Child's Nervous System (2021) 37:3003–3011
https://doi.org/10.1007/s00381-021-05287-5

REVIEW ARTICLE

A critical analysis of surgery for occult tethered cord syndrome

Marissa M. Michael[1] · Andrew L. A. Garton[1] · Claudia M. Kuzan-Fischer[2] · Rafael Uribe-Cardenas[1] · Jeffrey P. Greenfield[1]

Received: 13 January 2021 / Accepted: 6 July 2021 / Published online: 15 July 2021
© The Author(s), under exclusive licence to Springer-Verlag GmbH Germany, part of Springer Nature 2021

Abstract

Introduction Tethered cord syndrome (TCS) is an amalgamation of neurological, urological, orthopedic, and dermatologic signs and symptoms with radiographic evidence of a thickened filum and low-lying conus. Surgical sectioning of the filum and disconnection of any tethering entities such as dermal sinus tracts or lipomas has been shown to improve outcomes. The manifestation of TCS symptoms in the absence of a low-lying conus has been referred to as occult tethered cord syndrome (OTCS) and is much less well reviewed in the literature. To date, there has only been one randomized controlled trial examining the effect of intervention in OTCS; therefore, contemporary data is often elicited from limited cohorts.

Objective To perform a comprehensive literature review of management in OTCS and evaluate treatment response rates to sectioning of the filum terminale.

Results Seventeen papers met inclusion criteria for our review. Sample sizes ranged from 8 to 60 children, and results were mixed, often dependent on study design, definition of typical OTCS symptoms, and follow-up intervals. Symptomatic improvement was observed in > 50% of patients for all but one study; however, the recurrence rates were highly variable.

Conclusion The data regarding the efficacy of surgical treatment in OTCS is mixed and merits more rigorous scientific examination with strict and clear parameters regarding symptomatic operationalization and follow-up time points to monitor for TCS recurrence.

Keywords Tethered cord syndrome · Occult · Review · Pediatric neurosurgery · Filum terminale

Introduction

Tethered cord syndrome (TCS), a condition first described by Hoffman et al. in 1976 [1], is the clinical presentation associated with radiographic findings of a thickened filum terminale and/or a low-lying conus. A wide filum, presence of a lipoma, or fibrosis can cause loss of filum elasticity, leading to the anchoring of the conus medullaris and caudal traction of the spinal cord, predisposing to cord hypoxia and nerve root damage [2]. As a clinical entity, TCS includes neurological symptoms such as back or leg weakness, hyporeflexia, and abnormal gait. Urological symptoms include neurogenic bladder, incontinence, and frequent urinary tract infections. Orthopedic signs can range from lower extremity deformities to scoliosis. Cutaneous abnormalities (such as a sacral dimple) are sometimes observed over the lumbosacral region, occasionally being present over the attachment point of the filum. Any constellation of signs or symptoms from the above categories, or none at all, may be present in individuals with a tethered spinal cord.

The incidence of TCS is historically not well-published, although noted to be increasing steadily, likely due in part to increased awareness, improvements in MRI capabilities in pediatric populations, and incidental diagnoses [3]. Radiographic signs of TCS include (1) the presence of a low-lying conus (typically below L2, although a conus as low as the superior plate of L3 can be considered for children < 6 months of age) [4], (2) a thickened filum terminale (> 2 mm), and (3) lipomatous infiltration within the filum.

Marissa M. Michael and Andrew L. A. Garton contributed equally to this manuscript and are co-first authors.

✉ Andrew L. A. Garton
garton.ala@gmail.com

✉ Jeffrey P. Greenfield
jpgreenf@med.cornell.edu

[1] Department of Neurosurgery, Weill Cornell Medical Center, New York-Presbyterian Hospital, New York City, NY, USA

[2] Department of Neurosurgery, University Children's Hospital Zurich, Zurich, Switzerland

Occult tethered cord syndrome (OTCS), by contrast, is the manifestation of all the above symptoms without clear radiographic evidence of a low-lying conus. There has been difficulty defining this disease and determining whether it should be surgically treated. Otherwise unexplained urinary incontinence played an important role in initially defining this disease, and early evidence from the 1990s suggested that these symptoms could be resolved by detethering the filum in the absence of a low-lying conus [5–7, 9, 10]. The combination of the abovementioned neurological, urological, orthopedic, or dermatological signs and symptoms might be indicative of OTCS; however, the degree of dysfunction that may necessitate or benefit from surgical intervention is still hotly debated [11]. Some studies show that sectioning of the filum terminale can effectively relieve these symptoms [5–7, 10, 12–19]. However, others argue that OTCS is poorly defined, lacks clear in vivo evidence of its pathophysiology, and that, without evidence from a prospective clinical trial with a control group and validated outcome measures, the neurosurgical community cannot be certain of the efficacy of surgical management [20, 21]. We therefore offer here a comprehensive review of both historical and contemporary literature surrounding OTCS management and outcomes with the intention of solidifying a framework upon which subsequent clinical research efforts can be structured.

