Childs Nerv Syst (2013) 29:1635–1640
DOI 10.1007/s00381-013-2129-1

SPECIAL ANNUAL ISSUE

Occult tethered cord syndrome: a review

Albert Tu · Paul Steinbok

Received: 22 April 2013 / Accepted: 23 April 2013

Abstract
Background Tethered cord syndrome is a well-defined condition, the management of which is fairly uniform. In contrast, occult tethered cord syndrome is a recently defined entity, where the management is still controversial. The pathophysiology is unclear and may be conceptually incongruent with current understanding of typical tethered cord syndrome. Presentation, investigation, and management of this condition are reviewed, and current understanding is presented.
Purpose The aim of this study is to review the presentation, pathophysiology, investigation, and management of occult tethered cord syndrome.
Methods Literature review.
Results Patients with occult tethered cord syndrome presents predominantly with urologic symptoms. Adult and pediatric patients vary slightly in their presentation with a higher incidence of pain in the former and incontinence in the latter. Operative management for these patients is associated with consistent improvement in urologic function in particular, although surgery is also associated with risk of worsening symptoms. The natural history of untreated patients is unknown.
Conclusions As occult tethered cord syndrome becomes increasingly recognized, it is important to be aware of the potential benefits of operative intervention for appropriately selected patients. Given that the natural history of this entity remains unknown, a clinical trial is currently underway that may assist in defining the role for operative management in treating this condition.

Keywords Occult tethered cord syndrome · Normal conus · Tight filum terminale · Urinary dysfunction · Natural history · Review

Introduction

The clinical and radiographic constellation of tethered cord syndrome (TCS) secondary to a tight filum terminale is well-documented and accepted [8]. The characteristic features of this condition include neurologic, orthopedic, and/or urologic symptoms in the presence of an abnormally low conus [8, 17]. More recently, the concept of an occult tethered cord syndrome (OTCS) has been proposed [29]. In this condition, symptoms consistent with TCS are present and are thought to be related to tethering of the spinal cord by the filum, but the conus is in a normal position. A previous survey by Steinbok et al. (2007) [23] demonstrated that management of patients with classic TCS features is fairly uniform among pediatric neurosurgeons. However, the management and even the existence of OTCS is controversial. The current understanding of OTCS is reviewed.

Presentation

Khoury et al. (1990) first described sectioning of the filum to untether the spinal cord in patients with a conus at the normal level [7]. They identified 31 patients with predominantly urologic symptoms suggestive of TCS in the presence of an occult spinal bony dysraphism but a normally

A. Tu · P. Steinbok
Division of Neurosurgery, University of British Columbia, Vancouver, BC, Canada

P. Steinbok
Division of Neurosurgery, Department of Surgery, British Columbia Children's Hospital, Vancouver, BC, Canada

P. Steinbok (✉)
Division of Neurosurgery, B.C. Children's Hospital and UBC, 4480 Oak St., Room K3-159, Vancouver, BC, Canada V6H 3V4
e-mail: psteinbok@cw.bc.ca

positioned conus. After section of the filum, a majority experienced recovery or improved control of their urinary function. Warder (1993) later described 13 patients with a similar clinical presentation in the absence of obvious radiographic abnormality but with cutaneous findings suggestive of occult spinal dysraphism [29]. This group formed the basis of what was later termed an "occult tethered cord syndrome". Warder et al. (1993) found that up 18 % of TCS patients actually had a conus at the normal level, and other surgical series have found similar incidences ranging from 14–28 % [5, 28, 29].

