chart-based analysis.

Methods: Patients admitted to Gulhane Military Medical Academy Department of Neurosurgery for management of caudal spinal cord tethering from June 1999 through December 2006 (N = 22).

Results: Conus level was normal in 1 patient with split cord malformation and dermal sinus. Tight terminal filum was found in 21 patients, including postrepair myelomeningocele tethered cord in 4, lipomyelomeningocele/meningocele in 8, split cord malformation in 3, dermal sinus in 7, and syringomyelia in 3. The most common complaints were back pain (15 patients, 68.1%), bladder dysfunction (8, 36.3%), fecal incontinence (2, 9.09%), and leg pain (7, 31.8%). One patient had hydrocephalus (4.5%). Ten of 22 patients underwent surgery; 8 of 10 patients had detethering; and 12 patients refused surgery. Postoperative cerebrospinal fluid leakage requiring reinforcement sutures occurred in 1 patient. There were no infectious complications. Neurologic status and outcomes were compared with preoperative findings.

Conclusions: Some patients refuse surgery despite severe neurologic disturbances. Neurosurgeons should fully explain the risks and benefits of surgery for tethered cord to the patient and family. A much larger and prospective randomized series is needed to determine the effects of operative vs nonoperative management of tethered cord syndrome in adulthood.

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Key Words: Tethered cord syndrome; Filum terminale syndrome, adult; Split cord malformation; Meningocele; Lipomyelomeningocele; Split cord malformation; Dermal sinus; Syringomyelia

INTRODUCTION

Tethered cord syndrome (TCS) is a stretch-induced functional disorder of the spinal cord with its caudal part anchored by an inelastic structure (1). Garceau first described the “filum terminale syndrome” in 1953 in 3 patients (2). Two decades later, in 1976, Hoffman and colleagues coined the term “tethered spinal cord” to describe the symptoms of their patients with an elongated spinal cord and a thick filum terminale (3). Yamada and others broadened the stretch-induced functional disorder to patients with other anomalies in 1981 (4). Myelomeningocele, lipoma, lipomyelomeningocele, diastematomyelia, meningocele manqué, and

dermoid sinus were included in this category but only when parallelism between the oxidative metabolic changes and neurologic status exists.

Symptoms related to a congenital tethered cord occur most commonly in childhood, so it was initially regarded as a pediatric problem; but in many patients, the diagnosis is not established until symptoms manifest in adulthood. It is well established that early surgical intervention of congenital TCS in children prevents neurosurgical deterioration. The number of adults in whom congenital TCS is diagnosed continues to grow as a result of better imaging and recognition of this syndrome. Pediatric TCS has been well studied in the literature, but much of the information regarding the adult population is still being defined. Patients who never undergo treatment for TCS likely have an elevated risk of developing symptoms with advancing age (5).

Standard treatment for TCS diagnosed in adulthood remains controversial. Results of recent clinical studies of surgical intervention in adulthood are encouraging (5–7).

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Table 1. Summary of Demographic and Clinical Data Obtained in 10 Patients Who Underwent Surgery

Case No.	Age (y)	Sex	Diagnosis	Operation	Conus Level	Follow-up Time (mo)	Neurologic Examination
1	22	Male	TCS	DeT	S2	12	Urgency
2	22	Male	TCS + DS	DeT + DS resection	L5	6	Cutaneous stigmata (hypertrichosis), dermal sinus
3	20	Male	TCS + diastematomyelia at L4 syringomyelia	DeT + resection of bony septum	L4	12	Cutaneous stigmata (hypertrichosis), altered sensation
4	21	Male	TCS + DS + lipomeningocele	DeT + M and DS resection	L5	6	Cutaneous stigmata (hypertrichosis)
5	21	Male	TCS + lipomyelomeningocele	DeT	S1	6	Myelomeningocele repair in childhood, urge incontinence
6	25	Male	TCS + lipomyelomeningocele	DeT	S1	12	Urgency
7	20	Male	TCS + meningo-myelocele	DeT + M resection	L4–5	12	Altered sensation, urgency, fecal incontinence
8	21	Male	TCS + SCM + DS	DeT + resection of fibrous band and DS	L5	12	Cutaneous stigmata (hypertrichosis), altered sensation
9	21	Male	TCS + SCM	DeT + resection of fibrous band	L4–5	12	Urgency, sexual dysfunction, muscle weakness, altered sensation
10	23	Male	TCS + DS + hydrocephalus	DeT + DS resection	L5	6	Myelomeningocele repair in childhood, fecal incontinence, urgency, bilateral pes cavus