Literature review

A PubMed search was done for the terms "occult tethered cord syndrome," "filum terminale," and "surgery" or "sectioning." We included all papers that met our search terms then subsequently excluded studies that did not clearly delineate their measurements of symptomatology, although included a wide range of symptom operationalizations (see Table 1, for example, for the variability in outcome measures). We also excluded studies without any outcome data. There was no explicit attempt to exclude papers based on publication date; however, we found that all papers meeting our inclusion criteria were published from 1990 onwards. Of the 51 results, six studies directly undertook a determination of the outcomes of sectioning the filum terminale in pediatric patients with suspected occult tethered cord syndrome. The citation lists of these papers were reviewed to create a comprehensive list of all such studies to date, totaling 17 papers (see Fig. 1).

Results

In 1990, Khoury et al. offered the first investigation of clinical and objective urodynamic outcomes of surgery for a highly selective patient population with what would now be consistent with OTCS. Patients were 5 years of age or older, presented with urinary incontinence, had detrusor hyperreflexia despite conservative management, and had a thickened filum terminale [5]. Thirty-one patients met these criteria, only four of whom had a low-lying conus [5]. The long-term results were promising, with daytime incontinence resolving in 72% (21/29), stool incontinence resolving in 100% (7/7), and back and leg pain resolving in 100% of patients who presented with these symptoms at a mean follow-up of 13.3 months [5]. Urodynamic instability resolved in 59% (16/27) of patients who presented with uninhibited detrusor contractions [5]. Khoury et al. suspected that the progressive deterioration of neurological impairments was associated with increasing traction, eventually culminating in a low-lying conus when presenting too late for effective treatment. They advocated for the development of a sensitive and specific means of diagnosis and proposed a randomized prospective trial of conservative management versus surgery [5].

In 1993, Warder and Oakes reported on their experience with releasing the filum terminale of 12 patients who had a normal level conus (defined by them as at or above the L1–L2 disc space). These patients had combinations of abnormal neurological examinations, cutaneous stigmata of spinal dysraphism, presence of fat in the filum, and bony malformations [19]. They demonstrated post-operative improvement at a mean follow-up of 2.2 years in 75% of patients with bladder dysfunction, 100% of patients with bowel dysfunction, 75% of patients with lower extremity weakness, 67% of patients with lower extremity pain, and 100% of patients with paresthesias [19].

Nazar et al. presented a retrospective chart review of children with a normally positioned conus medullaris (above L2), a filum of a normal diameter (< 2 mm), and severe back and/or leg pain or significant daytime urinary dysfunction, assessing both refractory and conservative management. They assessed 32 patients, 97% (31/32) of whom experienced significant relief of their symptoms at follow-ups ranging from 2 to 48 months, with a mean of 22.4 months [16]. Pain was resolved in 100% of patients for whom that was the presenting symptom, 64% (14/22) developed normal daytime bladder function with 32% (7/22) showing significant (> 50%) improvement with respect to decrease in frequency, urgency and incontinence, and urodynamic function improved in 8 of the 9 patients who were tested post-operatively [16]. All patients who presented with stool incontinence had significant improvement, but in 2 patients who initially showed complete resolution, their improvement went down to 85% at 1 year and 75% at 4 months, respectively [16]. The difference in clinical symptoms between occult and traditional filum terminale syndromes (namely, the low frequency of lumbar cutaneous markings, neurological abnormalities, or skeletal abnormalities in the former) implied that a different, parallel pathophysiological mechanism may exist, and that

Table 1 Compilation of studies examining surgical efficacy in syndromes consistent with OTCS

