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REVIEW ARTICLE

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Evaluation and management of tethered cord syndrome in occult spinal dysraphism: Recommendations from the international children's continence society

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Aims: As awareness and frequency of tethered spinal cord (TSC) related to occult spinal dysraphism (OSD) has increased with magnetic resonance imaging (MRI), variability exists in its evaluation and management. Due to no published level I data, we summarize the current International Children's Continence Society (ICCS) recommendations for diagnosis and treatment of OSD.

Methods: Guidelines were formulated based on analysis of pertinent literature and consensus among authors. This document was vetted by the multidisciplinary members of the ICCS via its website before submission for peer review publication.

Results: The more frequent diagnosis of OSD is associated with increased operative intervention. Spinal cord untethering (SCU) has a highly variable risk profile, largely dependent on the specific form of OSD. Progressive neurological deterioration attributed to "tethered cord" may occur, with or without surgery, in selected forms of OSD whereas other cohorts do well.

Conclusion: Infants with classic cutaneous markers of OSD, with progressive neurologic, skeletal, and/or urologic findings, present no diagnostic or therapeutic dilemma: they routinely undergo MRI and SCU. Conversely, in asymptomatic patients or those with fixed, minor abnormalities, the risk profile of these OSD cohorts should be carefully considered before SCU is performed. Irrespective of whether or not SCU is performed, patients at risk for progression should be followed carefully throughout childhood and adolescence by a multidisciplinary team.

KEYWORDS
incontinence, neuro-orthopaedic syndrome, spinal dysraphism, tethered cord syndrome, tethered spinal cord

Abbreviations: ARM, anorectal malformation; CMG, cystometrography; DST, dermal sinus tract; EMG, electromyography; LDM, limited dorsal myeloschisis; LUT, lower urinary tract; LMM, lipomyelomeningocele; MMC, myelomeningocele; MRI, magnetic resonance imaging; OEIS Complex, omphalocele, exstrophy, imperforate anus, spinal defects; OSD, occult spinal dysraphism; OTCS, occult tethered cord syndrome, aka tight filum syndrome,; PURSMS, partial urorectal septal malformation sequence; SBO, spina bifida occulta; SCM, split cord malformation, aka diastematomyelia; SCU, spinal cord untethering; TCS, tethered cord syndrome, aka neuro orthopedic syndrome; TMC, terminal myelycystocele; TSC, tethered spinal cord; UDS, urodynamic studies; VACTERL, vertebral malformations, anal atresia, cardiac anomalies, TE fistula, renal abnormalities, limb defects.

Alan Wein led the peer-review process as the Associate Editor responsible for the paper.

1 | INTRODUCTION

Herein we summarize current evaluation and management strategies for infants and children with TSC due to OSD, a controversial topic among neurosurgeons, urologists and other care providers for these patients. This review excludes management of children with open myelomeningocele (MMC), though referenced where appropriate.

Children with complicated OSD have benefitted from management advances in MMC patients, but diagnosis and treatment options are often more nuanced. While prevalence of MMC has declined, OSD is now increasingly diagnosed due to greater availability, sensitivity and accuracy of MRI and ultrasound.

Greater access to good imaging promotes frequent, sometimes unnecessary imaging of patients with suspected cutaneous stigmata of OSD. Neurosurgeons encounter an abundance of patients diagnosed with OSD and face dilemmas about offering surgery, particularly for those who are ostensibly asymptomatic. Because the association of bladder and bowel functional issues exists, urologists can elucidate subtle, sub-clinical effects of some OSD through urodynamic studies (UDS), which aids in decisions to operate in selective cases.

This communication aims to analyze and present current data regarding diagnosis, evaluation, and treatment, either observational or surgical, from the ICCS. The multidisciplary makeup of this ICCS document reduces subjective bias and offers a consensus toward the challenging management of patients with OSD.