TCS, tethered cord syndrome; DS, dermal sinus; SCM, split cord malformation; DeT, detethering.

Surgical intervention is usually indicated based on an expected natural history of disease progression in the absence of treatment. In our clinical experience, some adults with TCS decline surgery despite severe neurologic deficit. To clarify the debate on the intervention among neurosurgeons in clinical practice, we studied the surgical indications and outcomes for TCS in a retrospective review of 22 patients.

METHODS

All patients admitted to Gulhane Military Medical Academy Department of Neurosurgery for management of caudal spinal cord tethering from June 1999 through December 2006 were identified. Because some cases were published previously, the patients who were operated on by K. M. Akay, MD, were excluded from the study (8). Three patients underwent surgery in other centers and had follow up with their previous records in our center. Patient age in this study ranged from 20 to 29 years (mean 22.3 y). There were 21 men and 1 woman. Clinical charts, operative records, and follow-up data were reviewed for each patient. Mean duration of follow up for the 12 patients who declined surgery was 7 months (range 1–

12 mo). Patients who refused surgery answered a series of questions regarding their decision, their symptoms, and their lifestyles (Table 3). Mean duration of follow up for the 10 patients who had surgery was 9.6 months (range 6–12 mo).

RESULTS

Overall, tight terminal filum was found in 21 patients. Conus level was normal in 1 patient who presented with split cord malformation and dermal sinus. This patient refused surgery. Our TCS series included postrepair myelomeningocele in 4 patients, lipomyelomeningocele/meningocele in 8, split cord malformation in 3, dermal sinus in 7, and syringomyelia in 3. The most common complaint was back pain in 15 patients (68.1%) followed by bladder dysfunction in 8 (36.3%), fecal incontinence in 2 (9.09%), leg pain in 7 (31.8%), and hydrocephalus in 1 (4.5%).

Operation

Ten patients had operations for detethering. During surgery, in patients with lipomyelomeningocele and myelomeningocele, as much dissection as possible was

Table 2. Clinical Outcomes of 10 Patients Who Underwent Operation

Case No.	Back Pain Preop/Postop	Leg Pain, Back Pain Preop/Postop	Muscle Weakness, Back Pain Preop/Postop	Bladder Dysfunction, Back Pain Preop/Postop	Fecal Incontinence, Back Pain Preop/Postop
1	+/-	+/-	-/-	+/improved	-/-
2	+/-	-/-	-/-	-/-	-/-
3	+/+	+/-	-/-	-/-	-/-
4	+/+	-/-	-/-	-/-	-/-
5	+/+	+/-	-/-	+/improved	-/-
6	-/-	-/-	-/-	+/+	-/-
7	-/-	-/-	-/-	+/+	+/+
8	+/+	-/-	-/-	-/-	-/-
9	+/-	+/-	+/-	+/improved	-/-
10	+/-	-/-	-/-	+/improved	+/+

Preop, preoperatively; postop, postoperatively.

performed around the dorsal cord. In the patient with diastematomyelia (case 3), the bony septum was removed. In 2 other patients with split cord malformation (cases 8 and 9), fibrous bands and attachments were excised. Dermal sinuses were excised when present (Table 1).

Complications

Cerebrospinal fluid leakage occurred in 1 patient after operation; this was controlled with reinforcement sutures. There were no infectious complications and no new neurologic deficits after detethering. In our series, urinary tract infection was not a problem in either the operative or the nonoperative groups. None of the patients was admitted to hospital for urinary tract infection during our follow-up period.