Like typical TCS, the presentation of OTCS is variable with multiple nonspecific symptoms. Broadly, symptoms may be classified into four main categories: neurologic (i.e., motor or sensory dysfunction), urologic, neurocutaneous, and neuro-orthopedic [19]. The most common in OTCS is urinary dysfunction, which is seen in 68–100 % of patients [4, 9, 13, 16, 21, 24, 31] and can have considerable deleterious effects on quality of life [1]. In pediatric series, patients present with a combination of urinary frequency and incontinence (both day and night). Secondary incontinence is much more prevalent than primary incontinence at a ratio of 2:1–6.7:1 [13, 31]. In patients unable to verbalize their urinary changes, frequent urinary tract infections may further suggest a dysfunctional bladder and is seen in as many as 50 % of OTCS sufferers [20]. On the other hand, adult patients tend to present with urinary frequency but are less likely to be incontinent [9]. As many 100 % of adult patients will also present with nondermatomal back and or lower extremity pain and may also be the only presenting symptom [9, 33]. Komagata et al. (2004) found that this pain could be distinguished from other spinal pathologies by its exacerbation with flexion posture but alleviation with neck extension [9]. In addition, a significant proportion of patients will further present with bowel complaints including both constipation and encopresis, which has been reported in as many as 58 % [13]. While up to 51 % of healthy people have at least transient urinary dysfunction [32], patients with OTCS not infrequently have additional neurologic findings. Metcalfe et al. (2006) observed that in patients referred for isolated urinary dysfunction, 19 % also had abnormal spinal reflexes on detailed examination [13]. Even in the presence of a normal neurologic exam, Selcuki et al. (2000) reported that 24 % of their patients demonstrated altered somatosensory evoked potentials (SSEPS) [20]. Some authors have argued that OTCS always presents with more than isolated urinary dysfunction, although these studies have been from retrospective reviews of surgically treated patients [30]. Fabiano et al. (2009) found that 22.7 % of patients had a single system dysfunction (i.e., isolated urologic or neurologic dysfunction) [4]. In addition, Yamada et al. (2007) proposed that the pathophysiology of cord tethering could facilitate a presentation of isolated incontinence alone [33].

Anatomy and embryology

TCS may be considered a developmental anomaly of the spinal cord. The CNS undergoes three key steps to form the spinal cord: neurulation, canalization of the tail bud, and differentiation. At approximately 20 days gestation, the neural plate folds onto itself and forms a neural tube. Multiple fusion points develop and extend in rostral and caudal directions. When the fusion reaches the tail of the neural placode, a terminal body known as the caudal cell mass is stimulated to form the tail bud. Although this process is not well understood in humans, it is believed the caudal cell mass undergoes canalization and regression until all that is left is the medulla spinalis below T12 and a distal filum terminale. Abnormalities in canalization of the caudal cell mass result in a pathologic filum terminale and the potential for future tethering [6].

Pathophysiology of tethering by the filum

In theory, the basic underlying problem in TCS secondary to the filum terminale is abnormal tension on the conus by the filum. The spinal cord and conus are secured to the surrounding spinal canal down to the level of T12 by the dentate ligaments, an extension of the overlying pia. Caudally, the conus is fixed to the canal by the filum terminale, a fibro elastic structure that continues as far down as S2 intradurally and then continues extradurally. The normal filum is less than 2 mm in diameter, and the normal location of the conus tip is above the lower part of the body of L2 [11]. It has been suggested that the primary role of the filum terminale is to secure and stabilize the conus during movements, which can significantly lengthen or shorten the spinal canal. Some studies have shown as much as a 7 % increase in length of the canal during flexion [25]. When applied to neural tissue, this stretch can result in metabolic derangements equivalent to ischemic injury [33]. It serves to reason that the filum is the structure responsible for accommodating these length changes. The failure of the filum to facilitate spinal cord movement is thought to be the pathologic mechanism that leads to symptoms of a tethered cord [33].

The spinal cord relies on oxidative metabolism to produce ATP and decreased elasticity of an abnormal filum transfers pathologic stretch to the distal cord and conus. The resultant reductions in oxidative metabolism mirror those seen in hypoxemia [33]. Mild to moderate stretch in experimental models results in transient reductions in metabolism, whereas more severe stretch causes persistent metabolic derangements that may not recover. The mechanism for these changes is not entirely clear, although it is thought to be related to manual deformation of cell and mitochondrial membranes, thereby causing a local energy

deficiency [33]. In addition, local blood flow changes have also been observed in direct response to sectioning of the filum suggesting a coexistent relative oligemia [33]. The fluctuating course that some patients report may be related to intrinsic plasticity of neuronal and glial cells that can accommodate some mechanical change over time [33].

