

# Outcome of Surgical Management for Tethered Spinal Cord

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**Objective :** The tethered cord syndrome results in progressive neurological deficits. Although it may remain controversial, many physicians recommend definitive surgery to untether the cord as soon as this condition is identified. We retrospectively evaluate the pre-operative and post-operative course of 38 tethered cord patients with spinal dysraphism in an attempt to learn the natural history of the disease and to determine the effectiveness of the surgical treatment.

**Methods :** The medical records, operation notes and radiographs were evaluated. The follow up period ranged from 4 months to 12 years with a mean follow-up of 28.6 months. Twenty-seven patients were younger than 15 years of age.

**Results :** At presentation, 26 of the patients were asymptomatic. In three of 11 adult symptomatic patients, their neurological deficits worsened after trauma or exercise. Improvement of motor strength was documented in two out of 5 patients. Five of nine patients with bladder symptoms improved, however, none had a complete return of their bladder function.

**Conclusion :** Childhood patients were less symptomatic than the adult patients. Adult patients showed progression of their symptoms that were not improved even after the operation in most of the cases. Asymptomatic tethered spinal cord can be symptomatic as time passes by and even at an old age. Future research should be focused on the operative methods to prevent the delayed deterioration after surgery, rather than on the usefulness of preventive surgery in asymptomatic patients.

**KEY WORDS :** Tethered cord syndrome · Surgical management · Neurological deficits.

## Introduction

Tethered spinal cord syndrome is a descriptive term used to indicate progressive neurological deficits in individuals with an abnormal fixation of the spinal cord, and it is seen in patients with spinal dysraphism. It is anatomically associated with a low-lying position of the conus, and the common causes of conus entrapment in occult spinal dysraphism are lumbosacral lipoma, diastematomyelia and fibrous adhesions<sup>4,5</sup>. Limitation in movement of the spinal cord results in stretching the spinal cord and so this compromises its blood supply. Ischemic damage to the neural tissue is thought to be the main cause of the neurological deterioration<sup>21</sup>.

The tethered cord syndrome results in progressive neurological deficits. The age at deterioration is variable and it not just related to the periods of growth spurts<sup>3,8,10</sup>. A late onset presentation in adults is also well recognized<sup>19,23,30</sup>. The patients usually develop urinary symptoms due to neurogenic bladder,

motor impairment of the lower extremities, sensory disturbance and secondary orthopedic deformities<sup>3</sup>.

There are many controversial issues in the management of the tethered cord syndrome, including timing, surgical method, and long-term outcome of the operative treatment. Many authors recommend definitive surgery to untether the cord as soon as this condition is identified early in life, and preferably before the onset of neurological deficits<sup>3,8,10,12,23,25</sup>. However, some groups reported disappointing results after long-term follow-up of their operative cases<sup>24,25</sup>. The debate can not be solved in near future, however, accumulated data about the results of the management of the disease will eventually give the answers.

The aim of this study is to elucidate the natural history of the tethered cord and the role of the surgical treatment. We retrospectively evaluated the pre-operative and post-operative course of 38 tethered cord patients with spinal dysraphism and discussed the role the surgery.

• Received : July 13, 2005 • Accepted : October 17, 2005

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## Materials and Methods

We retrospectively reviewed the files of 38 patients who underwent surgery for tethered cord syndrome in our hospital from May, 1990 to May, 2004. All operations were performed by the senior author. Operations were aimed to untether the spinal cord. The intraspinal lipomas were removed as much as possible, resecting behind the dorsal root entry area and below the exit of the last spinal rootlet. All tethered cords, except one, in which lipoma went out through the intervertebral foramen, were successfully untethered. During the closure of the dura, special care was taken to have the intradural space as wide as possible. Dural grafts using artificial dura were done if needed. The patients were irregularly followed up at the outpatient clinic by history taking and physical examination. Radiological and urodynamic follow-up were not performed routinely.

All the patients with lumbosacral lipoma and tethered cord were reviewed. Patients with postmyelomeningocele tethered cord, intramedullary dermoid and intrasacral meningocele were not included in this study. The medical records, operation notes and radiographs were evaluated, and the surgical results were collected from the patient's discharge summaries as well as the subsequent clinic records. The current status of the patients was determined from the most recent clinical evaluation or by telephone interview, and during the telephone interview, a standard set of questions was asked.

