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Journal of Neuroscience and Neurological Surgery
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Research Article

Functional Outcome after Fatty and Tight Filum Sectioning in a Diverse Cohort of Children with Tethered Cord Syndrome

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Received date: December 23, 2024; Accepted date: January 08, 2025; Published date: January 31, 2025

Citation: Stefanie Deininger, Marie Schuler-Ortoli, Christian Rainer Wirtz, Aurelia Peraud, (2025), Functional Outcome after Fatty and Tight Filum Sectioning in a Diverse Cohort of Children with Tethered Cord Syndrome, *J. Neuroscience and Neurological Surgery*, 17(1); **DOI:10.31579/2578-8868/350**

Abstract

Background: Tethered cord syndrome (TCS) encompasses a spectrum of conditions, from severe dysraphic disorders to milder forms like occult tethered cord. This study aims to evaluate the functional outcomes following surgical de-tethering in a pediatric population with TCS caused by a fatty or tight filum, including those with complex developmental anomalies.

Material and Methods: We conducted a retrospective analysis of 58 pediatric patients who underwent surgical de-tethering between January 2018 and February 2024. Data was collected on motor function, bladder and bowel dysfunction, and other preoperative symptoms. Functional outcomes were assessed via clinical exams, neurophysiological testing, and urodynamic studies.

Results: The cohort had a mean age of 5.5 years at surgery, with a follow-up period averaging 1.8 years. Preoperatively, 79.3% of patients presented with neurogenic bladder and 56.9% with bowel dysfunction. Significant improvements were observed in bladder function, with 26.1% of patients showing improvement and 73.9% maintaining stable function (p=0.04). Children with developmental anomalies, who had a higher incidence of bladder and bowel dysfunction, showed notable improvement, with 66.7% demonstrating motor function recovery and 100% achieving pain relief. No worsening of symptoms was noted during follow-up, and no cases of re-tethering were observed within the follow-up period.

Conclusion: Surgical de-tethering for TCS in children, including those with complex developmental anomalies, is safe and results in marked improvements in motor function, pain, and bladder control. Long-term follow-up is necessary to monitor potential re-tethering, particularly in patients with complex conditions. The results support a proactive approach to surgery, irrespective of the severity of preoperative symptoms.

Keywords: tethered cord; fatty filum; myelomeningocele; occult spinal dysraphism; spina bifida

Introduction

Tethered cord syndrome (TCS) encompasses a wide range of conditions, from severe dysraphic disorders like meningomyelocele and lipomeningomyelocele to milder forms such as occult tethered cord [1,2]. In the latter, patients may present with only external stigmata, while imaging reveals a tight or fatty filum and/or a low conus position (typically below L2). Regardless of the form, TCS is frequently associated with developmental comorbidities, such as complex anorectal and urogenital malformations [3,4].

Clinically, patients often present with progressive sensory and motor deficits in the lower limbs, alongside cutaneous, urological, and musculoskeletal abnormalities [5,6,7,8]. Research suggests that the traction on the spinal cord causes hypoxia and ischemia, leading to metabolic disruption and neuronal damage [9,10,11,12]. Symptom severity is thought to correlate with the extent of these metabolic disturbances [13,14]. However, postoperative improvement rates vary significantly, ranging from 10% to 90% [7,8,15]. While many studies have reported significant improvement in symptoms following surgery, long-term outcomes are inconsistent [6,7,16,17]. Some patients experience symptom recurrence, while others see little benefit from the procedure. Additionally, postoperative complications, including infections, cerebrospinal fluid leaks and re-tethering have been reported [6,18,19,20, 21].

There is a particular lack of outcome data for children with developmental comorbidities following TCS release [3]. These children often present with pre-existing bladder and bowel dysfunction, complicating the assessment of surgical impact. Some have undergone prior pull-through

surgeries for anal atresia, while others manage their conditions with a stoma or self-catheterization. Given these complexities, the effect of TCS release on this subgroup remains uncertain.

This study examines functional outcomes and the disease spectrum in children with tethered cord caused by fatty or tight filum, based on a diverse retrospective cohort from a high-volume pediatric neurosurgery unit.

Materials and Methods

The study protocol was approved by the local ethics committee (protocol #17-504 and protocol #202/20). Written informed consent for the surgery was given by all parents/patients.

All patients undergoing surgical de-tethering for fatty/tight filum at our institution from 01/2018 from 02/2024 were included. Two patients got external de-tethering by the same staff surgeon and follow-up at our institution. Routine follow-up consisted in patient (and/or parent) interview, clinical exam, neurophysiological and urodynamic testing [8,18].