It is easy to understand how a thickened filum may exert tension on the conus and therefore cause the radiographic and clinical features of TCS. It is more difficult to explain symptomatic tethering of a cord ending in the normal location, especially in cases where the filum appears normal. One possibility that has been explored is that the microstructure of the filum is abnormal, and hence the filum is functionally less elastic than normal. Histologically, the intradural rostral third of the filum contains structures similar to the spinal cord including a central canal and neural stem cells [11]. The caudal two thirds, in comparison, is composed predominantly of fibrous tissue with a mixture of elastin, elaunin, reticulin, and collagen. It is believed that the compliance of the filum is determined by the ratio of elastic fibers to collagen and the integrity of the reticulin framework. When comparing filum resected from TCS patients and normal controls, there is a significant reduction of reticulin and changes in the ratio of elastic /collagen proteins that consequently decrease the elastic properties of the filum [11]. Furthermore, fila from TCS patients demonstrate adipocyte invasion, fibrosis, and loss of the typical meningothelial cell architecture [26]. These changes are seen in macroscopically "thickened" filum, as well as in grossly normal appearing filum of patients with TCS, although to varying degrees [26].

Adult onset TCS is conceptually counter intuitive, since growth of the spine is no longer occurring but may be explained by changes to the filum in addition to environmental and age-related factors. These includes: (1) progressive fibrosis of the filum that leads to eventual loss of elasticity, (2) growth spurts in adolescence and early adulthood that increase tension on the filum, (3) increases in physical activity with strain on neurologic structures, and (4) osteoarthritic spinal canal stenosis with further restriction of movement of the spinal cord and filum, thereby accentuating vertical tension on the cord [33]. Whether the filum is congenitally predisposed to tethering or acquires changes that cause progressive tethering is not resolved, although may explain the different presentations seen in adult and pediatric OTCS.

Investigations

Lumbosacral imaging should be obtained in any patient investigated for suspected tethering of the spinal cord. Although MR is considered the definitive imaging modality, Rohrschneider et al. (1996) demonstrated that gray-scale ultrasound is comparable to MR in sensitivity up to 6 months of age or before the posterior elements calcify and occlude the acoustic window. While a low lying conus or spinal dysraphism is a reliable predictor for TCS, the radiographic evaluation of OTCS is more challenging. Multiple groups [15, 22] have attempted to improve the diagnostic sensitivity and specificity, and several MR sequences have been presented. Supine MR, in the absence of any other features of spinal dysraphism or abnormally positioned conus has not been found to be particularly sensitive for OTCS but does a have reasonable negative predictive value reaching 90 % in some series [22]. Prone MR has been advocated as a useful tool to detect a posteriorly-deflected filum relative to the dorsal nerve roots [15]. In normal patients, the filum is covered by the posterior roots and attaches anteriorly in the sacral canal. TCS patients in comparison have a posterior attachment of the filum, often at the most lordotic position of the spine (i.e., L4/5). This observation has been confirmed by intraoperative endoscope prior to opening of the arachnoid and potential displacement of the structures with CSF egress [33]. While some authors [15] advocate strongly for use of prone MRI to try and identify the posterior location of the tethering filum, the sensitivity of this tool has been reported to be as low as 62 % [22], with an interreader concordance of only 69 %. Furthermore, the additional anesthetic time/scanner time associated with prone MR tempers its wide acceptance. Cine MR similarly suffers from reported sensitivities ranging from 50–67 % [22], with accuracies only as high as 62 %. To date, while imaging studies are essential to the evaluation of the patient with TCS, their utility in diagnosing OTCS remains limited.

SSEPs have also been used in the investigation of patients with OTCS [2]. Longitudinal studies have verified the utility of SSEPs in detecting delayed retethering during follow-up of previously treated patients [20]. Their use, however, in OTCS is limited, as the sensitivity is poorer than in patients with typical TCS [20]. As such, the role of SSEPs in many centers has been in the evaluation of postoperative function rather than patient selection for surgery [20].