## Results

There were 16 men and 22 women. The age at surgery ranged from 2 months to 63 years with the median age of 19 months. Of these cases, 27 patients were younger than 15 years of age (Table 1). The mean follow-up period was 28.6 months (follow-up range: 4 months to 12 years).

The neuroimaging findings indicated that the conus medullaris was in a low position in all 38 patients. Based on the findings of the neuroimaging and the operations, the 34 intraspinal

**Table 1.** The age distribution of the 38 tethered spinal cord patients at the time of untethering surgery in our hospital

Age	Sex		Total
	Male	Female	
0 - 6 mo	7	8	15
6 - 12 mo	1	1	2
1 - 5 yrs	3	1	4
5 - 10	1	3	4
10 - 15	1	1	2
15 - 25	0	1	1
25 - 40	1	4	5
> 40	2	3	5

lipomas could be classified into three types according to Chapman<sup>3)</sup>: 1) the dorsal type or with lipoma attached to the placode and neural elements suspended from the undersurface of the placode (13 cases), 2) the caudal type or with lipoma terminating the caudal portion of the cord through the dural defect (12 cases), 3) the transitional type or lipoma mixed with neural elements and the placode through the dural defect (9 cases). Other types of these lesions included a dermal sinus tract in 3 patients and a split cord malformation in one patient.

At presentation, 26 patients of all 38 patients were asymptomatic. The asymptomatic patients were all referred for an evaluation of the cutaneous manifestations. Preoperative examinations revealed cutaneous manifestations in 34 of 38 patients, bowel and bladder dysfunction in 10 patients, sensory disturbance such as decreased superficial sensation in 9 patients, pain in the legs or in the lumbosacral region in 3 patients and new or progressive weakness involving one or both extremities presented in 5 patients. Among the 34 patients who had skin lesions, the most common were fatty mass and skin dimples for 14 patients each, respectively, and this was followed by tail-like appendages in 5 patients and a depigmented region in one patient (Table 2).

In the 27 childhood group patients, the clinical manifestations were lumbosacral cutaneous abnormalities, urologic symptoms, pain and sensory disturbance in the order of frequency. In the 11 adult patients, sensory disturbance was most common symptom, and this was followed by cutaneous manifestations, bladder-bowel symptom, weakness and pain (Table 3).

**Table 2.** The main presenting signs and symptoms of patients with tethered cord syndrome

Signs and Symptoms	No. of cases (%)
Cutaneous manifestation	34 (89.4)
Fatty mass	14
Skin dimpling	14
Tail like appendage	5
Depigmented region	1
Lower extremities weakness	5 (13.1)
Back/leg pain	3 (7.8)
Sensory disturbance	9 (23.7)
Bowel/bladder dysfunction	10 (26.3)
No neurological deficits	26 (68.4)

**Table 3.** Comparison of the main neurological signs between childhood and adult patients

Neurological signs	Childhood(≤15 yrs)	Adult(>15yrs)
Skin stigma	27	7
Pain	1	2
Urologic symptom	2	7
Motor deficit	0	5
Sensory disturbance	1	8
No neurological deficit	24	1

**Table 4.** The surgical outcome in patients according to the age group

Symptom	Improved		No interval change		Worsening	
	≤15 yrs	>15 yrs	≤15 yrs	>15 yrs	≤15 yrs	>15 yrs
Pain	1	2	0	0	0	0
Motor	0	2	0	3	0	0
Sensory	1	4	0	3	0	1
Bladder	2	3	0	4	0	0

In 3 of the 11 adult symptomatic patients, the neurological deficit worsened after trauma, and this trauma were two falls and one headstand exercise in the three patients, respectively.

There was no operative mortality or severe morbidity in this series. Two patients developed leakage of their cerebrospinal fluid and they were conservatively managed quite successfully.

The postoperative follow-up period ranged from 4 months to 12 years (the mean was 28.6 months). Very good results regarding pain relief were achieved. Improvement in motor strength was documented for two out of 5 patients of the 9 patients with sensory complaints, 5 improved, one worsened and 3 were unchanged. The results for bladder dysfunction were disappointing. Five of nine patients were helped by surgery however, none of the patients had a complete return of their bladder function (Table 4).