The data analyzed included gender, age at the time of surgery, and preoperative symptoms related to TCS. These symptoms encompassed motor function disturbances: degree of paresis, gait ataxia, ambulation ability, and results from neurophysiological tests such as somatosensory evoked potentials (SEPs) and, if performed, motor evoked potentials (MEPs), bladder function (urodynamic findings including bladder capacity, compliance, post-void residual volume, frequency of urination, catheterization frequency if applicable, incidence of urinary tract infections, and presence of urge or stress incontinence), bowel function, back pain, as well as the type and severity of foot or spinal deformities. Additionally, the presence of associated developmental disorders or genetic alterations. Each diagnosis was confirmed through lumbar magnetic resonance imaging (MRI) in conjunction with clinical and electrophysiological findings (exemplary illustration is provided in figure 1A and 1B).

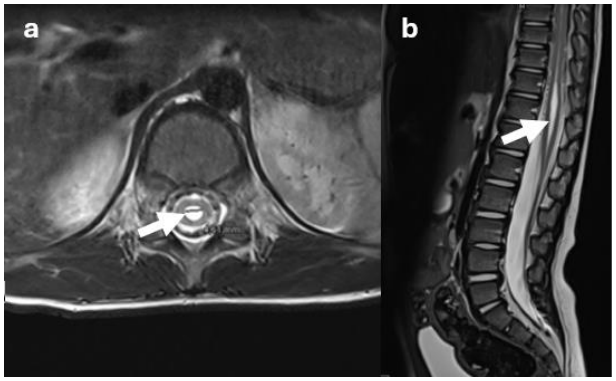
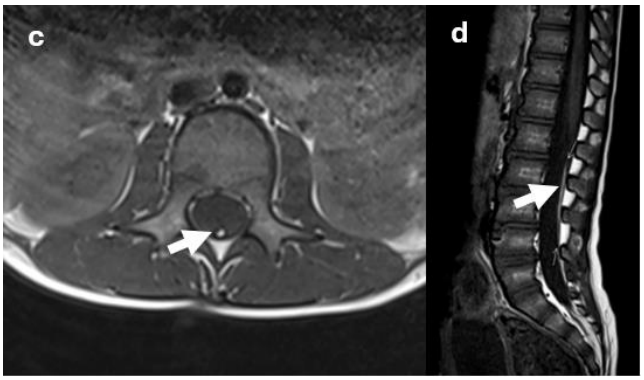


Figure 1A. Syringomyelia in a T2-weighted lumbar MRI

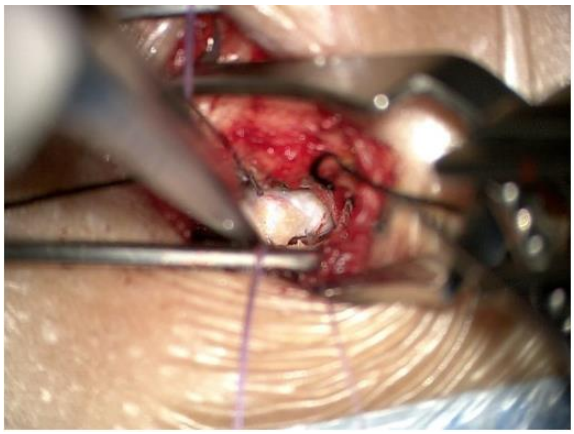


T2-weighted axial (a) and sagittal (b) MR image demonstrating a pointed conus medullaris with associated syringomyelia (arrows) with a maximum diameter of 5mm. The underlying reason is a tight fatty filum (arrows), which can be seen in the corresponding T1-weighted axial (c) as well as sagittal (d) MR images.

Figure 1B. Tight fatty filum in a T1-weighted lumbar MRI

The outcome measures included functional alterations of the symptoms and complications mentioned. The criteria for symptom improvement were defined as previously described [8].

The surgical management of the herein described cohort included the excision of any sinus tracts when present (n=3), as well as the removal of conus-lipomas (n=1) or dermoids (n=1). All patients underwent surgical de-tethering with continuous neurophysiological monitoring, including SEPs and MEPs, along with direct nerve root stimulation of the lower limb muscles and anal sphincters. The microsurgical procedure was performed through an ultrasound-guided, minimally invasive skin incision of approximately 1.5 cm and an interlaminar approach with undercutting of the ligamentum flavum. The dura was then opened in the midline over a length of 5 mm, secured with tag-up sutures on both sides, and CSF was allowed to drain. Subsequently, the filum terminale was identified among the nerve roots of the cauda equina using electrical stimulation, followed by coagulation and sectioning of the filum (figure 2 presents an illustrative example).