In comparison, urodynamic studies (UDS) are felt to be strong predictors for patients with a tethered cord [10]. In a study by Lavallee et al. (2013), 85 % of patients with abnormal urodynamics had an abnormal MRI, while 40 % subsequently underwent a tethered cord release [10]. Meyrat et al. (2003) introduced an objective UDS score based on parameters of bladder volume, compliance, detrusor activity, vesicosphincteric dysynergy, each graded on a scale of 1–5. Scores of 0–4 were considered normal, 5–6 suggestive of possible disorder, and >6 definitively abnormal [14]. In addition, a change of −2 on serial examinations was considered objective evidence of neurourologic deterioration. The objectivity and accessibility of this test makes it appealing

Table 1 Details of published series of OTCS

Author	Duration of follow up (months)	Number of patients in study	Number with urologic dysfunction	Number with improved urologic symptoms post op	Number with worse symptoms post op
Khoury et al. (1990)	Mean 13.3	31	31	22	0
Warder and Oakes (1993)	Range 6–72	13	13	4	0
Nazar et al. (1995)	Range 2–48	32	32	31	0
Selcuki et al. (2000)	Mean 18	17	17	3	4
Selcuki et al. (2003)	Range 3–84, mean 31.7	8	8	7	1
Komagata et al. (2004)	Not reported	37	33	26	0
Wehby et al. (2004)	Mean 13.9	60	60	58	0
Metcalfe et al. (2006)	Mean 49	36	36	26	0
Selden et al. (2006)	Range 3–30, mean 15.6	6	6	6	0
Steinbok et al. (2007)	Range 12–120, mean 37.2	8	8	7	1
Fabiano et al. (2009)	Mean 16	22	22	16	2
Fukui et al. (2011)	Mean of over 240 months	10	10	9	1
Cornips et al. (2012)	Mean 30.4	9	5	5	0

for the evaluation of patients with urologic symptoms suspicious for TCS and should be included in the primary workup. A variation of this scoring system was developed by MacNeily et al. (2007), specifically for assessing patients with potential OTCS [12].

Other tests for the diagnosis of OTCS have been proposed, but most are considered too invasive (i.e., direct stretch test of filum) or impractical to utilize on a screening basis (i.e., photospectrometry) [33]. At present, the primary modalities of identifying patients with OTCS remains history, clinical examination, and urodynamic studies to delineate the etiology of urinary dysfunction. Imaging is essential in the evaluation of these patients, but the absence of a radiographic abnormality does not rule out the presence of OTCS.

Management and outcome

The natural history of TCS and OTCS remains controversial. Some authors have suggested that neurologic progression and deterioration occurs in over 50 % of patients with clinical TCS [27], while others have suggested a much lower rate of progression [3]. In patients with urologic dysfunction, spontaneous improvements have been reported in 16 % per year [30]. Steinbok et al. (2007) retrospectively compared OTCS patients undergoing surgery versus conservative treatment, and found that while 88 % of patients improved with surgery, 29 % of patients managed conservatively also had improvement in their symptoms [24]. Furthermore, while no patient worsened without surgery, 12.5 % of patients deteriorated after operative intervention. These results reinforce that the role for intervention must be tempered against the potentially benign course of most patients [3]. In a separate study, a survey of AANS pediatric section neurosurgeons revealed that 85 % would detether a patient with clinical TCS and a radiographic evidence of a low-lying conus and fatty filum [23]. However, only 67 % would offer detethering if there was clinical evidence of TCS and only a fatty filum, and there was significant disagreement regarding the management of patients with clinical findings suggestive of TCS but with normal imaging.

Surgical management for OTCS has been well-described in the literature with a variety of approaches and techniques [13, 18, 20, 31]; the key tenet being to disconnect the filum. None have proven to be superior. To date, 13 publications exist describing the outcomes of surgery for OTCS. These studies are summarized in Table 1. All were retrospective studies with 12 publications focusing on pediatric patients and 1 study reviewing outcomes in adult patients [9]. Overall, 97.2 % of 289 patients presented with urologic dysfunction and 78.3 % improved after surgery. Follow up on average was 46.5 months, when all studies were included; however, one

Table 2 Summary of published series of OTCS

Average follow up	46.5 months
Corrected average follow up*	25.01 months
Total number of patients	289
Total number with urologic dysfunction pre op	281
Total number improved subjectively	220
Total number with worse urologic symptoms	9
Percent presenting with urologic dysfunction	97.23 %
Percent with improved urologic symptoms	78.30 %
Percent with worse urologic symptoms	3.28 %

