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Systematic Review of Urologic Outcomes from Tethered Cord Release in Occult Spinal Dysraphism in Children

Jeffrey T. White[1] · **Derek C. Samples**[2] · **Juan C. Prieto**[1,3,4] · **Izabela Tarasiewicz**[2]

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Abstract Tethered cord syndrome describes a condition of multisystem end organ dysfunction due to fixation of the spinal cord. This systematic review focuses on the closed skin variant of this condition, occult spinal dysraphism. The embryology, pathophysiology, presentation, and classification of occult spinal dysraphism are explained to develop a simple framework for discussions regarding this often confusing condition. Following Preferred Reporting Items for Systematic Review and Meta-Analyses (PRISMA) guidelines, we synthesized urologic outcome data after tethered cord release in children from 17 studies performed over the past 25 years. These results prompted several conclusions. First, the different subgroups and different nomenclature of tethered cord syndrome are often confused, making interpretation of results difficult. Second, untethering has a positive effect on urologic symptoms and urodynamics parameters. Third, timing of untethering is important: early intervention prevents significant long-term traction aiming to avoid irreversible neurologic damage. Fourth, pediatric urologists and neurosurgeons have an important role in diagnosing and treating this condition and should work closely as part of a multidisciplinary team.

Keywords Tethered cord syndrome · Pediatric urology · Occult spinal dysraphism · Urinary tract infection · Detrusor overactivity

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✉ Juan C. Prieto
jcprietos@hotmail.com

Jeffrey T. White
whitej9@uthscsa.edu

Derek C. Samples
samplesd@uthscsa.edu

Izabela Tarasiewicz
tarasiewicz@uthscsa.edu

[1] Department of Urology, University of Texas Health Science Center at San Antonio, 7703 Floyd Curl Dr, Mail Code 7845, San Antonio, TX 78229, USA

[2] Department of Neurosurgery, University of Texas Health Science Center at San Antonio, 7703 Floyd Curl Dr, Mail Code 7843, San Antonio, TX 78229, USA

[3] Children's Hospital of San Antonio, San Antonio, TX, USA

[4] Methodist Children's Hospital, San Antonio, TX, USA

Abbreviations

BMP	Bone morphogenetic protein
CCM	Caudal cell mass
DO	Detrusor overactivity
DSD	Detrusor-sphincter dyssynergia
FF	Fatty filum
GUS	Genitourinary system
LMMC	Lipomyelomeningocele
LUT	Lower urinary tract
MMC	Myelomeningocele
NTD	Neural tube defect
OSD	Occult spinal dysraphism
OTCS	Occult tethered cord syndrome
PVR	Post-void residual
SC	Spinal cord
SCM	Split cord malformation
TCR	Tethered cord release
TCS	Tethered cord syndrome
UDS	Urodynamic studies
UTI	Urinary tract infection

Introduction

Tethered cord syndrome (TCS) in children is described as an array of congenital anomalies, including cutaneous, urologic, neurologic, and orthopedic dysfunction. It is thought to result from abnormal fixation of the distal spinal cord (SC) during embryogenesis. Prognosis of this syndrome is highly dependent on the degree of traction. Any delay in diagnosis and treatment can have serious and possibly irreversible consequences. All disciplines involved in the care of TCS patients (urology, neurosurgery, orthopedic surgery, physiatry) must be aware of the syndrome and its presentation to achieve early detection. The following paper reviews the key aspects of the closed skin variant of TCS, occult spinal dysraphism (OSD), to facilitate a better understanding within the surgical community. This review is the product of a combined effort between pediatric neurosurgery and urology teams.

The aim of the present review article was to address three main issues in regard to OSD: (1) understanding the definition, terminology, embryology, pathophysiology and classification of OSD; (2) elucidating the role of urodynamics (UDS) in OSD; and (3) analyzing the indications for neurosurgical intervention.

Embryology

Tethered cord syndrome is caused by abnormal development of the central nervous system. The genitourinary system (GUS) is an organ system classically affected by tethered cord syndrome [1]. Understanding the embryology behind both spinal and urological development is critical to understanding this relationship.

The SC forms through primary and secondary neurulation. In primary neurulation, the cutaneous ectoderm separates from the neuroectoderm and fuses in the midline, internalizing the neural tube. This process is disjunction. The mesoderm then migrates between the ectoderm and neuroectoderm to form the posterior bony and soft tissue elements. Disruption of disjunction is responsible for many SC pathologies, including MMC, intraspinal lipoma, lipomyelomeningocele (LMMC), and dermal sinus tract. Failure of primary neurulation results in an open neural tube defect.

Secondary neurulation refers to formation of the spinal elements caudal to S2. In this process, the SC ascends; regression continues until the conus reaches the adult level. Errors during secondary neurulation contribute to formation of terminal lipomas/myelocystoceles and tight/fatty filum (FF) [2, 3].

Maldevelopment of the distal SC components affects other end organs such as the GUS. The etiology of GUS disease is twofold: (1) errors in neural tube formation leading to maldevelopment of the GUS and (2) changes in lower urinary tract (LUT) function due to altered innervation. Maldevelopment of the neural tube alters mesodermal development and consequently mesodermal organs, such as the GUS [4, 5]. Due to changes in spinal nerve function from NTD, altered innervation patterns cause smooth muscle transdifferentiation modifying the bladder smooth muscle layer and the extracellular matrix [6, 7•]. These bladders are characterized as small capacity and with poor compliance as early as 20 weeks gestation [8, 9].