Outcomes

Back Pain. Of the 8 of 10 patients who presented with preoperative progressive back pain, 4 were improved after operation.

Leg Pain. Of the 2 of 10 patients who presented with preoperative progressive leg pain, all who underwent surgery were improved after operation.

Bladder Dysfunction. Overall, 7 (31.8%) of 22 patients presented with urgency. Of 10 patients who chose surgery, 6 had preoperative urgency; 4 of them improved after surgery. Case 1, a patient with spina bifida occulta who presented with urgency, gradually improved to normal bladder function 6 months after surgery. Case 5, who had had myelomeningocele repair in childhood and presented with tethered cord and

Table 3. Questions Posed to and Responses of Patients Who Refused Surgery

Question Number	Question	Patient Response
1.	Were you operated on in childhood for tethered cord syndrome or meningocele?	Two patients answered yes.
2.	Is there any drainage from your back?	One patient answered yes.
3.	What is your urinary condition? (a) urinary incontinence, (b) urgency, (c) normal urinary function	Two patients indicated urinary incontinence, and 3 complained of urgency. Seven patients described their function as normal.
4.	Do you have fecal incontinence?	One patient answered yes.
5.	Have you ever been offered operative intervention by any neurosurgeon for tethered cord release?	All of the patients answered no.
6.	Is it your decision, refusing the operation?	All of the patients answered yes.
7.	Is it your family's preference to refuse the operation?	Six patients said yes.
8.	Have you ever been assured by any neurosurgeon that your condition cannot improve?	Four patients answered yes.
9.	What is your lifestyle? (a) very active, (b) active, (c) not active.	One patient was very active, 6 were active, and 5 were not active.
10.	How long is the duration of your symptoms?	One patient was suffering from back pain for 6 months; the rest did not give a specific duration.
11.	Are your symptoms progressive?	Case 3 complained of progressive back pain for 6 months, but this was related to exertion.

Figure 1. Patient with tethered cord syndrome and lipomyelomeningocele (case 8) who declined detethering. (a) T1-weighted image of sagittal lumbar magnetic resonance imaging. (b) T1-weighted image of axial lumbar magnetic resonance imaging at the level of lipomyelomeningocele.

Figure 2. Patient with tethered cord syndrome, meningocele, agenesis of corpus callosum, cutaneous stigmata (hypertrichosis), altered sensation, and leg length discrepancy who refused surgery (case 10). (a) T2-weighted sagittal magnetic resonance imaging of lumbar region. (b) T1-weighted image of sagittal cranial magnetic resonance imaging.

myelomeningocele, had urinary incontinence that improved to urgency after detethering.

Fecal Incontinence. Overall, 3 of 22 patients presented with fecal incontinence and 2 of them underwent surgery. Fecal incontinence was not a progressive symptom, and they had no improvement in this complaint (Table 2).

Reasons for Rejection of Operation. Of the 12 patients who declined surgery, 2 with spina bifida aperta had undergone meningomyelocele repair during childhood (cases 2 and 9) and did not want to undergo any new surgery. Case 8, whose conus level was at L4 with tethered cord and myelomeningocele, refused surgery despite fecal and urinary incontinence and muscle weakness (Figure 1a and b). Case 10, who presented with dysgenesis of corpus callosum, cutaneous stigmata (hypertrichosis), altered sensation, and leg length discrepancy, also refused detethering (Figure 2a and b). Case 11, whose conus level was at L3, presented with a dermal sinus. Radiologic intervention revealed split cord malformation and meningocele at L2-L3 level, but neurologic examination was completely normal (Figure 3a and b). This patient refused closure of dermal sinus, meningocele repair, and fibrous band detachment. Five patients (cases 3, 5, 6, 11, and 12) with asymptomatic tethered cord refused surgery. Four patients (cases 1, 2, 8, and 9) refused surgery because they had been told in the past that their condition could not be improved (Table 4).