Intraoperative view of the tight fatty filum mobilized through a minimal dural opening before microsurgical transection.

Figure 2: Intraoperative view of a tight fatty filum

Quantitative analysis was conducted using GraphPad Prism 8, with data presented as proportions or mean with standard deviation (sd). Categorical data were analyzed using the chi-square test, and continuous data with paired or unpaired t-test. A p-value of ≤ 0.05 was considered statistically significant.

Results

The cohort (shown in table 1) consisted of 58 patients with a mean age at surgery of 5.5 ± 6.2 years and a follow-up duration averaging 1.8 ± 2.0 years. The male-to-female ratio was 62.1% to 37.9%.

Variable	Value
Patients (n, %)	**58 (100)**
Age at surgery (yrs, mean ± sd)	**5.5 ± 6.0**
Gender	
Male (n, %)	**36 (62.1)**
Female (n, %)	**22 (37.9)**
Follow-up since operation (yrs, mean ± sd)	**1.8 ± 2.0**
Additional neurological condition[1]	
Caudal regression syndrome (n, %)	**13 (22.4)**
Anorectal malformations (n, %)	**14 (24.1)**
Urogenital malformation (n, %)	**7 (12.1)**
Syndromic disease (n, %)	**10 (17.2)**
Radiological findings[1]	
Filum lipoma (n, %)	**14 (24.1)**
Syringomyelia (n, %)	**15 (25.9)**
Diastematomyelia (n, %)	**2 (3.4)**
Associated pathologies[1]	
Chiari malformation (n, %)	**2 (3.4)**
Type I (n, %)	**1 (1.7)**
Type II (n, %)	**1 (1.7)**
Hydrocephalus (n, %)	**3 (5.2)**
Encephalocele (n, %)	**1 (1.7)**
Craniosynostosis (n, %)	**1 (1.7)**
Dermal sinus (n, %)	**3 (5.2)**

[1] *more than one may apply; Abbreviations: n, number; yrs, years; sd, standard deviation.*

Table 1: Demographic characteristics

Of the entire cohort, 31 children (53.3%) presented with complex anorectal or urogenital malformations associated with tethered cord syndrome or additional syndromic disease. Among them, 13 exhibited caudal regression syndromes with 5 cases involving anal atresia, two children displaying a persistent urogenital sinus, and two identified with a VACTERL association. Additional four children were diagnosed with VACTERL (without caudal regression syndrome), including one case with anal atresia. One child showed a persistent urogenital sinus, and another presented with anal atresia (both without additional syndromic conditions). Other rare syndromic conditions included Trisomy 21 (n=2), Coffin-Siris syndrome (n=1), Kasabach-Merritt syndrome (n=1), and X-linked hydrocephalus (n=1) and 7 other various combined developmental disorders. Conus lipoma was present in 24.1% of patients, while syringomyelia was observed in 25.9%, and diastematomyelia in 3.4%. Chiari malformation and hydrocephalus were present in 3.4% and 5.2% of patients, respectively with 5.2% requiring a VP shunt. Children with

developmental anomalies underwent surgery at 3.9 ± 3.2 years, compared to 7.4 ± 7.7 years for healthy children (p=0.025).

Of particular interest was a 15-year-old girl who had undergone postnatal MMC closure, decompression of a Chiari II malformation and a ventriculoperitoneal shunt in former history. On presentation, she showed deteriorating urodynamics, and imaging revealed a tight filum terminale with a lipoma at the filum terminale.

Symptomatic tethering was the indication for surgery in most children. Table 2 presents the preoperative symptoms in descending order of frequency in the overall cohort: the most prevalent symptom was neurogenic bladder, affecting 79.3% of patients, followed by bowel dysfunction (56.9%).

Symptoms[1]	Overall cohort	Children with developmental anomalies	Healthy children	P Value
	N=58	*N=31*	*N=27*	
Neurogenic bladder (n, %)	**46 (79.3)**	**27 (87.1)**	**19 (70.4)**	**0.12**
Bowel dysfunction (n, %)	**33 (56.9)**	**25 (80.6)**	**8 (29.6)**	**<0.0001***
Motoric dysfunction (n, %)	**11 (19.0)**	**6 (19.4)**	**5 (18.5)**	**0.94**
Ambulatory (n, %)	**45 (77.6)**	**25 (80.6)**	**20 (74.1)**	**0.56**
Foot deformity (n, %)	**12 (20.7)**	**7 (22.6)**	**5 (18.5)**	**0.71**
Pain (n, %)	**10 (17.2)**	**2 (6.5)**	**8 (29.6)**	**0.02***
Asymptomatic (n, %)	**5 (8.6)**	**0**	**5 (18.5)**	**0.01***