Finally, neural tube growth is responsible for anatomical positioning of the cloacal membrane [5]. Improper neural tube development will affect the partitioning of the distal genitourinary and gastrointestinal tracts. Birth defects of the distal genitourinary or gastrointestinal tracts (cloaca, imperforate anus, exstrophy-epispadias complex, etc.) should prompt an inquiry into SC malformation.

Pathophysiology

Pang and colleagues reported that the degree of traction on the conus determines the pathology of TCS [10]: significant traction causes earlier presentation with more severe symptoms. Otherwise, patients remain asymptomatic with subclinical dysfunction; additional stretching from growth spurts or other events (strenuous exercise, pregnancy, trauma, etc.) may trigger symptoms.

Yamada and colleagues presented a mechanism of injury in animal models of TCS [1]. They showed that the degree of caudal traction on the SC correlates to the severity of the neurological deficit, a principle termed traction-induced hypoxia. A reduction in blood flow secondary to the force of traction causes proportional SC damage resulting in end organ dysfunction [11]. Low amplitude traction produces reversible injury; high amplitude forces cause irreversible SC injury. This would suggest that earlier release of a tethered cord should prevent further permanent SC damage and possibly reverse any temporary organ dysfunction.

Diagnosis: Clinical Presentation

TCS in children presents with a myriad of subtle symptoms [12]. Providers must be aware of the different possible signs and symptoms to achieve early diagnosis and improve outcomes. Cutaneous lesions, including midline hairy patches, hemangiomas, dermal pits/sinuses, hypertrichosis, subcutaneous lipoma, "cigarette burns," lumbosacral appendage, and nevi, are seen in 80 % of OSD patients [3, 13, 14]. Neurological manifestations, which are due to disruption of motor and sensory pathways to the lower extremities, include delayed gait, hyper-/hyporeflexia, muscular atrophy, spasticity, poor sensation or proprioception, and painless ulcerations of the feet or legs [12, 15]. Pain as the presenting symptom is much less common in pediatrics. Orthopedic abnormalities, including foot deformities, limb length discrepancies, gluteal

asymmetry, scoliosis, and vertebral anomalies, such as bifid vertebrae, laminar anomalies, hemivertebra, and sacral agenesis, are found in 90 % [3, 12]. Urologic symptoms range from incontinence, urgency, frequency, and recurrent UTIs to subtle changes observed on urodynamic studies (UDS) [12]. Bladder symptoms are difficult to assess in infants; therefore, patients may not present until toilet training is attempted. UDS abnormalities, which precede clinical symptoms, can be used prior to toileting as an assay for GUS dysfunction. This highlights the importance of a complete urological workup to prevent delayed diagnosis and treatment.

TCS is often associated with other congenital syndromes such as caudal agenesis and anorectal atresia syndromes: omphalocele, cloacal exstrophy, imperforate anus, and spinal anomalies (OEIS), vertebral anomalies, anal atresia, cardiac anomalies, TE fistula, renal and limb anomalies (VACTERL), and Currarino/anorectal malformation, sacrococcygeal osseous defect, presacral mass (ASP) triad [16]. Screening for OSD should be standard of care in syndromic patients.

Diagnosis: Radiographic and Urodynamic Studies

Diagnosis of TCS requires radiographic findings correlating with clinical symptoms. Though ultrasound can be useful in infants less than 6 months of age as a screening tool, variability among ultrasonographers and difficult interpretation limits its use [17•]. MRI is the imaging procedure of choice for assessment of OSD [18]. T1-weighted imaging provides clear anatomical detail of neural tissue and the diameter of the filum, allowing for evaluation of vertebral levels and the presence of fat/thickening [18, 19]. For this paper, a normal conus is considered terminating at or above L2. A low-lying cord in TCS refers to a filum below the L2 vertebral body; a filum diameter greater than 2 mm is considered abnormally thickened in children [10, 18, 20–23].

Classification of TCS Presentation

It is important to differentiate TCS according to natural history, comorbidities, and differing severities. Van Leeuwen et al. suggested a tethered cord classification of four groups based on the origin of tethering (Table 1): (1) post-MMC repair, (2) fatty/tight filum terminale, (3) LMMC/conus lipoma, and (4) split cord malformation (SCM) [24]. These groups are unfortunately intermingled in published articles. From a diagnosis and management perspective, it is important to acknowledge and understand the different subgroups.

The first group describes TCS following MMC repair. For the purpose of this article, we will not discuss initial repair or retethering.

Group 2 is comprised of patients with a fatty/tight filum terminale (filum lipoma); this is caused by fat infiltration of the filum during secondary neurulation. This group often includes other caudal developmental abnormalities (VACTERL, Currarino/APS, etc.) and occurs in 0.1 % of school children [19]. These patients present without cutaneous markers or neurologic/urologic symptoms; thus, patients are older at diagnosis and present with severe symptoms.

Patients with LMMC, or conus lipoma, constitute the third group of OSD. The anomaly occurs during early disjunction: mesodermal elements fuse with the SC preventing bony element formation. With an estimated incidence of 1:400, LMMC is the most common spinal anomaly [2]. Diagnosis is typically made in infancy due to cutaneous findings: usually a non-tender, subcutaneous fatty mass [25]. The most common initial neurologic manifestation is bladder dysfunction [15], and the natural history of these lesions is progressive neurological deterioration [2, 25]. Early diagnosis and intervention is critical.