DISCUSSION

Tethered cord syndrome is a descriptive term used to indicate progressive neurologic deficits. In our paper, we have not used the term “syndrome” for asymptomatic tethered cord (Table 3). Three categories of TCS were recently described by Yamada and colleagues (1). The first category includes lumbosacral cord anchored by an inelastic filum. The second category includes caudal myelomeningoceles and many sacral myelomeningoceles. The third category is divided into 2 groups. The first group includes patients with paraplegia and lipomyelomeningocele and myelomeningocele who apparently have no functional lumbosacral neurons. No neurologic benefit is expected from surgery in this group. The second group includes asymptomatic patients with an elongated cord and a thick filum. They need close observation for onset of subtle symptoms, particularly incontinence, which if untreated becomes quickly irreversible (1). Besides Yamada’s classification, Van

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Figure 3. Patient with tethered cord, split cord malformation at L1 level, dermal sinus, and meningocele (case 11), who declined detethering. (a) Proton density-weighted image of sagittal lumbar magnetic resonance imaging. (b) T2-weighted image of axial lumbar magnetic resonance imaging at the level of split cord malformation.

Table 4. Summary of Demographic and Clinical Data for 12 Patients Who Declined Surgery

Case No.	Age (y)	Sex	Diagnosis	Conus Level	Follow up (mo)	Neurologic Examination	Reason
1	20	Male	SCM at L1 + DS	L2	6	Paresthesia at legs + cutaneous stigmata + hypertrichosis + urgency	Counseled in another center
2	20	Male	TCS	L5	12	Myelomeningocele repair in childhood + muscular weakness	Counseled in another center
3	24	Male	Tethered cord	L5	12	Normal examination	Patient preference
4	29	Male	TCS	L4	12	Paresthesia at legs	Patient preference
5	20	Male	Tethered cord + DS	L4	6	Cutaneous stigmata (hypertrichosis)	Patient preference
6	29	Male	Tethered cord + diastematomyelia at L1	L4	1	Normal examination	Patient preference
7	24	Male	TCS + diastematomyelia at L1 + syringomyelia	L4	1	Muscle weakness	Patient preference
8	20	Male	TCS + lipomyelomeningocele	L4	1	Fecal incontinence	Counseled in another center
9	21	Male	TCS + meningocele	L5	12	Meningocele repair in childhood + foot deformity + bilateral pes cavus	Counseled in another center
10	21	Male	TCS + meningocele	L5	12	Dysgenesis of corpus callosum + cutaneous stigmata (hypertrichosis) + altered sensation-leg length discrepancy	Patient/family preference
11	21	Male	Tethered cord + SCM at L1 + DS + meningocele	L3	3	Normal examination	Patient preference
12	26	Female	Tethered cord + DS + syringomyelia	L4	6	Cutaneous stigmata (hypertrichosis)	Patient preference

TCS, tethered cord syndrome; DS, dermal sinus; SCM, split cord malformation; S, syringomyelia.

Leeuwen and others reported the clinical outcomes after detethering surgery in 4 patient subgroups based on the origin of tethering: (a) postrepair myelomeningocele, (b) terminal filum lipoma and tight terminal filum, (c) lipomyelomeningocele and conus lipoma, and (d) split cord malformation (7).

The incidence of occult spinal dysraphism is unknown, and although it is likely that some patients remain asymptomatic and a diagnosis is never made, a subset of patients with the congenital syndrome develops progressive symptoms and signs in adulthood.

Regarding the diagnosis and treatment of TCS, most neurosurgic experience is in pediatric patients. Because it is generally established that signs and symptoms of TCS worsen as patients age, most pediatric neurosurgeons believe that an infant or young child in whom TCS has been diagnosed, regardless of origin, should undergo a detethering procedure (3,9,10). However, there is much debate over the surgical management of congenital TCS presenting in childhood. Whereas some authors advocate prophylactic surgery, others suggest that surgery should be performed only when symptom progression develops. In our series, 10 symptomatic patients underwent

surgery. Of the 12 patients who declined surgery, 5 were asymptomatic. Three of the 5 asymptomatic patients had normal physical examinations and 2 had cutaneous stigmata (hypertrichosis) (Table 3).