1.1 | Embryogenesis and classification of OSD

OSD encompasses a range of developmental anomalies of the spinal cord, occurring at every level, but most notably in the lumbosacral region. Given the association of lumbosacral anomalies to bladder and bowel function, these lower spinal lesions will be emphasized forthwith.

The natural history, mechanisms of neurological deterioration and surgical complexity vary considerably between different OSD types. Failure to appreciate this heterogeneity has undermined applicability of clinical studies; for example, thickened filum terminale is commonly included alongside spinal lipomas in untethering series.

OSD includes spinal cord malformations that are skin covered, in contrast with open spinal dysraphism (spina bifida aperta) that characteristically have skin defects and exposed neural tissue. Compared with "open" lesions, current knowledge of OSD is less advanced due to lack of appropriate animal models that permit detailed analyses of underlying embryological mechanisms.[1] Embryological classification of OSD is based on developmental stage (gastrulation, primary neurulation, or secondary neurulation) during which the dysraphic anomaly is presumed to arise (Table 1).[2]

1.1.1 | Gastrulation

During gastrulation the ectoderm is established as one of the three germ cell layers; this layer leads to formation of the spinal cord, skin, and many of its adnexae.[3,4]

Defects arising during gastrulation affect all germ cell layers, leading to skeletal and enteric associations (eg, split spinal cord malformations [SCM]) and neurenteric cysts.

1.1.2 | Primary neurulation

Folding of the ectodermal plate is governed by the interaction of molecular, genetic (planar cell polarity genes) and cytoskeletal factors that result in formation of the neural tube and in separation of the ectoderm into its neural and cutaneous components.

Neurulation defects include anomalies involving failure of neural tube closure (myelomeningocele) and separation of the neural from cutaneous ectoderm (dysjunction). The process of dysjunction may occur prematurely or incompletely. With incomplete closure, premature separation

TABLE 1 Classification of spinal dysraphism according to presumed developmental origin[2]

	Developmental stage				
		Primary neurulation			
			Dysjunction		
	Gastrulation	Neural tube closure	Premature	Incomplete	Secondary neurulation
Dysraphism diagnosis	Split cord malformations	Myelomeningocele	Lipoma (dorsal)	Dermal sinus track	Thickened filum
	Neurenteric cysts			Limited dorsal myeloschisis	Lipoma (caudal, transitional and chaotic)
					Terminal myelocystocele

permits formation of a lipoma on the surface of the neural placode from the exposed underlying mesoderm.[5,6]

Anomalies of delayed or incomplete dysjunction refer to a group of dysraphic defects whereby neurulation appears to have taken place, yet persistent connections remain between cutaneous and neural elements.[2] The nomenclature is confusing[7] but it is the nature of the connection that distinguishes each; a predominantly cutaneous lesion is termed dermal sinus track (DST), whereas a connection that is neuroglial is termed limited dorsal myeloschisis (LDM).[8]

1.1.3 | Secondary neurulation

Secondary neurulation leads to formation of the terminal spinal cord (distal to S2), including the conus medullaris, filum terminale, and cauda equina. Cells responsible for secondary neurulation originate in the caudal cell mass of the tail bud, a region that contributes to formation of the lower gastrointestinal and lower genitourinary tracts.

Disorders of secondary neurulation are prone to cause neurogenic sphincter disturbances, and frequently co-exist alongside cloacal and partial urorectal septal malformations (PURSMS) due to their common origin in the caudal cell mass.[5,6,9] Complete absence of secondary neurulation characterizes caudal regression syndrome (sacral dysgensis), the only dysraphic malformation where the spinal cord ends in a higher than normal position and has a blunted appearance. Thickened or fatty filum terminale is the most common and anatomically mildest secondary neurulation anomaly; a more complex variant is the caudal lumbosacral lipoma. Other lipomas include transitional and chaotic types, these also originate during secondary neurulation and represent some of the most anatomically complex dysraphisms. Terminal myelocystocele (TMC) completes the group of secondary neurulation disorders; a severe developmental abnormality of the termination of the spinal cord where the conus balloons into a large cyst-like structure, the cystocele.[10] Associated structural malformations of the lower genitourinary tract are common with terminal myelocystoceles.[10]