The final group is SCM or diastematomyelia. This anomaly accounts for 25 % of OSD and results from ectoderm-endoderm adhesions during early gastrulation bisecting the SC [26]. Tethering occurs at the bisecting bony spur/dorsal band as well as the fatty/thickened filum. Cutaneous stigmata (usually lumbosacral hair tuft), orthopedic anomalies and scoliosis are common. Up to 85 % of patients have tandem neurodevelopmental lesions (FF, LMMC, MMC, meningocele manqué, and chiari). Seventy-five percent of SCM patients develop urological abnormalities [22, 26–29].

Methods

Protocol, Information Sources, Study Selection

We performed a systematic literature review via PubMed and Ovid; search terms “spinal dysraphism,” “tethered cord release,” and “urodynamics” were assayed in the pediatric literature since 1990. Intercollegial discussion produced two unpublished manuscripts which were included in our analyses. We followed Preferred Reporting Items for Systematic Review and Meta-Analyses (PRISMA) guidelines; details of the protocol for this systematic review were registered on PROSPERO (CRD42015024762).

Study Selection, Eligibility Criteria, Exclusion Criteria

From 536 publications, 167 were duplicates, leaving 369 records to screen. Full text articles were available on 234 of these records; 2 were unpublished manuscripts. Records reporting results only in languages other than English as well as those regarding open spinal dysraphism, secondary tethered cord, and adult patients were excluded. Likewise, records that did not report preoperative or postoperative urological symptoms or UDS were excluded. Of the original 536 records, 17 manuscripts were examined.

Table 1 Classification of spinal dysraphism causing tethered cord syndrome [24]

Classification	Signs	Urologic symptoms	Comorbidities
I. Myelomeningocele	Open defect (repaired at birth)	First sign of retethering; neurogenic bladder	Chiari II; hydrocephalus; SCM
II. Tight/fatty filum (filum lipoma)	Skin covered; often no overlying cutaneous marker	Asymptomatic; if present, more likely irreversible	More often seen with caudal syndromes (VACTERL, Currarino, sacral agenesis, etc.)
III. Lipomyelomengingocele (Conus lipoma)	Skin covered; fat pad; skin dimple	Often first sign; 50 % symptomatic at birth; 25 % of asymptomatic patients will progress	Other urogenital malformations (~25 %) [25]
IV. Split cord malformation	Skin covered; hair tuft (most common); scoliosis; limb asymmetry	Urologic dysfunction (75 %)	Multiple lesions (50–85 %; spinal lipoma, MMC, meningocele manqué, chiari)

Data Collection, Data Items

The following information was obtained from each study: number of patients studied, types of spinal dysraphism lesions, age at presentation and at intervention, length of postoperative follow-up, cutaneous presenting signs, preoperative neurological examination, postoperative neurological examination, preoperative urological symptoms, postoperative urological symptoms, preoperative urodynamic data, and postoperative urodynamic data. For each study, the pre- and postoperative data (neurological examination, urological symptoms, and urodynamic data) were compared to obtain a number and percentage of patients that were unchanged, improved, or worsened. These data elements were collected and summarized in a table format (Table 2).

Bias Detection, Summary Measures and Result Synthesis

Bias was assessed according to the recommendations of Hayden et al. [30]. The following biases were assessed for each study: study participation, study attrition, prognostic factor measurement, outcome measurement, confounding measurement and account, and analysis (Supplement table 1). Any study with high risk for bias in any category was excluded from the review. Though their results were included in Table 2, the studies by Abrahamsson et al. [31] as well as Broderick and colleagues [32•] were excluded from the synthesis due to high level of confounding study participation, outcome assessment, and analysis biases. Due to intermingling of OSD lesions as well as nonuniformity of reporting, statistical analyses of pooled study elements could not be performed. Data are summarized and reported without further statistical interpretation.

Results with Discussion

TCS UDS findings have been described previously by Kearns et al. [33••]. The most common UDS findings in TCS are detrusor overactivity (DO), detrusor-sphincter dyssynergia (DSD), and decreased compliance. Kearns and colleagues recommended performing UDS prior to untethering surgery and 3 to 6 months after the procedure [32•, 33••].

Analyzing the literature for effects of untethering in patients with primary TCS is difficult. Authors often confuse obsolete terminology, use different classification systems, and combine patients with varying etiologies (e.g., including OSD patients with MMC or secondary TCS patients). Here, we remind practitioners of a classification system for tethered cord syndrome based on the work of Van Leeuwen et al. [24]. This system regroups TCS presentations based on natural history and disease severity, preventing further intermingling in future studies regarding TCS presentations.

Publications from both pediatric neurosurgeons and pediatric urologists report varying degrees of success from untethering: improvements in UDS range from 5 to 93 % and improvement in urological symptoms range from 17 to 70 %. These wide ranges are likely due to the varied timing of surgical intervention: a longer tethered time and greater amount of conus traction results in further permanent damage, making untethering less successful (Wang et al., 2015, unpublished).

Group I: Myelomeningocele

Literature supports closure and untethering at or before birth [34, 35]. As a result of untethering, patients are at risk for retethering. Urological dysfunction will often be the first indication. If left untreated, the natural history of this process has demonstrated symptomatic progression in up to 60 % of patients in the first 5 years [36, 37]. These patients require both neurosurgical and urological follow-up to permit early diagnosis of neurologic dysfunction and correctional interventions.