Authors	Sample size	Urologic results	Fecal results	Pain results	Other results
Khoury et al. [5]	27 (plus 4 patients with a low conus)	Resolution: 72% daytime incontinence , 36% nocturnal enuresis, 59% urodynamic instability	Resolution: 100% stool incontinence	Resolution: 100% back and leg pain	N/A
Warder and Oakes [19]	12	Improvement: 75% bladder dysfunction	Improvement: 100% bowel dysfunction	Improvement: 100% low back pain, 67% lower extremity pain	Improvement: 75% lower extremity weakness, 33% asymmetric hyporeflexia and 25% spasticity, hyperreflexia
Nazar et al. [16]	32	Resolution: 64% daytime bladder dysfunction, 88% urodynamic function	Resolution: 82% stool incontinence	Resolution: 100% pain	Significant (> 50%) relief of symptoms in 97%
Palmer et al. [7]	8	Resolution: 57% daytime incontinence. Improvement: 43% daytime incontinence	Resolution: 100% nocturnal enuresis	N/A	N/A
Selcuki et al. [17]	17	Improvement: 58% urodynamics	N/A	N/A	Subjective improvement of symptoms in 76.4%
Wehby et al. [10]	60	Resolution or improvement: 100% urinary urgency/frequency, 69% recurrent UTI. Resolution: 52% urinary incontinence/retention, 46% nocturia. Improvement: 46% urinary incontinence/retention, 25% nocturia	Resolution: 56% fecal incontinence. Improvement: 41% fecal incontinence	N/A	N/A
Komagata et al. [15]	37	Resolution: 79% bladder-bowel dysfunction	N/A	Resolution: 40% pain Improvement: 58% pain	Muscle weakness in 74%, sensory disturbances in 68% and spinal stiffness in 82%
Nogueira et al. [22]	3	Resolution: 50% voiding dysfunction (optional post-op urodynamics)	N/A	N/A	N/A
Selden [23]	6	Improvement: bladder function 100% (post-op urodynamics). Moderate improvement: 67% voiding dysfunction Mild improvement: 33% voiding dysfunction (subjectively)	N/A	N/A	N/A
Metcalfe et al. [6]	36	Resolution: 42% urinary symptoms Improvement: 72% urinary symptoms, 57% urodynamics	Resolution: 53% bowel incontinence. Improvement: 88% bowel incontinence, 92% constipation	N/A	Resolution: 63% use of medications
Bao et al. [8]	21	N/A	N/A	N/A	Improvement: 93% sphincter dysfunction, 100% sensory neurological deficit, 44% motor neurological deficit

Table 1 (continued)

Authors	Sample size	Urologic results	Fecal results	Pain results	Other results
Steinbok et al. [18]	8	Improvement: 88% urinary incontinence, 57% urodynamics	N/A	N/A	Improvement: 83% non-urological symptoms
Fabiano et al. [13]	22	N/A	N/A	N/A	Subjective improvement: symptoms in 73%. Objective improvement: symptoms in 50%
Fukui et al. [14]	10	Improvement: 90% lower urinary tract symptoms	N/A	N/A	N/A
Cornips et al. [12]	9	Improvement: 100% urological function	N/A	N/A	Improvement: 100% neurological function, 50% orthopedic symptoms (scoliosis)
Kulwin et al. [24]	16	Improvement: 100% urodynamics	Improvement: 100% bowel incontinence	Improvement: 87.5% back or leg pain	Improvement: 100% gait disturbances, 67% scoliosis
Steinbok et al. [18]	21*	Mean change in: urodynamics 0.77 (S) vs. 0.42 (M), bowel bladder dysfunction score − 5.30 (S) vs. − 5.64 (M)	Bowel bladder dysfunction score − 5.30 (S) vs. − 5.64 (M)	N/A	Child impact scale of PEMQOL 19.64 (S) vs. 10.71 (M)

*In both arms

sustained, subradiographical injury might be the cause of the former [16].

A smaller retrospective review of 8 children was published by Palmer et al. in 1999. Their patients had urinary and/or stool incontinence refractory to conservative treatment and spina bifida occulta but had normal urological and neurological examinations and MRIs. Six months after surgery, nocturnal enuresis and stool incontinence had resolved in all children [7]. Daytime urinary incontinence completely resolved in 57% of patients who presented with it, and "markedly improved" in the remainder [7].

In 1998, Selcuki and Coskin investigated 13 patients with urinary incontinence and a normal conus who underwent filum terminale sectioning. Ninety-three percent regained continence in the immediate postoperative period, but almost half became symptomatic again within the first month after surgery [9]. A secondary retrospective review of two groups yielded mixed outcomes as well: 17 patients with TCS without a low-lying conus (all of the patients in the former study were included) were compared to a group of 60 patients with secondary cord tethering who had a low-lying conus. At a mean follow-up of 18 months, 76.4% of the primary tethered cord group had subjective improvement, but only 58.8% had both subjective and objective (by urodynamic data) improvements [17]. The group with a low-lying conus had slightly poorer outcomes, with only 68.5% having subjective improvement at a mean follow-up of 15 months and 28.5% having both subjective and objective improvements [17].

The same group histologically examined 21 filum terminale specimens, providing the first histopathological comparison between OTCS and TCS patients: 5 controls from cadavers (group 1), 8 from patients with normal radiological findings but abnormal urological findings (group 2, OTCS), and 8 from patients with abnormal radiological findings and abnormal urological findings (group 3, TCS). They found that groups 2 and 3 had greater amounts of connective and adipose tissue than group 1, as well as larger capillaries and denser bundles of collagen [26]. This was notable especially in the specimens of group 2 patients in whom these properties, as well as increased amounts of hyalin, were attributed to causing a loss of elasticity in these macroscopically normal fila, thereby leading to transference of a tethering effect to the conus which theoretically could lead to hypoxia and ultimately detrusor irritability and hyperreflexive bladder contractions [26]. This data raises the possibility that TCS and OTCS are manifestations of the same pathophysiologic disease process, in different stages of radiographic clarity.