## Discussion

The tethered cord syndrome was initially described by Garceau et al<sup>5</sup>) as the “filum terminale syndrome.” Primary tethering of the cord is found in the various anatomical types of closed spinal dysraphism, the most common of which are the lumbosacral lipomas. In many cases, they are associated with skin lesions<sup>8,20,22,26</sup>). In our series, the two main clinical features leading to diagnosis were cutaneous manifestations on the lower back and neurological disturbances. Cutaneous manifestations were present in 89.4% of our patients. The most common cutaneous lesion was a subcutaneous lipoma.

Chapman<sup>3</sup>) classified intraspinal lipomas into three types: caudal, dorsal and transitional, and they were related to the attachment pattern of the lipoma on the caudal portion of the cord. In our experience, 38.2% of the lipomas of the conus were dorsal, 35.2% were caudal, and 26.6% were transitional.

The pathophysiological mechanism for the deterioration of the neurological deficit is not clear. A stretching effect of the cord has been proposed as the main mechanism<sup>32,33</sup>). The oxidative metabolism is impaired in the spinal cord and the severity of the neuronal dysfunction parallels the degree of impairment for the oxidative mechanism<sup>32</sup>). Decreases in blood flow and glucose metabolic impairment are also known to contribute the deterioration<sup>13</sup>). Untethering and decompression of the mass effect are the beneficial mechanisms of surgery. Other

postulated mechanisms of the neurological deficit include myelodysplasia and arachnoiditis<sup>25</sup>). These are mechanisms that can not be unsolved by surgery, and they make the disease's natural history and long term surgical results unpredictable.

The natural history of primary tethered cord caused by intraspinal lipoma is not clearly known. Although it is likely that some patients remain asymptomatic and a diagnosis is never made, a subset of patients with the congenital syndrome develop progressive symptoms and signs in adulthood<sup>11,15,23,27,28</sup>). Sudden deterioration is possible, and such deterioration may occur after sudden movements or trauma that causes traction of the spinal cord, e.g. heavy lifting, delivery, bending movements, intercourse, straight leg raising exercises, prolonged sitting, trauma and others<sup>6,23</sup>). Most of the pediatric neurosurgeons in the major children's hospitals prefer early operation for the asymptomatic children<sup>18,31</sup>). Xenos et al<sup>31</sup>) showed, in their 69 case surgical series, that prophylactic surgery for asymptomatic patients did provide some protection from future neurological deterioration. Furthermore, symptomatic conus lipoma surgery was effective in stopping any further deterioration. Improvement in the neurological function can occur, but few patients experience a return to normal. The overall function and pre-existing sphincter dysfunction is not significantly altered by surgery<sup>6</sup>). Pang and Wilberger<sup>23</sup>) recommended surgical treatment for adults with a physically active life, as trauma can precipitate symptoms. Hoffman et al<sup>8</sup>) also recommended untethering surgery for patients with lumbosacral lipoma (lipomyelomeningocele) before they reached 6 months of age. According to McLone et al, individuals who have cutaneous manifestations of a tethered spinal cord should be studied and operated on as soon as is reasonable<sup>21,22</sup>). However, the long term effects of preventive surgery are a subject of debate. Hood et al<sup>9</sup>) indicated that surgical intervention is unnecessary in the absence of neurological deterioration. According to the series of Kulkarni et al<sup>17</sup>), among 53 asymptomatic children with spinal lipoma of conus, 13 patients (25%) exhibited neurological deterioration during the mean follow-up period of 4.4 years. There was no significant difference for the risk of neurological deterioration for patients who were treated conservatively and for those who underwent early surgery. Van Calenbergh et al<sup>29</sup>) reported on the results after surgery for symptomatic lipomas that showed evolution of the different symptoms. According to their series, the orthopedic symptoms most commonly progressed during the natural course of the disease, while pain and sphincter symptoms showed postoperative improvement. The debate will be solved only after more reliable research is done, including randomized long term studies for clarifying the natural course of the disease, and the natural disease course might be impossible to uncover in the near future.