Group II: Tight or Fatty Filum (Filum Lipoma)

The true incidence of this condition is unknown. Cadaveric studies estimate its prevalence at 3.7 %, MRI studies at 1.5 to

Table 2 Synthesis of studies on tethered cord release in closed spinal dysraphism in children

Reference	*N*	Type of lesion (*n*)	Mean (age range) at baseline, months or years	Mean (range) postoperative follow-up in months	LUTS, *n*/*N* (%)	LUTS at baseline (*n*/*N*)	Postoperative improvement urological symptoms (*n*/*N*)	Abnormal preoperative/baseline UDS, (*n* or *n*/*N*)
Khoury et al., 1990 [44]	31	OSD (31): suspected TCS secondary to a thickened filum (4) or normal conus (OTCS, 27)	8.6 years (3.5–17 years)	13.3 (3 to 33)	97	Daytime incontinence (29/31); enuresis (30/31)	Improved daytime incontinence 8/29 (72 %)	DO (27); areflexic detrusor with elevated PVR (1); abnormal compliance (24)
AAtala et al., 1992 [45]	35	LMMC (35)	Bimodal: younger group 3 months (birth–15 months; *n*=29); older group 10 years (4.5–19 years; *n*=6)	N/A	100 (6/6 in older group)	Older group 6/6: urgency (5); incontinence (1)	Older group 1/6 (17 %)	Younger group 11/29 (38 %): DSD, DO, abnormal motor unit potentials; older group 6/6 (100 %): detrusor areflexia, DO
Wu et al., 1998 [46]	43	LMMC (43): conus medullaris (38), conus and filum (3), filum terminale (2)	1 month–25 years. divided into two groups according to age at surgery: before or after 1.5 years	68 for early group and 69 for late group	N/A	N/A	N/A	Young group (<1.5 years): normal (16), poor empty/synergic (4), poor empty/DSD (1), flaccid/DSD (1), DO/synergic (1), DO/DSD (1), poor compliance/synergic (1) Older group (>1.5 years): normal (3); poor empty/synergic (5); flaccid/synergic (2); DO/synergic (2); DO/DSD (3); DO/incontinent (1); poor compliance/DSD (2)
Arikan et al., 1999 [53]	17	OSD (16); LMMC (1); diastematomyelia (1)	8.7 years	(6–14)	100	Urgency-frequency (12); daytime incontinence (11); UTI (6); urge incontinence (2); intermittency (1); overflow incontinence (1); nocturnal incontinence (1)	N/A	DO (16); high DLPP (8); decreased compliance (4); DSD (3); areflexia (1)
Nogueira et al. 2004 [39]	19	OSD (19)	Group 1, orthopedic: 8 years (8 months to 14 years)	N/A	32	Daytime incontinence (4); UTI (4); nocturnal incontinence (1)	Improved incontinence (4, only patient without improvement was daytime incontinence)	Normal (16) DO (3)
	16	OSD (16)	Group 2, cutaneous lesions: 1 year (1 week to 11 years)	N/A	All prior to toilet training	N/A	N/A	Normal (12) DO (4)
	13	OSD (13)	Group 3, voiding symptoms: 9 years (4.5 to 4 years)	N/A	100	Daytime/nocturnal incontinence (12); UTI (7); urgency (5); frequency (4)	None (7); daytime/nocturnal incontinence (5); UTI (1)	DO (11) Detrusor areflexia (2)
	6	OSD (6)	Group 4, congenital syndromes: 5 years (birth to 10 years)	N/A	50 %, 2 patients not toilet-trained	Daytime urinary incontinence (1); nocturnal incontinence (1); UTI (1); urgency-frequency (1)	None (1); nocturnal incontinence (1); UTI (1); urgency-frequency (1)	1/2 (50 %)

Table 2 (continued)