Adults with TCS present with pain more often and demonstrate bladder dysfunction less often than do children with TCS (8). It is a challenge for the neurosurgeon to offer a surgical intervention to a patient who has no neurologic deficit or bladder dysfunction and only mild back and/or leg pain. Lee and colleagues' series of 60 patients with adult tethered cord showed clinical outcomes with low risk of neurologic complications. In this series, back pain and leg pain improved significantly (78–83%) regardless of the origin of the tethering (6). Rajpal and others reported that the symptoms that improved the most after surgical correction were back pain (65%) and bowel/bladder dysfunction (62%) (5). However, in our series, improvement in back pain was seen in 57.1% with this symptom, improvement in leg pain was 100%, and improvement in bladder dysfunction was 66.6%.

Lad and colleagues presented nationwide estimates of outcomes and complication rates after surgery for TCS in the United States from 1993 through 2002 (11).

According to that study, approximately 90% of patients were 44 years of age or younger, 71.0% were 17 years of age or younger, and 18.6% were 18 to 44 years of age. The 1.8% of patients who were 65 years of age or older experienced a nearly threefold increase in complication rate (20.3%) compared with those who were 17 years of age or younger (7.0%). Patients 65 years of age or older had a threefold increased risk of adverse outcome compared with patients 18 to 44 years of age. Adult patients with tethered cord often have preoperative deficits secondary to tethering, such as bladder dysfunction, which may worsen postoperatively (6). Other authors have reported a similar adverse association between age and lumbar spine surgery (12).

Rates of surgical complications have not been reported in all studies. These areas of concern highlight the need for more accurate and consistent reporting of the surgical outcomes of adult patients with TCS (6). In our series, cerebrospinal fluid leakage occurred in 1 patient postoperatively. There were no infectious complications. Two patients experienced transient postoperative urinary retention that resolved 2 days later. This may have been related to either the operation or the anesthesia. None of the patients experienced neurologic deterioration after the operation.

It is remarkable that only 10 of 22 patients accepted surgical intervention even though some of them had severe symptoms affecting quality of life, such as fecal incontinence, urinary incontinence, and muscle weakness. Whereas the decision to undergo surgery must be left to the patient, we used a questionnaire to explore the decisions of the 12 patients who did not accept operation. According to their responses, 2 patients did not accept the operation because they had no neurologic symptoms. One patient who had undergone meningocele repair in childhood did not want to undergo a second operation. The rest of the patients, who had such symptoms as peripheral paresthesias and cutaneous stigmata (hypertrichosis), were afraid of unexpected neurologic complications. Four patients refused operation because they had been convinced in the past that their condition was not amenable to intervention. For example, the patient with tethered cord plus lipomyelomeningocele and fecal incontinence had been assured in another center that his condition would never improve (Table 4).

The recommendations based on the survey are as follows:

1. If the patient has an active lifestyle with tolerable symptoms during the day without a pattern of progression, do not advise operation for tethered cord release.
2. If the patient does not have an active lifestyle and has mildly tolerable symptoms during the day without an

overall pattern of progression, do not offer operation for tethered cord release.

3. If a patient does not have an active lifestyle and has intolerable symptoms during the day, the decision should be based on patient/family preferences and advice of the neurosurgeon or medical center.

CONCLUSIONS

Some patients do not consent to detethering procedures despite severe neurologic disturbances. Monitoring for progressive symptomatology is warranted for asymptomatic patients who decline prophylactic surgery. We recommend that neurosurgeons explain the risks and benefits of the operation for tethered cord to the patient and family and let them decide. A larger prospective, randomized series is needed to determine the effect of operative vs nonoperative management of TCS in adulthood.

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