1.2 | Prevalence, etiology, and genetics of OSD and associated conditions

Specific incidence and prevalence data are unknown but slight female predominance exists for most OSD. Simple spina bifida occulta (SBO), an incomplete fusion of posterior lumbosacral elements, often discovered incidentally on radiographic imaging, occurs in 25-30% of people; it is inconsequential in asymptomatic patients without other findings.[11] Benign sacral dimples are noted in 2-4% of babies. These are easily distinguished from more rare DSTs, occurring in approximately 1 in 1500-2500 live births.[12] Cadaveric studies demonstrate a 4-6% prevalence of filum terminale lipomas, whereas MRI suggests a prevalence of 0.24-4%.[13] Spinal cord lipomas are detected in 3-8/100 000 births.[9,11] Prevalence of rarer forms of OSD is unknown; split cord malformation is estimated to account for 3-5% of all OSD.[3]

Environmental and nutritional links to OSD are tenuous. Folic acid supplementation is associated with a decreased incidence of imperforate anus and open neural tube defects (MMC), but this has not led to fewer OSD.[9,14] Maternal medical conditions have not been strongly associated with most complex dysraphic anomalies, except caudal regression syndrome's link to maternal diabetes and myelocystocele's link with retinoic acid use.[15]

Recent genetic and twin studies support a genetic basis for OSD.[16] Further evidence indicating a genetic role includes higher prevalence rates of OSD in Hispanics and familial reports of LMM and fatty fila in identical twins.[9,17] Trisomy 21 and chromosomal 22 deletions (TBX1) are associated with OSD.

Several syndromes have genetic links associated with OSD and include VACTERL (Vertebral malformations, Anal atresia, Cardiac anomalies, TE fistula, Renal abnormalities, and Limb defects) and Currarino triad (sacral dysgenesis, presacral mass, anorectal malformation [ARM]). VACTERL is related to deletions of GLI2 and GLI3 and associated with spinal dysraphism in 39%.[16] Currarino triad is linked to a mutation in MNX1 and HLXB9 genes and associated with spinal dysraphism in 60%.[16,18]

1.3 | Pathophysiology of tethered spinal cord

While all dysraphic conditions can be associated with neurologic, urologic, and orthopaedic deficits (sometimes called the neuro-orthopedic syndrome or tethered cord syndrome [TCS]) the term "tethered spinal cord" is inconsistently used and misleading. Animal models of "tethered spinal cord" have shown mechanical traction reduces spinal cord blood flow, leading to impairments in oxidative metabolism.[19,20] These changes in oxygenation are associated with reversible changes in motor evoked potentials, somatosensory potentials, and neurological examinations in animals.[19,20]

Such observations are extrapolated to spinal dysraphism and cited as evidence supporting untethering surgery. However, it would be inappropriate to conclude that "tethering" is the sole, or main mechanism underlying the neuro-orthopedic syndrome. Primary dysplasia of neural tissue, while acknowledged in MMC, is given scant consideration in OSD lesions. In caudal regression syndrome an intrinsic neural dysplasia is self-evident with complete absence of the conus, and in SCMs, where one hemi-cord is less well developed (the ipsi-lateral limb is also smaller). It is probable some element of dysplasia exists in complex lipomas as deficits due to dysplasia are likely irreversible

with treatment. Other mechanisms of neurological deterioration in OSD include mass effect from an enlarging lipoma or intraspinal dermoid, ascending syringomyelia in TMC and mechanical compression from bone abnormalities (diastematomyelia spur in SCM or where there is severe spinal kyphosis due to anomalous segmentation).

In summary, spinal cord tethering may be one component responsible for neurological deterioration that is alleviated by surgery; however, other intrinsic mechanisms (dysplasia, mass effect and mechanical compression) causing deterioration should not be overlooked.