Reference	*N*	Type of lesion (*n*)	Mean (age range) at baseline, months or years	Mean (range) postoperative follow-up in months	LUTS, *n*/*N* (%)	LUTS at baseline (*n*/*N*)	Postoperative improvement urological symptoms (*n*/*N*)	Abnormal preoperative/baseline UDS, (*n* or *n*/*N*)
Guerra et al., 2006 [40]	24	Thickened filum (9); lipoma (9); syrinx (8); spina bifida occulta (5); diastematomyelia (4)	6 years (1 month–12 years)	6.4	54	Daytime urinary incontinence (7); urgency-frequency (7); UTI (6)	Daytime urinary incontinence (1); UTI (1)	Normal (3) DO (17) Low bladder capacity and compliance (4)
Metcalfe et al., 2006 [41]	36	Normal (27); conus L2–L3 (2); hydromyelia (1); minimal fatty infiltration (6)	8.3 years (1.2–15 years)	49	N/A	Daytime urinary incontinence (30); nocturnal incontinence (28); UTI (13)	Urinary improvement in 26/36 patients; 22/36 were able to discontinue anticholinergics	DO (25); less than 50 % of expected capacity (21)
Abrahamsson et al., 2007 [31]	20	MMC (20)	8 years (2–13 years)	12 (6–19)	20	Incontinence (4)	Incontinence improved (3), worsened (1)	Severe dysfunction (9, baseline detrusor pressure >40 cm H2O); moderate dysfunction (6, baseline detrusor pressure 20–4 cm H2O; mild dysfunction (5, baseline detrusor pressure <20 cm H2O)
Macejko et al., 2007 [47]	79	FF (26); LMMC (26)	9.6 months	5.2 years (6 months to 11.2 years)	N/A	Urinary retention (1); UTI (1)	Normal (2)	Hypotonia (14); DSD (7); high voiding pressure (6);
Kumar et al., 2008 [48]	15	LMMC (7); SCM (4); thickened filum (3); neuroenteric cyst (1)	7.6 years (3 months–18 years)	6–8	53	Frequency, urgency, straining, incomplete emptying (3); occasional incontinence with dry periods (1); continuous incontinence (4)	7 improved, 1 worsened	13/15 abnormal preoperative urodynamic (DO, detrusor areflexia, incontinence, decreased compliance, elevated PVR)
Lavallee et al., 2013 [54]	123	OSD (123)	11 month	19.6 (6–39)	10	UTI (10)—only 11 patients toilet-trained	UTI (3)	Normal (100, 81 %); DO (33, 27 %); low bladder capacity (43, 35 %); low compliance (28, 23 %)
Broderick et al., 2014 [32•]	38	OSD (38)	3 years (0.2–16.3 years)	N/A	0	Asymptomatic	Asymptomatic	Normal (31)
Frainey et al., 2014 [42•]	59	FF or low-lying cord	59 months (2–277 months)	84 (12–192)	44	Incontinence (11)	Incontinence (5)	Normal (23/52); high risk (2/52); indeterminant (27/52)
Kim et al., 2014 [49•]	44	LMMC (37); filum terminale lipoma (7)	38 months (2–384 months)	57.3 (26–200)	27	Incontinence (7); febrile UTI (4); weak stream (3); straining (1)	Febrile UTI (9); incontinence (2); urgency (1)	UDS scores before and after TCR were not statistically significant. Subset analysis showed lower overall scores 6 months postoperatively reflected long-term outcomes.
(Steinbok et al., 2015, unpublished)	21	OSD (19), FF (1)	9.3 years (6–18 years)	12	N/A	No statistically significant difference in QOL or dysfunctional voiding scores between the medical and surgical groups.		There was no statistically significant change in UDS scores between the medical and surgical groups. A sample size of 1400 patients is required to show a statistically significant result.
(Wang et al., 2015, unpublished)	102	Low conus (48), FF (30), syringomyelia (29), vertebral body	4.3 years	6.8 years	33	Incontinence (34), LUTS (15), UTI (8)	Incontinence (17)	N/A

Table 2 (continued)

Reference	*N*	Type of lesion (*n*)	Mean (age range) at baseline, months or years	Mean (range) postoperative follow-up in months	LUTS, *n*/*N* (%)	LUTS at baseline (*n*/*N*)	Postoperative improvement urological symptoms (*n*/*N*)	Abnormal preoperative/baseline UDS, (*n* or *n*/*N*)
		dysmorphism (20), lipoma (19)						
Yener et al., 2015 [43••]	40	Tight filum (30); dermal sinus (3); filum lipoma (6); SCM (4), intradural lipoma (2); neuroenteric cyst (2); syrinx (11); lipoma (2); lipomyelocele (1); sacral cyst (1); spinal cord tension factor (7)	7.2 years (1.5–16 years)	2.8 years (1–6.6 years)	60	Incontinence (9); urgency (1); frequency (2); UTI (5); urinary retention (1)	Incontinence (4); urgency (1); urinary retention (1); UTI (2)	Abnormal leak pressure (23); abnormal maximum pressure (33); elevated PVR (20); abnormal compliance (17)

Reference	Postoperative/baseline UDS, (*n* or *n*/*N*)	Postoperative improvement in UDS after TCR (*n*/*N*)	Postoperative detereloration of preopeartive normal UDS (*n*/*N*)	Lumbosacral cutaneous lesions (*n*/*N*)	Preoperative abnormal neurological examination (*n*/*N*)	Postoperative improvement neurological examination (*n*/*N*)
Khoury et al., 1990 [44]	DO (27); areflexic detrusor with elevated PVR (1); abnormal compliance (24)	DO improved 16/27 (59 %)	0	Sacral dimple (2)	0	N/A
AAtala et al., 1992 [45]	DSD (15); DO (7); detrusor areflexia	Younger group 9/11 (82 %); older group 1/6 (17 %)	Younger group 1/18 (6 %); DSD	Subcutaneous lipoma (35); skin dimple (10); hairy nevus (2); skin appendage (1); dermal vascular malfunction (7)	Younger group 14/29 (48 %); older group 6/6 (100 %)	Younger group 71 % (10/14); older group 1/6 (17 %)
Wu et al., 1998 [46]	Young group (<1.5 years): normal/synergic (13); poor empty/incontinent (1); poor empty/synergic (1); flaccid/synergic (2); flaccid/DSD (1); DO/DSD (1)	0/25 (0 %)	5/25 (20 %)	N/A	N/A	N/A
	Older group (>1.5 years): normal/synergic (3); flaccid/synergic (2); flaccid/DSD (1); poor empty/synergic (3); poor empty/DSD (1); DO/synergic (2); DO/DSD (3); poor compliance/DSD (2)	2/18 (11 %)	1/18 (6 %)			
Arikan et al., 1999 [53]	8 patients improved in 6 months (47 %) but only 5 (29 %) had long-term improvement at 14 months	5/17 (29 %)	N/A	6/17 (35 %)	N/A	N/A