Without precise knowledge about the exact mechanism of

deterioration, it is difficult to know the role of prophylactic surgery for the asymptomatic patient. However, the important things that we have to consider are what the prognoses of patients are when they are operated upon after neurological deterioration. According to Xenos<sup>31)</sup>, even if 68% of symptomatic patients improved after surgery, few patients return to their normal overall function and their preexisting sphincter dysfunction is not significantly altered by surgery. In our series, even though five of nine adult with sphincter impairment demonstrated some degree of improvement, none of them accomplished an effective recovery of their sphincteric symptoms. Some of the deterioration of symptoms developed suddenly after relatively minor trauma. We can not say whether their serious symptoms would not have developed if they had received early prophylactic surgery before becoming symptomatic. It is not reasonable to recommend observatory management to asymptomatic patient because we do not know their prognosis. Instead, the thing that we have to develop is the best surgical method for preventing the late deterioration. Several procedures were suggested, however, none of them have proved to be effective. Kang et al<sup>14)</sup> used the mega dural sac rebuilding method and they reported those patient on whom they used this method experienced no deterioration. Future research should be focused on developing the operative methods to prevent the delayed deterioration after surgery, rather than evaluating the usefulness of the preventive surgery in asymptomatic patients. Lipoma of the filum is not considered in this controversy because it has little operative risk and a good prognosis. Type I (dorsal) and II (caudal) lipomas are known to be the lower risk group for surgery<sup>14)</sup>. Bulsara et al<sup>2)</sup> and Kang et al<sup>14)</sup> reported that age was a factor for neurological deterioration. Kang et al<sup>14)</sup> reported less pronounced neurological deterioration after surgery before three years of age than after that age. Differentiating the high risk group from the low risk group is of crucial important to determine the effectiveness of preventive surgery.

In this paper, we retrospectively analyzed the clinical records of 38 patients, and all the asymptomatic cases were under 3 years old, while most of the patients older than 15 years showed urological or neurological abnormalities. Such a late onset of the symptoms in our series is consistent with the earlier reports on this clinical condition<sup>7,8,11,12,16,24)</sup>. Despite the above evidence, the natural history of tethered spinal cord cannot be determined from the data in this series because most of the patients were selected and referred to our department from other departments or hospitals. There will always be asymptomatic patients who are not detected by any form of medical survey or examination. The present study revealed that at least some tethered spinal cord patients develop neurological deficits with age, and surgical intervention will be of benefit to these patients.

## Conclusion

The childhood patients were less symptomatic than adults, however, a much longer follow-up is needed to elucidate the usefulness of preventive surgery in the asymptomatic patients. Adult patients showed progression of their symptoms, which were not improved even after the operation in most of the cases. Asymptomatic tethered spinal cord could become symptomatic as time passes by and even at old age.

Future research should be focused on developing operative methods to prevent the delayed deterioration after surgery, rather than evaluating the usefulness of the preventive surgery in asymptomatic patients.

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## Commentary

This excellent review summarizes the retrospective evaluation results of preoperative and postoperative course of 38 tethered cord patients with spinal dysraphism who underwent untethering surgery for tethered cord syndrome at a single institution over a 14-year period.

Of these cases, 27 patients were younger than 15 years of age

and 26 patients were asymptomatic on presentation. The authors also observed that only partial improvement in motor and sphincter function for the symptomatic adult patients even after surgery.

In my limited experience, I have also applied prophylactic surgery at an earlier age, after 6 months of age, for the asymptomatic infant patient in order to prevent deterioration.

The management of asymptomatic intraspinal lipomas is still an controversial issue, although many articles and book chapters on this condition recommend early prophylactic surgery at the time of diagnosis regardless of patients age or neurological function, on the basis of the idea that surgery offers improved outcomes compare with the natural history.

However, Pierre-Kahn et al suggested that tethering might not be the only factor causing neurological worsening. The incidence and patterns of neurological deterioration seemed to be very similar, regardless of whether early surgery was performed. This group suggest that conservative treatment of asymptomatic patients is preferred treatment option.

The present study is sizable, but is limited by non-randomization and lack of information on the long-term natural history. Despite of these limitation and the fact that does not fully answer the question, it does provide current information on recent treatment modality and it includes an appropriate discussion of the prevailing literature.

A more definitive randomized study will be required to clarify the relative efficacy of early surgery for tethered cord syndrome with intraspinal lipoma among asymptomatic patients.

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