With one of the largest sample sizes to date, Wehby et al. performed a retrospective review in 2004 of 60 children operated on for "tethered cord syndrome," but with a normally positioned conus. Their study included children with

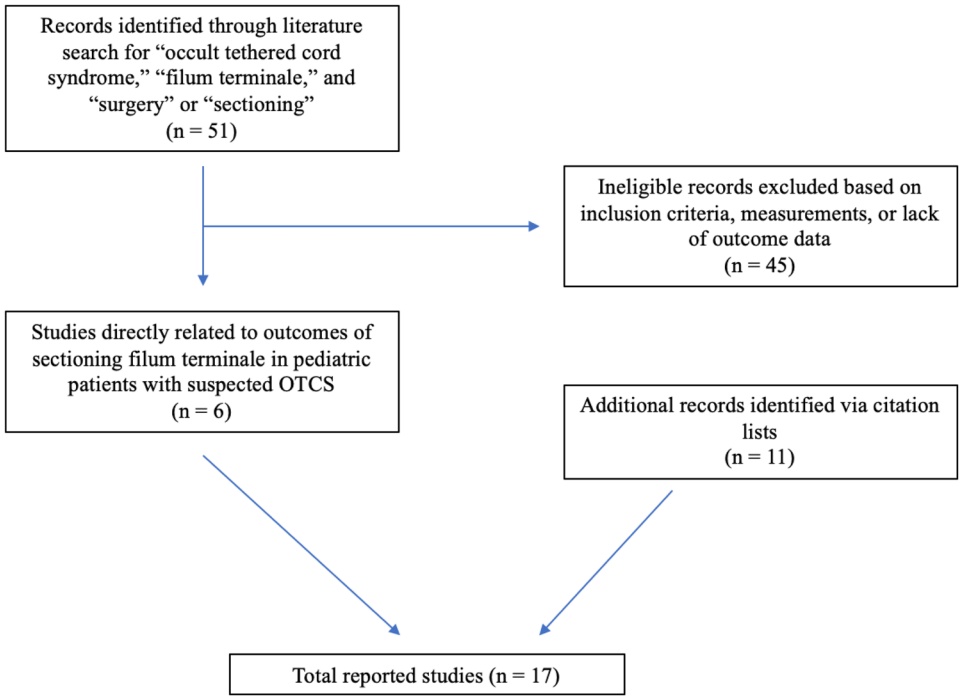

Fig. 1 Schematic of paper selection for review

bladder incontinence despite conservative treatment, spina bifida occulta, and at least two other symptoms (e.g. bowel instability and lower extremity pain or weakness) [10]. They reported that 97% of patients experienced resolution or improvement of their symptoms, with first signs of improvement noted at week 1 after surgery, and almost all experiencing improvement by month 1 [10]. After a mean follow-up of 13.9 months, urinary frequency/urgency resolved or improved in 100% (58/58) of cases, urinary incontinence/retention resolved or improved by more than 50% in 98% (53/54) of cases, fecal incontinence resolved or improved in 97% (31/32) of cases, nocturia resolved or improved in 71% (20/28) of cases, and recurrent UTIs resolved or improved in 69% (11/16) of cases [10]. Notably, Wehby et al. were limited in that their evaluation did not include urodynamic testing or other objective operationalization of incontinence.

In an effort to create a valid and reliable diagnostic assessment for tight filum terminale, Komagata et al. developed a provocation test in which a case was considered positive if low back or leg pain induced by lumbar and neck anterior flexion was relieved by neck extension only. They conducted a review of 37 adult and pediatric patients who had tethered cord syndrome with a conus medullaris above the L1–L2 disc space and at least 4 of the following symptoms: low back pain, non-dermatomal leg pain, bladder-bowel dysfunction, spinal stiffness, positive provocation test [15]. They found that low back or leg pain completely resolved in 40% of patients and improved in an additional 58%. Spinal stiffness improved in 82%, bladder-bowel dysfunction in 79%, muscle weakness in 74%, and sensory disturbances in 68% of patients over a mean follow-up period of 41 months, with a range of 1–8 years [15].