Table 2 (continued)

Reference	Postoperative/baseline UDS, (*n* or *n*/*N*)	Postoperative improvement in UDS after TCR (*n*/*N*)	Postoperative detereloration of preopeartive normal UDS (*n*/*N*)	Lumbosacral cutaneous lesions (*n*/*N*)	Preoperative abnormal neurological examination (*n*/*N*)	Postoperative improvement neurological examination (*n*/*N*)
Nogueira et al. 2004 [39]	Normal (15); DO (1) Normal (2); DO (1)	N/A 2/3 (66 %)	1/16 (6 %)	3/19 (16 %)	N/A	N/A
	Normal (5), only performed in 5 patients Normal (4)	N/A 4/4 (100 %)	0	16/16 (100 %)	N/A	N/A
	Normal (4); DO (6); no postoperative UDS (1) Normal (1); detrusor areflexia (1)	5/11 (45 %) 1/2 (50 %)	0	0	N/A	N/A
	Normal (1) DO (5)	N/A Normal (2); DO (3)	N/A 2/5 (40 %)	0	N/A	N/A
Guerra et al., 2006 [40]	DO (2); low capacity and compliance (1) Normal (10); DO (1); low capacity and compliance (1) Low capacity and compliance (4)	0 10/17 (59 %) 0	3/3 (100 %) 1/17 (6 %) 0	21/24 (88 %)	13/24 (54 %)	N/A
Metcalfe et al., 2006 [41]	28/36 patients with both pre- and postoperative urodynamics; 16 patients (57 %) noted improved parameters; DO resolved in 8/25 (32 %),	16/28 (57 %)	0	7/36 (19 %)	16/36 (44 %)	12/16 (75 %)
Abrahamsson et al., 2007 [31]	Severe (5); moderate (6); mild (9)	7/20 (35 %)	1/20 (5 %)	N/A	N/A	N/A
Macejko et al., 2007 [47]	Hypotonia (5); DO (1); high voiding pressures (6)	8/30 (27 %)	10/36 (28 %)	69 (87 %)	14 (18 %)	N/A
Kumar et al., 2008 [48]	Improved in 6/15, 3/15 worsened, 6/15 unchanged	6/15 (40 %)	3/15 (20 %)	15/15 (100 %)	Motor 11/15 (73 %); sensory 8/15 (53 %)	Motor 7/11 (64 %); sensory 3/8 (38 %)
Lavallee et al., 2013 [54]	DO (14, 35 %); low bladder capacity (13, 33 %) and compliance (12, 30 %)	N/A	N/A	123/123 (100 %)	N/A	N/A
Broderick et al., 2014 [32•]	15 postoperative UDS only: 14 improved, 1 worsened s/p wound infection with areflexic detrusor	14/15 (93 %)	1/15 (7 %)	33/38 (87 %)	N/A	N/A
Frainey et al., 2014 [42•]	Normal (18/49); high risk (3/49); indeterminant (28/49)	N/A	N/A	20/59 (34 %)	39/59 (66 %)	N/A
Kim et al., 2014 [49•]	UDS scores before and after TCR were not statistically significant. Subset analysis showed lower overall scores 6 months postoperatively reflected long-term outcomes.	N/A	N/A	N/A	20/44 (45 %)	N/A
(Steinbok et al., 2015, unpublished)	There was no statistically significant change in UDS scores between the	N/A	N/A	N/A	N/A	N/A

Table 2 (continued)

Reference	Postoperative/baseline UDS, (*n* or *n*/*N*)	Postoperative improvement in UDS after TCR (*n*/*N*)	Postoperative deterioration of preoperative normal UDS (*n*/*N*)	Lumbosacral cutaneous lesions (*n*/*N*)	Preoperative abnormal neurological examination (*n*/*N*)	Postoperative improvement neurological examination (*n*/*N*)
	medical and surgical groups. A sample size of 1400 patients is required to show a statistically significant result.					
(Wang et al., 2015, unpublished)	N/A	17/34 (50 %) symptoms-based only	N/A	N/A	N/A	N/A
Yener et al., 2015 [43••]	Abnormal leak pressure (16); abnormal maximum pressure (21); elevated PVR (15); abnormal compliance (10)	13/24 (54 %) completely resolved urinary symptoms	3/26 (12 %) developed de novo urinary symptoms	22/40 (55 %)	N/A	N/A

5 % [19, 38]. Patients often present at older ages due to a lack of cutaneous findings.

For patients with VACTERL syndrome, there is a high risk of FF. According to Nogueira, these patients had a high rate of clinical symptoms and UDS abnormalities (50 %) which improved after untethering by 33 and 40 % of patients, respectively [39]. Therefore, it is imperative to perform urological evaluation on VACTERL patients.

Guerra et al. published a retrospective review of 24 patients after TCR, including 9 FF patients [40]. Preoperatively, 58 % were toilet-trained with median age of 8.1 years and 42 % were not toilet-trained with median age of 8 months. Untethering resolved daytime incontinence in 93 % (p=0.04), corrected neurogenic DO in 59 %, and caused deterioration in few patients (n=3). Interestingly, the authors were able to stratify response by conus position: with conus at or below L3, there was a 50 % response in UDS parameter normalization as compared to a 100 % response with the conus at L1 and L2. This supports the pathophysiology of TCS: greater severity of traction results in worsened outcomes from untethering.