*With removal of study by Fukui et al. (2011)

study [9] reported outcomes in patients with over 20 years of follow up; and when this study was excluded, the average follow up was 23.5 months. Five studies also reported on deterioration after surgery for a total of nine patients or 3.28 % (Table 2). This figure includes only those with development of new or worsening of preexistent symptom(s). Studies also reported on the success of detethering for other symptoms, including bowel abnormalities and pain. Success of treatment of these symptoms varied from 88 % for bowel dysfunction [13] to 98 % for pain [9, 33]. Although these results are difficult to summarize, as reporting methods and criteria vary for each study. While these findings suggest good initial outcomes, it is unclear whether improvements are durable and persistent. The incidence of retethering has also not been established in this population. Current literature suggests that patients with a simple filum snip for TCS have a 3–8 % risk of delayed retethering; whether OTCS patients face the same probability is not yet known.

Conclusion

OTCS is a relatively recently identified entity that is still poorly understood. While the most common symptom remains urologic dysfunction, patients may also experience pain and neurologic deficits. By definition, diagnosis is dependent on clinical examination supplemented by urodynamic or functional studies. The absence of objective radiographic abnormalities makes the decision about a detethering operation difficult. In uncontrolled case series, intervention in appropriately selected patients can generate substantial meaningful improvement. The longevity of these benefits in the long term remains to be determined. Because management of these patients remains controversial, a randomized controlled trial is underway to compare outcomes after section of the filum with best medical management in children over the age of 5 years and possible OTCS [24], but accrual of patients has been slow, and the study is not expected to be completed for another 2 years.

Disclosure The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

References

1. Avery JC, Stocks NP, Duggan P, Braunack-Mayer AJ, Taylor AW, Goldney RD, Maclennan AH (2013) Identifying the quality of life effects of urinary incontinence with depression in an Australian population. BMC Urol 13:11
2. Boor R, Schwarz M, Reitter B, Voth D (1993) Tethered cord after spina bifida aperta: a longitudinal study of somatosensory evoked potentials. Childs Nerv Syst 9:328–330
3. Drake JM (2006) Occult tethered cord syndrome: not an indication for surgery. J Neurosurg 104:305–308
4. 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
5. Hendrick EB, Hoffman HJ, Humphreys RP (1983) The tethered spinal cord. Clin Neurosurg 30:457–463
6. Hertzler DA 2nd, DePowell JJ, Stevenson CB, Mangano FT (2010) Tethered cord syndrome: a review of the literature from embryology to adult presentation. Neurosurg Focus 29:E1
7. 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, discussion 428–429, 443–424
8. Klekamp J (2011) Tethered cord syndrome in adults. J Neurosurg Spine 15(3):258–270
9. Komagata M, Endo K, Nishiyama M, Ikegami H, Imakiire A (2004) Management of tight filum terminale. Minim Invasive Neurosurg 47:49–53
10. Lavallee LT, Leonard MP, Dubois C, Guerra LA (2013) Urodynamic testing–is it a useful tool in the management of children with cutaneous stigmata of occult spinal dysraphism? J Urol 189:678–683
11. Liu FY, Li JF, Guan X, Luo XF, Wang ZL, Dang QH (2011) SEM study on filum terminale with tethered cord syndrome. Childs Nerv Syst 27:2141–2144
12. MacNeily AL, Leonard MP, Metcalfe PD, Casale A, Guerra L, Steinbok P, Garton H (2007) Development of an objective score to quantify the pediatric cystometrogram. J Urol 178:1752–1756, discussion 1756–1757
13. Metcalfe PD, Luerssen TG, King SJ, Kaefer M, Meldrum KK, Cain MP, Rink RC, Casale AJ (2006) Treatment of the occult tethered spinal cord for neuropathic bladder: results of sectioning the filum terminale. J Urol 176:1826–1829, discussion 1830
14. Meyrat BJ, Tercier S, Lutz N, Rilliet B, Forcada-Guex M, Vernet O (2003) Introduction of a urodynamic score to detect pre and postoperative neurological deficits in children with a primary tethered cord. Childs Nerv Syst 19:716–721
15. Nakanishi K, Tanaka N, Kamei N, Nakamae T, Izumi B, Ohta R, Fujioka Y, Ochi M (2013) Use of prone position magnetic resonance imaging for detecting the terminal filum in patients with occult tethered cord syndrome. J Neurosurg Spine 18:76–84
16. Nazar GB, Casale AJ, Roberts JG, Linden RD (1995) Occult filum terminale syndrome. Pediatr Neurosurg 23:228–235
17. Ogiwara H, Lyszczarz A, Alden TD, Bowman RM, McLone DG, Tomita T (2011) Retethering of transected fatty filum terminales. J Neurosurg Pediatr 7:42–46
18. Ostling LR, Bierbrauer KS, Ct K (2012) Outcome, reoperation, and complications in 99 consecutive children operated for tight or fatty filum. World Neurosurg 77:187–191
19. Selcuki M, Coskun K (1998) Management of tight filum terminale syndrome with special emphasis on normal level conus medullaris (NLCM). Surg Neurol 50:318–322, discussion 322
20. Selcuki M, Unlu A, Ugur HC, Soygur T, Arikan N, Selcuki D (2000) Patients with urinary incontinence often benefit from surgical detethering of tight filum terminale. Childs Nerv Syst 16:150–154, discussion 155
21. Selden NR, Nixon RR, Skoog SR, Lashley DB (2006) Minimal tethered cord syndrome associated with thickening of the terminal filum. J Neurosurg 105:214–218
22. Singh S, Kline-Fath B, Bierbrauer K, Racadio JM, Salisbury S, Macaluso M, Jackson EC, Egelhoff JC (2012) Comparison of standard, prone, and cine MRI in the evaluation of tethered cord. Pediatr Radiol 42:685–691
23. Steinbok P, Garton HJ, Gupta N (2006) Occult tethered cord syndrome: a survey of practice patterns. J Neurosurg 104:309–313