*The proportions are shown relative to the number of children in the respective cohort. [1]more than one may apply; Abbreviations: n, number. *p<0.05.*

Table 2: Preoperative symptoms

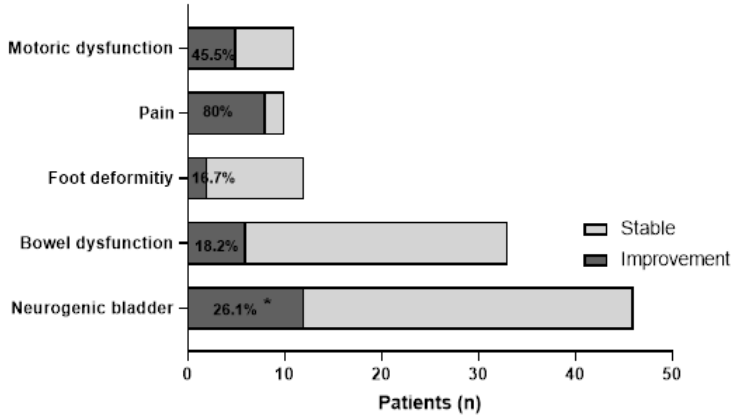
One 7-year-old boy with genetically confirmed Saethre-Chotzen syndrome was operated due to symptomatic re-tethering 4 years post-surgery with worsening somatosensory evoked potentials and progressive bowel incontinence. His initial surgery involved transection of the fatty filum and Chiari I decompression.

Five patients were asymptomatic preoperatively diagnosed due to stigmata like dermal sinus (n=2), sacral dimple (n=1), lumbosacral hairy patch (n=1) and lumbar hemangioma (n=1). Besides fatty/tight filum MRI showed syringomyelia in one case and three conus lipomas. The dermal sinuses were additionally removed during surgery.

When comparing symptoms, patients with developmental anomalies showed a higher incidence of bowel dysfunction (80.6% vs. 29.6%, p< 0.0001), whereas pain was more commonly reported among the healthy children (29.6% vs. 6.5%, p=0.02). Notably, all five asymptomatic patients were found exclusively within the healthy subgroup (p=0.01).

Figure 3 illustrates the functional improvements across all symptoms, highlighting the substantial postoperative recovery in pain and motor function. Of the children with developmental anomalies, 66.7% showed significant improvements in motor impairments, while 100% achieved complete pain relief. In contrast, 75% of healthy children also reported a reduction in pain symptoms.

Post-surgery, bladder dysfunction improved in 26.1% of patients (p=0.04), while 73.9% maintained stable symptoms. Two previously non-ambulatory patients gained the ability to walk. Foot deformities improved in 16.7% of cases, and bowel dysfunction in 18.2%, with most patients showing stable symptoms. Notably, no symptoms worsened during the follow-up period. Scoliosis was present in nine children (15.5%) preoperatively, though no follow-up data is available on its improvement. The five asymptomatic children remained symptom-free throughout the follow-up period.



1 more than one may apply; Abbreviations: n, number. *p<0.05

Figure 3: Functional outcome. Improvement of symptoms1 (%) in the overall cohort

The subgroup analysis comparing the functional outcomes of healthy children with those of children with disabilities revealed no significant differences.

Postoperative complications were minimal. During the follow-up period, no deteriorations were observed that required reoperation. A superficial wound healing disorder was managed conservatively, one patient experienced temporary paresthesia in the legs.

Discussion

In summary, this descriptive analysis highlights the heterogeneity of the patient population and underscores the general efficacy and safety of de-tethering surgery in improving or stabilizing neurological function and associated symptoms in children with tethered cord syndrome due to fatty or tight filum. Children with TCS related pain had a 80% chance of clinical improvement. The fact that all preoperatively asymptomatic patients remained symptom-free throughout the follow-up period supports the potential value of the surgery as a prophylactic measure. As no patients experienced deterioration, it can be concluded that ultimately all patients benefited from surgery.

The demographic data from our cohort indicate that surgery was performed at a relatively early age, with an average age of 5.5 years. This reflects a clinical focus on intervening promptly after diagnosis to address symptoms and prevent progression. Young patients in our cohort demonstrated favorable outcomes, including improvement in bladder function and pain. These findings align with existing literature emphasizing the benefits of timely surgery in minimizing long-term neurological damage [7, 19, 22-26].