2 | PRESENTING CLINICAL FINDINGS AND EVALUATION METHODS

2.1 | Signs and symptoms of OSD

Presentation of OSD varies widely, from completely asymptomatic despite a cutaneous abnormality to paraplegia with cloacal exstrophy. A detailed description of symptomatology is beyond the scope of this article and descriptions of cutaneous stigmata, and neurologic, orthopedic, and urologic manifestations have been described previously.[3,5,15,21–24]

Lumbosacral skin lesions (dimples, lipomas, gluteal asymmetries, or hairy patches) are common. Neurologic findings and/or foot or limb deformities are infrequent in infancy but increasingly common in undiagnosed children over time. Presentation of OSD is discussed subsequently.

2.2 | Imaging for suspected cases of OSD

Ultrasound is ideal for screening neonates and infants due to its ease of performance, no radiation exposure and low cost. Slow ossification of posterior sacral elements creates a window of opportunity to visualize spinal canal contents up to 3 months of age (Figure 1). Unfortunately, ready access to ultrasound has led to overabundant screening and often, equivocal findings.[25] Inexperienced sonographers may overlook subtle findings (fatty fila),[26,27] but in experienced hands, spinal ultrasonography has high specificity, making it a useful tool in "at risk" cases.[28]

Borderline "low conus" on ultrasound is rarely associated with a pathological abnormality but often leads to further unnecessary testing (spinal MRI).[29] Thakur et al found "borderline low conus" in 12% of 748 infants undergoing sonography for a sacral dimple: none of the 11 undergoing further imaging (MRI or US) had TSC and none underwent surgery.[29]

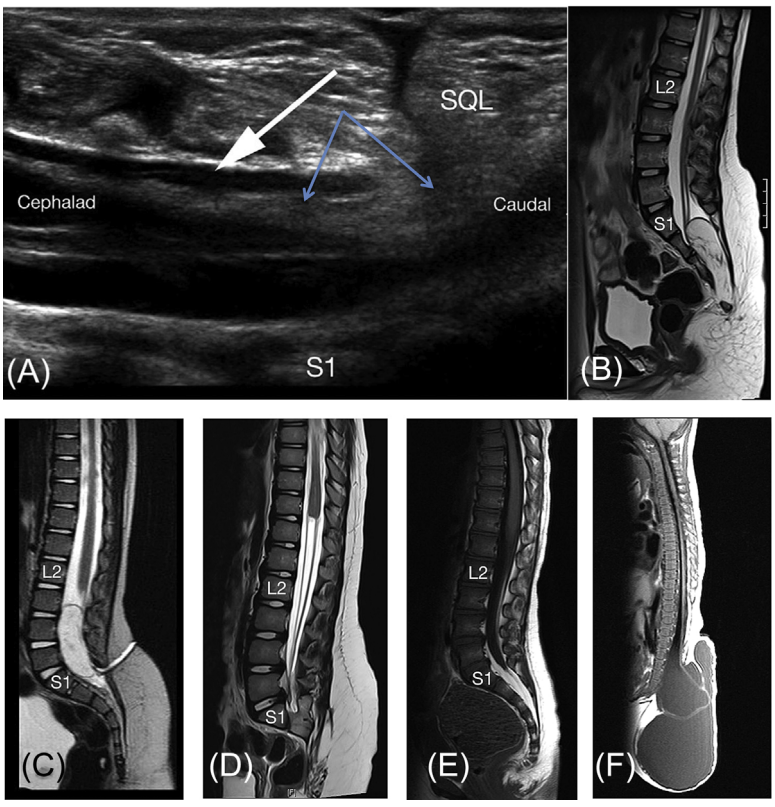

FIGURE 1 Imaging of OSD. (A) Lumbosacral ultrasound demonstrating a subcutaneous lipoma (SQL) connected to an intradural mass (arrow); (B) MRI demonstrating lipomyelomeningocele and low conus; (C) MRI of dermal sinus track with intradural dermoid mass; (D) MRI of caudal regression syndrome with a typical squared appearance of the conus, which is higher than normal; (E) MRI of fatty infiltration of filum terminale; and (F) MRI of myelocystocele

MRI is the modality of choice for evaluating patients with OSD. Because sedation is often required for optimal imaging in infants, MRI is delayed till 3 months of age, as surgery is rarely done shortly after birth, unless specifically indicated (Figure 1).