Metcalfe et al. evaluated 36 children, six with fatty infiltration, who underwent untethering after 2 years of failed medical therapy [41]. Clinical improvement in urinary symptoms occurred in 72 %. UDS improvements were documented in 57 % of cases.

Frainey et al. reported on the untethering of 59 children identified with MRI to have a FF or low-lying conus in order to identify factors predicting postoperative continence [42•]. Only two factors were statistically significant for postoperative continence: cutaneous lesions and preoperative continence status. Early normal postoperative UDS did approach significance (p=0.087) and may be an important indicator of long-term continence, but must be verified in a larger study.

Wang et al. performed a retrospective review of TCR in 102 children, 30 had FF, to identify preoperative variables predictive of postoperative continence (Wang et al., 2015, unpublished). They reported a 50 % resolution rate in incontinence. Preoperative urinary incontinence was the only variable significantly associated with increased odds of long-term bladder or bowel incontinence (OR=6.2, p=0.003). From these data, they conclude that once preoperative symptoms worsen, long-term beneficial results of untethering are limited.

Yener et al. prospectively followed 40 patients (30 had a tight filum) through TCR, recording changes in urinary symptom scores and UDS [43••]. Untethering improved urodynamic parameters, the most drastic of which was neurogenic DO. They noted a nearly overall 10 % increase in bladder capacity, 41 % with improved compliance and 25 % with improved PVR. Most importantly, they noted improved urodynamic parameters in patients even without urinary symptoms. They postulated that there is a negative effect on bladder dynamics from TCS despite the lack of symptoms. Taken

in conjunction with prior studies, this would suggest that this dysfunction is reversible since permanent neurological damage has not yet occurred.

Prior literature suggested that close observation may be prudent in asymptomatic tight filum/FF/filum lipoma. Steinbok and colleagues presented a prospective trial comparing untethering to medical observation (Steinbok et al., 2015, unpublished). They discovered no difference among groups, likely due to the long interval of tethering: surgical intervention was considered only after a year of failed medical therapy. Since the youngest patient was 5 years old, this allowed 6 years of traction prior to untethering. Similarly, any period of observation would only prolong tethering, leading to worsened permanent damage.

Our review on group II TCR patients suggested that TCR is beneficial for this class of patients. If untethering is performed prior to puberty, a 40 to 60 % rate of symptom as well as urodynamic resolution can be achieved. Most interestingly, multiple studies suggest a correlation between the natural history of the disease and the severity of the deficit: the greater degree of tethering (lower conus) as well as the greater time of tethering (greater age at presentation and untethering) resulted in worsened neurological deficits. These observations would suggest that earlier intervention or intervention prior to evidence of tethering symptoms would be beneficial [Wang et al., 2015, unpublished, 40, 43••, Steinbok et al., 2015, unpublished, 44].

Group III: Lipomyelomeningocele (Conus Lipoma)

As discussed above, LMMC is the most common OSD anomaly and these lesions produce progressive neurological deterioration. Thus older children and adults are more likely to present with irreversible urological findings. Early diagnosis and untethering is vital to prevent permanent sequelae.

The importance of early intervention can be gleaned from Atala et al. [45]. They described UDS findings in 35 children with LMMC before and after TCR. Most presented at an early age (before 15 months, mean age of 3 months); however, an older group was included with a mean age of 10 years. In the younger group, 83 % improved postoperatively; in the older group, only 17 % improved after surgery. Intervention improved the neurological examination in 71 % as well as LUT function in 82 %. This study supported prior observations that there is a high risk of progressive neurological deterioration in patients with untreated LMMC; furthermore, intervention at an older age produced poor results.

Wu et al. performed a retrospective review of 43 patients who underwent early TCR [46]. The patients were grouped by age at intervention, before or after age 1.5 years. They did not find statistically significant advantage for early neurosurgical repair ("advantage" was strictly defined as normal bladder and sphincter function at a follow-up greater than 5 years). Subgroup analysis demonstrated a favorable outcome in the late surgery group. This effect was likely due to detection of LMMC before onset of LUT dysfunction. Wu et al. continued to support early neurosurgical intervention in LMMC patients since there is a higher likelihood of normal preoperative LUT function that can be preserved with surgery.

Macejko et al. published a retrospective review of 79 cases of TCR, 26 of which were LMMC patients [47]. Cutaneous symptoms were the most common presenting sign (n=69). Preoperatively, 55 % had abnormal UDS; 13 % had postoperative poor urological outcomes. Seven of the ten patients were LMMC patients, leading to the conclusion that preoperative status of lipomatous spinal dysraphism was a risk factor for poor outcomes after prophylactic TCR.

Kumar et al. published a prospective study of tethered cord release in 25 children, 7 had LMMC [48]. Untethering produced the following improvements: 47 % in urological complaints, 73 % in motor dysfunction, and 53 % in sensory impairment. With regard to urodynamic outcomes, 40 % improved, 40 % were similar, and 20 % worsened.

Kim et al. published a retrospective review on 44 TCR patients, 37 of which were LMMC patients [49•]. A 71 % resolution rate of incontinence in untethered patients was reported. Furthermore, they discovered that early favorable UDS results 6 months after untethering reflected long-term outcomes.

Multiple studies suggest that prophylactic early intervention in LMMC is beneficial, though results may not be as encouraging as in group II TCS patients [45–47]. These benefits must be weighed against the risk of intervention: there is a risk of neurological damage up to 4 % and a surgical complication rate of 20 to 33 % in repairing LMMC lesions [21, 25]. Additionally, SC retethering may occur in 10 to 20 % of LMMC patients [50–52].