There are a few studies that have specifically examined neurological outcomes following de-tethering of an isolated fatty or tight filum [6, 7, 18, 19, 20, 21, 27-29]. While these studies generally suggest favorable functional outcomes, it is important to note that many exclude children with complex conditions, such as anorectal malformations and urogenital anomalies, limiting the applicability of these findings to a more vulnerable patient population. Additionally, healthy children, particularly those without complex conditions, tend to have better functional outcomes post-surgery [7, 30-33]. This underscores the challenges faced by managing children with more complex conditions, who are often excluded from studies but remain at higher risk for poorer outcomes.

Diagnosing TCS in young children, particularly those with developmental anomalies, remains a significant challenge. Symptoms such as pain and sensory deficits can be difficult to assess in this population, complicating the clinical decision-making process. Additionally, children with developmental anomalies may present with overlapping or atypical symptoms, further obscuring the diagnosis. The availability of objective diagnostic tools, such as neurophysiological monitoring and urodynamic studies, is often limited in young children with complex etiologies, underscoring the need for standardized assessment protocols. Addressing these diagnostic challenges is essential to ensuring timely and effective intervention [8, 18, 21, 22, 24, 25, 27].

Studies suggest that the outcomes of surgery vary depending on follow-up duration and patient comorbidities. Improvement or stabilization in bladder function is commonly reported [6, 19, 34-37], while motor dysfunction shows more variable rates of improvement [6-8]. However, bowel function appears to benefit less consistently, indicating that preexisting bowel dysfunction may limit the impact of the procedure [6-8, 34].

Apart from neurological outcome the re-tethering risk remains a notable concern, especially in children with more complex etiologies. A re-tethering rate of 33.6% was reported in children with complex conditions, such as those with previous anorectal or urogenital surgeries [20], compared to markable lower rates in children without these complications [18, 19, 27-29]. In our cohort, no cases of re-tethering were observed during the 1.8-year follow-up period. This aligns with findings from previous research, where re-tethering rates were low in younger cohorts [18, 19, 27, 28]. Besides etiology, early surgical intervention may reduce the likelihood of symptomatic recurrence, particularly when combined with meticulous surgical techniques and intraoperative monitoring [26, 30, 31, 37, 38]. However, long-term follow-up is essential to fully evaluate recurrence risks [16, 25, 26, 39, 40-44]. The largest published cohort of 99 patients following fatty filum surgery reported a re-tethering rate of 5.1% after 58 months [18], with children who experienced surgical complications, such as CSF leaks, being more prone for re-tethering. In conclusion, while filum transection can lead to improvements in bladder and motor function, the risk of re-tethering is particularly high in children with complex anatomical anomalies. This reinforces the need for careful postoperative monitoring over years, especially in those with anorectal malformations and urogenital anomalies.

This study has several key strengths. It analyzes a diverse cohort of pediatric patients with TCS, including underrepresented subgroups such as those with developmental anomalies. Comprehensive outcome assessment using clinical, neurophysiological, and urodynamic evaluations ensures a thorough analysis of postoperative improvements. The standardized surgical approach, featuring continuous neurophysiological monitoring, resulted in consistent outcomes with low complication rates. Significant improvement in bladder function highlights the procedure's effectiveness, while the sustained symptom-free status of asymptomatic patients underscores its potential prophylactic value. These findings provide valuable insights into the management of TCS in diverse pediatric populations.

Despite these strengths, the study also has limitations. The retrospective design and single-center setting may introduce selection bias and limit the generalizability of the findings. The relatively short follow-up period may underestimate late complications, such as re-tethering, while the lack of a control group limits definitive conclusions regarding the efficacy of surgical intervention. Additionally, the inclusion of a heterogeneous cohort adds complexity to interpreting subgroup-specific outcomes, highlighting the need for future studies with longer follow-ups and larger, multi-center populations. Despite this, the available studies indicate positive surgical outcomes, though there is variability in long-term efficacy and complication rates. National registries could facilitate the standardization of outcome measures, enable a more comprehensive evaluation of post-surgical improvements, and assess quality of life after de-tethering, thereby generating more consistent data and enhancing patient care globally.

Conclusion

This study demonstrates that surgery for TCS in a pediatric population is safe, with minimal complications, and leads to marked improvements in motor function, pain relief, and bowel as well as bladder function, even among symptomatic patients from vulnerable groups with preexisting developmental anomalies. We recommend a proactive approach to surgery in children with TCS, regardless of the severity of their preexisting symptoms.

Declarations

Conflict of interests

The authors declare no competing interests.

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DOI:10.31579/2578-8868/350