24. Steinbok P, Kariyattil R, MacNeily AE (2007) Comparison of section of filum terminale and non-neurosurgical management for urinary incontinence in patients with normal conus position and possible occult tethered cord syndrome. Neurosurgery 61:550–555, discussion 555–556
25. Tani S, Yamada S, Fuse T, Nakamura N (1991) Changes in lumbosacral canal length during flexion and extension–dynamic effect on the elongated spinal cord in the tethered spinal cord. No To Shinkei 43:1121–1125
26. Tehli O, Hodaj I, Kural C, Solmaz I, Onguru O, Izci Y (2011) A comparative study of histopathological analysis of filum terminale in patients with tethered cord syndrome and in normal human fetuses. Pediatr Neurosurg 47:412–416
27. Valentini LG, Selvaggio G, Visintini S, Erbetta A, Scaioli V, Solero CL (2011) Tethered cord: natural history, surgical outcome, and risk for Chiari malformation 1 (CM1): a review of 110 detethering. Neurol Sci 32(Suppl 3):S353–S356
28. Vogelzang NJ, Raghaven D (1990) Criteria of response to neoadjuvant chemotherapy in the treatment of invasive bladder cancer: a critical assessment. Prog Clin Biol Res 353:225–236
29. Warder DE, Oakes WJ (1993) Tethered cord syndrome and the conus in a normal position. Neurosurgery 33:374–378
30. Warder DE (2001) Tethered cord syndrome and occult spinal dysraphism. Neurosurg Focus 10:e1
31. Wehby MC, O'Hollaren PS, Abtin K, Hume JL, Richards BJ (2004) Occult tight filum terminale syndrome: results of surgical untethering. Pediatr Neurosurg 40:51–57, discussion 58
32. Wolin LH (1969) Stress incontinence in young, healthy nulliparous female subjects. J Urol 101:545–549
33. Yamada S, Won DJ, Pezeshkpour G, Yamada BS, Yamada SM, Siddiqi J, Zouros A, Colohan AR (2007) Pathophysiology of tethered cord syndrome and similar complex disorders. Neurosurg Focus 23:E6