In 2006, Selden et al. found a roughly 87% response rate to surgery, but only 67% improvement when urological symptoms were specifically assessed with urodynamics [11]. The study included 6 patients with bladder dysfunction, some of whom also had pain and bowel incontinence. At a mean follow-up of 16 months, all patients improved clinically, with 4 having moderate improvement and 2 mild [23]. Three of the patients had pre-operative hyperreflexia, which resolved in 1 and improved in 2 [23]. One of the 2 patients with detrusor-sphincter dis-synchrony improved [23]. All 4 patients who underwent postoperative urodynamic assessments had some level of improvement, which was associated with clinical improvement [23].

Shortly after, Metcalfe et al. published a retrospective review of 36 patients who had failed medical management with abnormal urodynamic findings. Nine patients had subtle MRI findings such as slight fat infiltration [6]. The clinical improvement rate of patients with urinary symptoms was 72% with 42% having complete resolution, yet only 57% (16/28) having urodynamic improvement [6]. Bowel incontinence improved in 88% and resolved in 53% within 3 months of surgery, constipation improved in 92% (23/25), and anticholinergic medications were able to be discontinued by 1 year after surgery in 63% of patients [6]. The mean follow-up period was 49 months, and after the 3-month follow-up appointment, no clinical deterioration was noted [6], showing lasting improvement in contrast to prior studies [9].

A review by Bao et al. in 2007 analyzed three different groups of patients: group 1 consisted of 21 children with a normally positioned conus (above L1–L2), 14 of whom did not exhibit a lipoma on imaging [8]. Group 2 patients had a low-lying conus ($n=18$), and patients in group 3 had a low-lying conus with an accompanying lumbosacral lipoma [8]. Thirteen out of 14 of the children in the subgroup with a normal level conus and no lipoma had improvement in their pre-operative symptoms after surgical release of the filum [8]. Seven of the patients in this subgroup had cutaneous stigmata, in contrast to the 100% of patients in groups 2 and 3 who presented with this sign [8]. Microscopic examination of the filum terminale of these patients revealed fat and fibrous tissue infiltration although it was difficult to identify on MRI [8]. Of the patients in group 1, 1 presented with sensory neurological deficit and improved, 4 of the 9 with motor neurological deficit improved, and 13 out or 14 improved in sphincter dysfunction [8]. The mean follow-up for all patients in this study was 2.3 years [8].

Steinbok et al. (2007) conducted a comparative study in which they compared children who underwent surgery for OTCS ($n=8$) to those who did not ($n=7$). Patients were included if they had urinary incontinence refractory to medical management, a conus ending above the lower end plate of L2, no other explanations except for possible OTCS, and were offered surgery [18]. Urinary incontinence improved clinically in 7 out of the 8 surgical patients (88%) at follow-ups that ranged from 1 to 10 years, with a mean of 3.1 years [18]. The subject who did not improve had normal pre-operative urodynamics but other symptoms as well, such as back and lower limb pain, which did resolve after surgery [18]. Urodynamics improved in 4 of the 7 patients who underwent repeat testing for it after surgery [18]. Non-urological symptoms improved in 5 of the 6 surgical children who presented with them, and the 1 who failed to improve had normal urodynamic findings pre-operatively [18]. By contrast, urinary incontinence only improved in 2 out of the 7 non-surgical patients at a mean follow-up of 3.3 years, and none of them improved with respect to their other symptoms [18].

In an attempt to identify pre-operative predictors of successful surgery, Fabiano et al. performed a retrospective review of 22 patients with a conus medullaris above the L3 vertebral body, filum less than 2 mm thick, and clinical symptoms suggestive of OTCS, divided into dermatologic, urologic, orthopedic, and neurologic categories. They found that the patients who presented with clinical symptoms in more than 1 of these 4 groups ($n=17$) were more likely to experience improvement after surgical untethering than those who only presented with symptoms from only 1 of the aforementioned groups (88% and 20%, $p=0.009$) [13]. They also found a correlation between the number of categories of symptoms patients presented with and their likelihood of improving after surgery ($X^2=9.40$, $p=0.024$) [13]. However, they did not find a significant correlation between postoperative outcomes and conus level ($p=0.65$), presence of minimal fatty infiltration in the filum ($p=0.56$), or presence of a syrinx ($p=0.36$) [13]. Overall, at a mean follow-up of 15.9 months, 16 of the 22 patients (73%) had subjective improvement, and 11 (50%) had objective improvement, as measured by neurological exam, bladder control, or urodynamic testing, as appropriate [13].

Cornips et al. attempted to compare outcomes of untethering surgery between patients who had normal and abnormal filum terminale radiographic findings. They found that improvements in neurologic and urologic functions were greater in the group with OTCS than that of the children in the TCS group. Of the 9 children in the former, all improved in neurological function, 100% of those who presented with urological dysfunction improved, and 1 of 2 patients with scoliosis improved, with it being unchanged in the other 1. Of the 2 children with a normal CM and an abnormal filum, 1 improved in their neurological symptoms, and 1 was unchanged [12]. Overall across both groups, symptoms improved subjectively in 75–79% and objectively in 38–68% of patients. Neurological symptoms improved or stabilized in 100% (25/25), urologic symptoms in 1/17, and orthopedic symptoms in 7/8 of the affected children [12].