Most clinicians consider conus termination below the L2 vertebral body abnormal, but radiological determination of the normal conus position is age dependent. Prenatal ascent of the conus, confirmed with MRI, continues for three months after a term birth.[30] The conus terminates below L2 with decreasing frequency during the first 3 months in normal term infants: 13% in the first, 11% in the second, and 5% in the third month of life.[31]

Therefore, conus termination below L2 after 3 months of age should raise suspicion for TSC, but diagnosing TSC solely based on conus level must be juxtaposed to age in preterm infants during early postnatal life. Determining conus level can be complicated by sacralization of lumbar vertebrae resulting in a higher normal conus position or lumbarization of sacral segments leading to lower conus termination.[32]

2.3 | Urologic evaluation of patient with suspected OSD and TCS

Urologic evaluation in OSD is an important component for treatment decisions and the gold standard remains UDS (cystometrography [CMG] and sphincter electromyography [EMG]).[33–36] When carefully performed, UDS yields information about lower urinary tract (LUT) function that guides neurosurgeons' decisions to operate and serves as a basis for comparison of surgical outcomes.[37] CMG reveals bladder reactivity when it is artificially filled and when it empties. Sphincter EMG monitors pelvic floor muscle activity (external urethral sphincter function) in response to filling and emptying of the bladder (Figure 2). Many urologists favor patch electrodes to measure muscle responses but some feel EMG needle electrodes are the most precise way to determine denervation involving the external sphincter and sacral cord reflex function during the micturition cycle, and for comparative assessment.

In infant and pre-toilet-trained children, performance and interpretation of UDS can be challenging, yet oftentimes UDS provides information involving LUT function that can be vital to timely intervention. Detrusor overactivity is the most common UDS finding associated with TCS. However, it may be seen in neurologically normal children with immature or "infantile" bladders as a result of delayed maturation involving inhibition from the pontine micturition center. Characteristic UDS tracings of detrusor overactivity supportive of an underlying neurologic condition include a repetitive, sinusoidal detrusor contraction pattern on CMG or findings of detrusor sphincter dyssynergy (Figure 2). There is limited normative data in this group and prognostic significance of findings (atypical voiding patterns and small post void residuals) is frequently vague. It is imperative urologists and neurosurgeons corroborate the clinical picture with UDS findings. Urgency, urge incontinence, sudden wetting or stress incontinence may be signs of LUT dysfunction; unless specifically characterized, they may result from subtle comorbidities (UTI, constipation, functional behavior disorders) and not OSD.

In addition to bladder dysfunction and urinary incontinence, disturbances of bowel function and rectal continence are frequent in OSD, particularly the severe forms associated with ARM. Bowel dysfunction rarely occurs in isolation and without LUT dysfunction.