Group IV: Split Cord Malformation/Diastematomyelia

The SCM group accounts for approximately 25 % of OSD. Though urological dysfunction will be present in 75 % of cases, symptoms are rare, highlighting the importance of early formal urological evaluation with UDS [22, 25–29].

There are no formal studies regarding TCR outcomes in purely SCM patients. These patients are combined with other TCR patients such as in the studies by Guerra et al. [40], Kumar and colleagues [48], and Yener and colleagues [43••], each with four SCM patients included in their analyses. Another published manuscript by Arikan et al. discussed SCM [53]. Seventeen patients were diagnosed with OSD by MRI after presenting with LUT dysfunction and normal physical exam. All patients noted abnormal preoperative UDS. Clinical and UDS improvement occurred in 29 % of patients at 14 months of follow-up after untethering surgery. The authors

concluded that MRI was important in diagnosing these patients with OSD and, if suspected sooner, better results might have been realized with earlier untethering.

Our systematic review of the literature revealed that untethering surgery in children including SCM can improve both clinical symptoms and UDS parameters [40, 43••, 48, 53]. The fact that studies combine SCM patients with other TCR patient groups makes it difficult to separate this patient group. Thus, until SCM patients are reported separately, this group can only be characterized similarly to other TCR presentations.

Occult Tethered Cord Syndrome

There is a consensus among pediatric neurosurgeons that symptomatic patients with group 2, 3, or 4 classification should be treated surgically [12]. A more controversial situation is occult tethered cord syndrome (OTCS)—the symptomatic, medication refractory patient with normal imaging.

Nogueira et al. evaluated 54 children who underwent untethering [39]. The patients were divided in four subgroups according to the etiology of initial presentation (see Table 1). The group of children presenting with LUT dysfunction and a normal radiologic evaluation had the highest percentage of UDS abnormalities (100 %). After untethering, half of their patients improved both clinically (54 %) and objectively with UDS (50 %).

Khoury et al. [44] reported on sectioning of the filum in 31 children who failed conservative treatment for persistent urinary incontinence. Though only four patients had a FF, untethering was performed in all. Daytime incontinence improved in 72 %, urodynamic DO resolved in 59 %, and compliance increased in 66 % of the patients. This was a controversial study: surgery was performed in children with OSD, normal neurological examination, and normal imaging (except the four patients with a FF).

Despite these encouraging results, there is no type I evidence to firmly support surgical release of the filum for OTCS (medical refractory LUT symptoms with normal imaging). Our literature review showed that sectioning the normal filum in OTCS patients may be beneficial [39, 44]. It has been suggested that surgical release of the normal filum relieves tension and improves cranial migration of the SC [12]. Better results are obtained when these patients undergo intervention prior to permanent neurologic dysfunction which is heralded with end-organ symptoms, i.e., incontinence.

To coordinate care for these complex patients, close communication and a robust referral pattern between pediatric neurosurgeons and pediatric urologists is paramount. Pediatric urologists are better equipped to diagnose, manage and follow urodynamic and urologic abnormalities. Pediatric neurosurgeons may be able to identify clinical signs unnoticed by pediatric urologists such as abnormal reflexes, minimal asymmetry in the feet, scoliosis, MRI results, etc.

Conclusions

There is a lack of class I evidence regarding TCR in OSD. Studies are mostly retrospective in nature and lack uniformity in regards to definition, terminology, classification, and standardized UDS evaluation. This ambiguity has created discrepancies and inaccuracies when analyzing surgical outcomes and prognoses for these conditions. For this reason, we propose that clinicians stratify OSD patients according to the classification system of van Leeuwen [24].

Daytime urinary incontinence is the number one symptom associated with OSD and, in most cases, improves significantly after untethering [Wang et al., 2015, unpublished, 39, 41, 43••, 44, 47, 48, 54]. Similarly, neurogenic DO is the most common UDS finding and also shows high resolution rate after surgery [32•, Wang et al., 2015, unpublished, 39–41, 44, 45, 48]. It is important to note that the degree of resolution after untethering is inversely proportional to the permanent damage caused to the nerve fibers (Wang et al., 2015, unpublished). A longer period of tethering as well as a greater degree of tension creates worsening damage. The earliest form of this damage is heralded by bladder dysfunction first seen on UDS. This dysfunction, when extreme, results in urologic symptoms which are suggestive of permanent organ damage. It is important to intervene in TCS prior to the development of symptoms; UDS can provide an objective evidence of this dysfunction prior to symptomatology. For this reason, UDS is an important part of the workup in OSD.

It is paramount to have a dedicated multidisciplinary team to evaluate patients with OSD and primary TCS including pediatric neurosurgeons, pediatric urologists, physiatrists, and pediatric orthopedic surgeons. A close working relationship among these practitioners will ensure accuracy and precision in selection of surgical candidates. The real challenge for pediatric neurosurgeons is the prompt identification of patients with TCS or those at risk for TCS who would benefit from early surgical intervention to avoid future neurological deterioration. A pediatric urologist can assist in this endeavor with UDS.

Compliance with Ethics Guidelines

Conflict of Interest Jeffrey T. White, Derek C. Samples, Juan C. Prieto, and Izabela Tarasiewicz each declare no potential conflicts of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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