Subsequently, Kulwin et al. conducted another study that examined a comorbidity of tethered cord syndrome. They performed a retrospective review of 16 patients operated on for TCS with what they defined as a normal conus level, above the rostral border of the L2–L3 disc space, and syringomyelia [24]. Two of the 16 patients had a fatty filum on T1 and 12 of them presented with abnormal urodynamics. The results were again promising, with all 11 who had post-operative urodynamics assessments improving both on formal testing and subjectively at a mean follow-up of 17 months. Seven of the 8 children who presented with back or leg pain had subjective improvement, along with 2 of the 2 with gait disturbances, and the 1 who presented with bowel incontinence. Two of the 3 with scoliosis either stabilized or improved, with the remaining 1 progressing [24].

However, surgical release of the filum was only minimally effective at improving syringomyelia in these patients. Only 4 patients (25%) had improvement in their syrinx at a mean follow-up of 14.5 months [24]. A fatty filum appeared to be predictive of an improved syrinx, as both of the 2 patients who presented with one had improvement of their syrinx (100%), while only 2 of the remaining 14 (14%) did ($p=0.05$). Additionally, all of the syrinxes that improved were between T5 and the conus, with 3 of the 4 below L1-2 [24].

In 2016, Steinbok et al. reported the first randomized, controlled pilot study which compared medical treatment to surgical sectioning of the filum terminale [25]. They randomized patients refractory to conservative management for

at least 1 year who had a normal conus position (above L2) and abnormal urodynamic findings. They excluded patients who had bladder outlet obstruction, an alternative diagnosis associated with neuropathic bladder, anorectal malformations, an alternative urological diagnosis, an atonic bladder, progressive lower extremity motor/sensory deficits, spinal dysraphism, or other spinal cord abnormalities, but accepted those with lumbar bifid spinal lamina or with lumbar cutaneous stigmata. Unfortunately, the target enrollment was not attained (goal 44 patients over 2 years at 3 different sites), in that 21 patients were randomized in 8 years. No significant differences in improvements between groups were found. However, there was a trend of a greater difference in urodynamics scores and scores on an enuresis quality of life questionnaire in the surgical arm than the medical arm, although the reverse was true for a bladder-bowel dysfunction assessment [25]. There were several possible reasons for their absence of findings, including small sample size, rigid exclusion criteria, duration and severity of symptoms, and the sensitivity of their outcomes assessments to interventions [25].

Discussion

At least a dozen retrospective studies have demonstrated some level of utility in untethering surgery in patients who present with a conus of a normal level but any combination of a wide array of urological, neurological, orthopedic, and dermatologic symptoms. In almost all of these studies, subjective resolution or significant improvement of most symptoms was at least 70%, and in some even reached 100% across all symptoms. Counterintuitively, a few report greater improvement in patients with a normal level conus than in those with a low conus [12, 26].

Khoury et al. suspected that tethered cord syndrome was a progressive disease which eventually resulted in a low-lying conus, and Nazar et al. noted that, in some of the patients whose symptoms did not improve, pre-operative pain had been present for the longest time (4 + years) and in those that it resolved in completely, their symptoms were present for less than 3 years or were comparatively less severe preoperatively. Therefore, the fact that many of the patients in Steinbok's study had a greater duration of symptoms (5 years) is a reasonable explanation for a higher proportion of refractory patients reducing the overall sensitivity of the study to detect group differences.

In current practice, if offered, neurosurgical management is typically a single-level laminectomy, durotomy, and selective sectioning of the filum terminale. Children who undergo this procedure are 5.8 years old on average and are a majority white (78.6%) and share an equal gender distribution [27, 28]. An international quality improvement database of 3682 pediatric patients found this procedure to have a complication rate of 8.1% (superficial wound infection: 5%, UTI: 2%, deep wound disruption: 1%) and a reoperation rate of 2.7% [27]. Smaller retrospective chart reviews reported post-operative complication rates between 1.7 and 13.2% [29–32] and re-operation rates between 2.7 and 8.6% [29–33]. One study found retethering was related to post-operative unchanged conus level, spinal cord thickness, and syrinx, which was thought to be representative of lower elasticity of the spinal cord [29] and another to a higher complication rate after initial surgery [30]. Mindful patient selection regarding surgical intervention in OTCS is important to maintain these complication rates as low as possible, given the nature of symptoms patients present with.