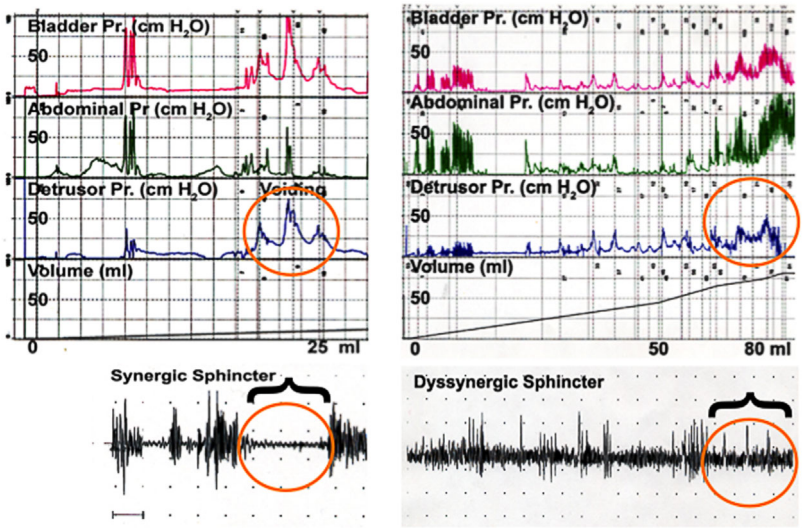

FIGURE 2 Cystometry and sphincter EMG showing synergy (left) and dyssynergy when voiding (right)

2.4 | Other evaluation methods for suspected OSD and TCS

Plain X-rays have no role in screening for OSD. Asymptomatic SBO is common in normal children; it is no more likely when LUT dysfunction of unknown origin is present.[38] Controversy exists whether or not an increasing lumbosacral angle might suggest progressive TSC.[39,40] In utero MRI is sensitive in diagnosing the obvious OSD.[41] Various other supplementary MRI techniques (cine, flexion, extension, supine, and prone) are ineffective diagnostic tools that do not reliably identify good candidates for surgery.[15,42]

Fecoflowmetry and electrophysiological testing (SSEP, MEP) may also detect sub-clinical decline, as deterioration often proceeds at a glacial pace, often imperceptible to patients.[43,44] The specificity of these tests is unclear presently. They are not routine procedures when investigating an OSD.

3 | PATTERNS OF CLINICAL PRESENTATION OF OSD

Affected infants and young children are recognized by cutaneous markers or orthopedic deformities; older children present with bladder and bowel dysfunction, pain or sensorimotor lower extremity deficits; adolescents with scoliosis and/or urinary incontinence.[5] Presenting signs and symptoms, previously categorized by Cochrane, are summarized below.[15]

3.1 | Symptomatic child with bowel and bladder dysfunction/evolving neurologic and orthopaedic abnormalities

When cutaneous markers are inconspicuous or absent, mildly symptomatic OSD are often overlooked until children are old enough to walk and/or expected to have control over bladder and bowel function (Figure 3). Subtle neurologic deficits, foot