The rate of symptomatic recurrences reported in a few of the studies in Table 1 is worth noting. Given the variability in diagnostic criteria for OTCS across these studies, comparing recurrence rates is difficult (e.g., different patients meet different criteria for OTCS for different reasons). A patient who undergoes sectioning of a fatty filum for leg pain may experience symptomatic relief but continue to have enuresis for several months afterward; by some criteria, this would be recurrence, while by others, it would be successful remission. The best estimation of a natural history of OTCS comes from Steinbok et al.'s 2016 pilot RCT; however, the sample size in the medical arm was 11, and as such, conjectures are limited. The natural history of OTCS warrants further study. For example, on average, enuresis regardless of etiology tends to improve throughout childhood without surgery, and so, therefore, it is imperative to understand the efficacy of surgery relative to observation when considering management of OTCS.

Enuresis is inherently highly variable between families, cultures, and practices. Therefore, every effort should be made to create objective measures when deciding if patients could make suitable operative candidates or not: it would be helpful, for example, to derive rates and incidences of urinary and bowel incontinence from patient voiding diaries and cross-reference those with objective urodynamic assessments when possible.

A need remains for more objective evidence regarding characterization of the clinical manifestations of OTCS and assessment of the efficacy of surgical intervention versus observation for expediting symptomatic relief. Decision-making would be aided by a prospective randomized clinical trial designed to identify children who are most likely to benefit from surgery. For example, as Fabiano et al. found that patients who presented with symptoms in more than 1 of the 4 main categories (urological, neurological, orthopedic, and dermatological) were significantly more likely to experience improvement than those who only presented with symptoms from 1 of these groups ($p = 0.009$), and since Metcalfe et al. noted a similar trend although not significant, it stands to

reason that children with multiple symptoms across these categorizations could benefit the most from surgery and should be closely examined.

There are many arguments against surgical intervention. The definition of the syndrome itself is unclear owing to its wide array of possible symptoms and the common nature of the primary symptoms (of incontinence and back pain) which are therefore not highly specific to the syndrome [20]. Furthermore, the natural history of OTCS is not well understood: in a retrospective review of OTCS patients with anorectal malformations, as few as 18% of patients actually deteriorated [34], while the risks of complications from prophylactic surgery are not insignificant [20]. Furthermore, there is absent experimental support for the ischemic hypothesis: no histological studies have demonstrated caudal ischemia or quantitative proof for tension in the filum [20].

Conclusion

Understanding the optimal management of TCS has not itself been without controversy. The establishment of consensus guidelines for the management of symptomatic presentations in the absence of a low-lying conus has been challenging for the neurosurgical community. While there are several historical cohorts that implicate surgical management as a viable, low-risk alternative to observation in these children, these studies lack clear follow-up paradigms, internal and external controls, and reliable, consistent symptomatic characterizations. We hope that this review will contribute to an ongoing understanding of this condition as well as guide future research efforts and clinical trials [35].

Declarations

Conflict of interests The authors declare no competing interests.

References

1. Hoffman HJ, Hendrick B, Humphreys RP (1976) The tethered spinal cord: its protean manifestations, diagnosis and surgical correction. Pediatr Neurosurg 2:145–155
2. Filippidis AS, Kalani MY, Theodore N, Rekate HL (2010) Spinal cord traction, vascular compromise, hypoxia, and metabolic derangements in the pathophysiology of tethered cord syndrome. Neurosurg Focus 29:E9
3. Bui CJ, Tubbs RS, Oakes WJ (2007) Tethered cord syndrome in children: a review. Neurosurg Focus 23:E2
4. Asil K, Yaldiz M (2018) Conus medullaris levels on ultrasonography in term newborns: normal levels and dermatological findings. Journal of Korean Neurosurgical Society 61:731–736
5. Khoury AE, Hendrick EB, McLorie GA, Kulkarni A, Churchill BM (1990) Occult spinal dysraphism: clinical and urodynamic outcome after division of the filum terminale. J Urol 144:426–428
6. Metcalfe PD, Luerssen TG, King SJ, Kaefer M, Meldrum KK, Cain MP et al (2006) Treatment of the occult tethered spinal cord for neuropathic bladder: results of sectioning the filum terminale. J Urol 176:1826–1830
7. Palmer JS, Maizels M, Grant JA, Richards I, Kaplan WE: Transection of filum terminale remits urinary and stool continence in children with neuropathic bladder and spina bifida occulta.(Abstract). Pediatrics 104:809, 1999
8. 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
9. 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
10. Wehby MC (2004) O'hollaren PS, Abtin K, Hume JL, Richards BJ: Occult tight filum terminale syndrome: results of surgical untethering. Pediatr Neurosurg 40:51–57
11. Selden NR (2006) Occult tethered cord syndrome: the case for surgery. J Neurosurg 104:302–304
12. Cornips EMJ, Vereijken IMP, Beuls EAM, Weber JW, Soudant DLHM, van Rhijn LW et al (2012) Clinical characteristics and surgical outcome in 25 cases of childhood tight filum syndrome. Eur J Paediatr Neurol 16:103–117
13. Fabiano AJ, Khan MF, Rozzelle CJ, Li V (2009) Preoperative predictors for improvement after surgical untethering in occult tight filum terminale syndrome. Pediatr Neurosurg 45:256–261
14. Fukui J, Ohotsuka K, Asagai Y (2011) Improved symptoms and lifestyle more than 20 years after untethering surgery for primary tethered cord syndrome. Neurourol Urodyn 30:1333
15. Komagata M, Endo K, Nishiyama M, Ikegami H, Imakiire A (2004) Management of tight filum terminale. Minim Invasive Neurosurg 47:49–53
16. Nazar GB, Casale AJ, Roberts G, Linden D (1995) Occult filum terminale syndrome. Pediatr Neurosurg 23:228–235
17. Selçuki M, Ünlü A, Çağlar Uğur H, Soygür T, Arıkan N, Selçuki D (2000) Patients with urinary incontinence often benefit from surgical detethering of tight filum terminale. Childs Nerv Syst 16:150–154
18. Steinbok EP, Kariyattil ER, Macneily EA (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
19. Warder ED, Oakes JW (1993) Tethered cord syndrome and the conus in a normal position. Neurosurgery 33:374–378
20. Drake JM (2006) Occult tethered cord syndrome: not an indication for surgery. J Neurosurg 104:305–308
21. Steinbok P, MacNeily AE (2007) Section of the terminal filum for occult tethered cord syndrome: toward a scientific answer. Neurosurg Focus 23:1–4
22. Nogueira M, Greenfield SP, Wan J, Santana A, Li V (2004) Tethered cord in children: A clinical classification with urodynamic correlation. J Uro 172:1677-1680
23. Selden NR, Nixon RR, Skoog SR, Lashley DB (2006) Minimal tethered cord syndrome associated with thickening of the terminal filum. J Neurosurg 105:214
24. Kulwin CG, Patel NB, Ackerman LL, Smith JL, Boaz JC, Fulkerson DH (2013) Radiographic and clinical outcome of syringomyelia in patients treated for tethered cord syndrome without other significant imaging abnormalities. J Neurosurg Pediatr 11:307
25. Steinbok P, Macneily AE, Hengel AR, Afshar K, Landgraf JM, Hader W et al (2016) Filum section for urinary incontinence in children with occult tethered cord syndrome: a randomized, controlled pilot study. J Urol 195:1183–1188
26. Selçuki M, Vatansever S, Inan S, Erdemli E, Bağdatoğlu C, Polat A (2003) Is a filum terminale with a normal appearance really normal? Childs Nerv Syst 19:3–10

27. Bhimani AD, Selner AN, Patel JB, Hobbs JG, Esfahani DR, Behbahani M et al (2019) Pediatric tethered cord release: an epidemiological and postoperative complication analysis. Journal of Spine Surgery 5:337–350
28. Shweikeh F, Al-Khouja L, Nuño M, Johnson JP, Drazin D, Adamo MA (2015) Disparities in clinical and economic outcomes in children and adolescents following surgery for tethered cord syndrome in the United States. J Neurosurg 15:427–433
29. Finger T, Schaumann A, Grillet F, Schulz M, Thomale U-W (2020) Retethering after transection of a tight filum terminale, postoperative MRI may help to identify patients at risk. Childs Nerv Syst 36:1499–1506
30. Ostling LR, Bierbrauer KS, Kuntz C (2012) Outcome, reoperation, and complications in 99 consecutive children operated for tight or fatty filum. World neurosurgery 77:187–191
31. Vassilyadi M, Tataryn Z, Merziotis M (2013) Retethering in children after sectioning of the filum terminale. Pediatr Neurosurg 48:335–341
32. Yong RL, Habrock-Bach T, Vaughan M, Kestle JR, Steinbok P (2011) Symptomatic retethering of the spinal cord after section of a tight filum terminale. Neurosurgery 68:1594–1602
33. Ogiwara H, Lyszczarz A, Alden TD, Bowman RM, McLone DG, Tomita T (2011) Retethering of transected fatty filum terminales clinical article. J Neurosurg Pediatr 7:42–46
34. Tuuha SE, Aziz D, Drake J, Wales P, Kim PCW (2004) Is surgery necessary for asymptomatic tethered cord in anorectal malformation patients? J Pediatr Surg 39:773–777
35. Tu A, Steinbok P (2013) Occult tethered cord syndrome: a review. Childs Nerv Syst 29:1635–1640

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.