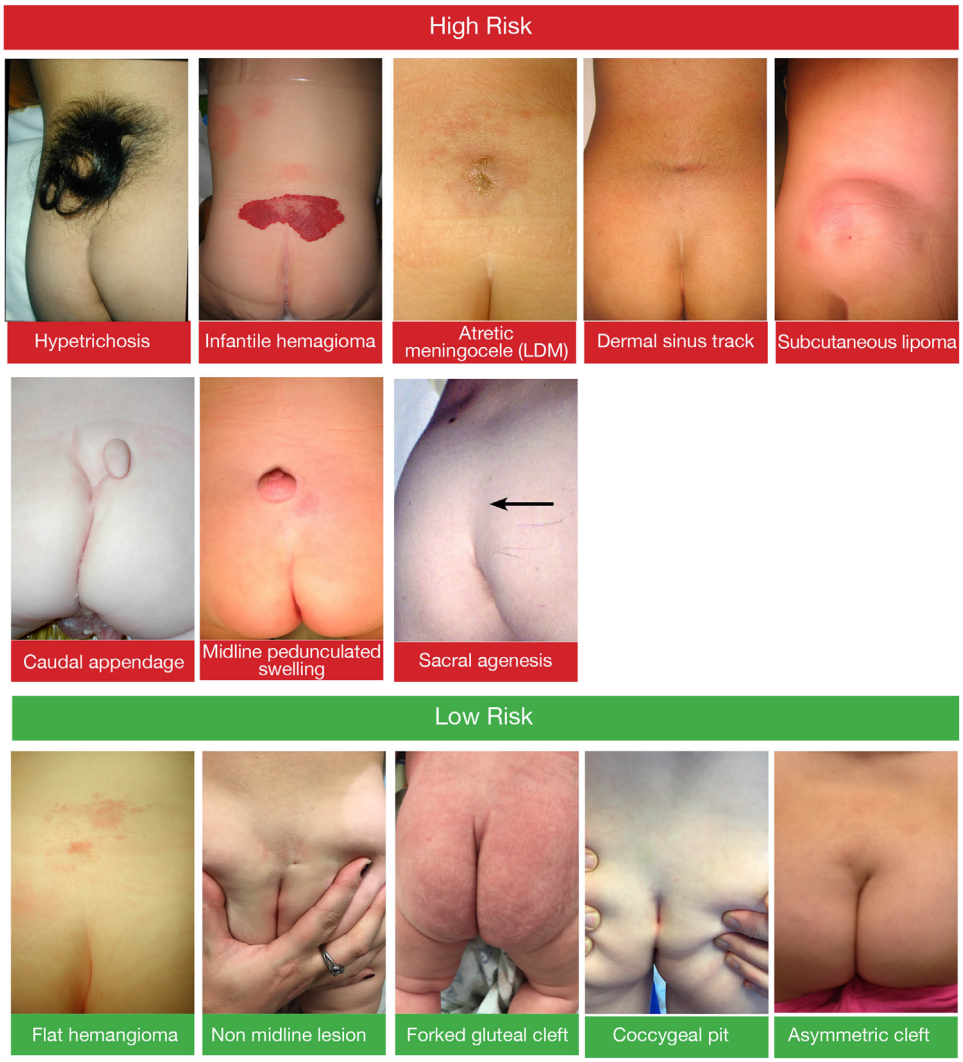

FIGURE 3 Typical lumbosacral cutaneous abnormalities stratified by their risk of association with OSD on MRI, adapted from Dias et al[5]

abnormalities, or impaired bladder and bowel control may only become evident as children grow, mature, and approach developmental milestones.

3.2 | Asymptomatic infant or child with urogenital and anorectal anomalies

Patients with caudal (sacral) agenesis have a variety of anorectal and urogenital abnormalities, from imperforate anus to cloacal exstrophy.[15] These anomalies are often associated with complex conditions—VACTERL association or omphalocele, exstrophy, imperforate anus, spinal defects (OEIS) syndrome.

Infants and children with ARM, PURSM, and cloacal exstrophy frequently have OSD, that correlates with severity of the associated malformation.[15] 10-53% of children with ARM have a spinal dysraphism.[26] Prevalence of a spinal cord anomaly is increased with the more cephalad positioned imperforate anus (above the levator ani muscle). Rarely are cutaneous markers present; this lack should not dissuade clinicians from obtaining MRI in these "high-risk" patients.

Children with sacral agenesis (absence of a variable number of sacral vertebrae) have a characteristic appearance to the lower back—a low gluteal crease with flattened buttocks—but no motor or sensory deficit involving the lower extremities (Figure 3). A sharp cut-off of the spinal cord at T12/ L1 is seen on spinal ultrasound or MRI (Figure 1). Although neurogenic bladder and sphincter dysfunction is present, rarely does progressive neurologic deterioration occur, so neurosurgical intervention is infrequently warranted.

3.3 | Asymptomatic infant with a cutaneous marker

80% of children with OSD have a noticeable cutaneous marker over the lumbosacral region, which can occur in isolation or in combination with other skin abnormalities. Conversely, only 3% of normal neonates have such an abnormality.[15,45] Any lumbosacral cutaneous marker in a newborn should raise suspicion of an OSD;[16] however, diagnostic specificity is unpredictable. Dias et al categorized cutaneous abnormalities according to their risk of OSD.[5] Commonly encountered cutaneous markers are demonstrated in Figure 3, organized according to risk of an OSD on MRI (Figure 3).

Midline abnormalities—"faun's tail," hypertrichosis associated with SCM, dermal appendage, a large midline subcutaneous lipoma, or a hairy midline dimple cephalad to the intergluteal folds, are recognized OSD manifestations. If these cutaneous markers are seen in conjunction with neurological deficits, foot or leg deformities, or neurogenic bladder/bowel dysfunction, the diagnosis warrants MRI confirmation (Figure 3).

The connection of lumbar hemangiomas and OSD is less clear. Large, raised reticulated, midline hemangiomas (telangectatic heamangiomas) require imaging, but miniscule lesions off the midline are normal variants that do not necessitate investigation.[5,46–48] The LUMBAR syndrome (Lower body hemangioma, Urogenital abnormalities, Myelopathy, Bony defects, Anorectal malformations, and Renal anomalies) is the severest form.[49] Midline lumbosacral nevus, flavus simplex (salmon patch) or port wine stain are normal variants, considered low risk for OSD; these necessitate imaging if they are midline and/or if the child has other findings.[5]

When cutaneous markers are found in isolation they can be confused with normal skin findings (stork bite, Mongolian spot). A benign sacrococcygeal dimple raises concerns for potential OSD, leading to inappropriate imaging.[26,27,50] A conical dimple within the gluteal cleft, where the tip of the coccyx is palpated at its base, is characteristic of a benign sacrococcygeal dimple that does not warrant imaging. Of healthy infants with sacrococcygeal dimples undergoing ultrasound screening, only 4 of 3884 had TSC.[25] Spinal cord imaging in "normal" children is recommended if the dimple has atypical features.[16] The importance is distinguishing benign dimples from DST tracks and LDM—the latter occur above the gluteal cleft, with the epithelium thin and the margins irregular (similar to cigarette burn appearance). When coarse hairs, discharge or infection is present MRI is mandatory.

3.4 | Symptomatic child with incontinence and normal MRI (occult tethered cord syndrome)

A normal spinal MRI associated with medically refractory neurogenic bladder dysfunction characterizes the dubious entity of OTCS.[51] Untethering (division of a normal filum) for children with delayed toilet training or secondary incontinence without a recognizable cause has been controversial[51–53] (see below –Group II).

4 | INDICATIONS FOR SURGICAL SPINAL CORD UNTETHERING (SCU)

Since the advent of MRI, a low-lying spinal cord is readily diagnosed in asymptomatic patients or those with fixed deficits. A U.S. inpatient hospital database review highlighted a steady rise in operations performed for "TCS."[54] While recommending surgery is tempting in asymptomatic children with an abnormal MRI or any deficit or deformity, improvement in long-standing, fixed deficits is uncommon after SCU.[55] Surgery is only advocated if observation risks outweigh intervention.[11]

Despite these risks, our understanding of the natural history of most OSDs is incomplete. This gap, combined with lack of randomized controlled trials, has produced controversy regarding indications for surgery for many OSD in asymptomatic or stable children. This debate is poignant because once deficits develop (lower extremity motor weakness, foot deformity, and/or urinary and bowel dysfunction) they are often irreversible following surgery.[24,56–58]

The range of surgical risks for different OSDs varies widely. Until more long-term data are available from well-designed studies, surgeons continue to "walk the fine line between over and under treatment."[59] A pragmatic approach to treatment should be taken, based on level II and level III evidence, and clinical experience.

The importance of accurate diagnostic classification and recognition of variable natural history and surgical complexity in managing OSD must be balanced during the treatment decision process. The indications for neurosurgical intervention can be considered threefold (Table 2):

(i) Group I: When the natural history is known and can be improved by surgical intervention.
- DST with discharge or infection (local abscess or meningitis).
- Type I split cord malformations (particularly where there is coexisting spinal deformity that is likely to require correction)

(ii) Group II: When the natural history might be unpredictable yet the anomaly is known to predispose to neurological injury and intervention can be performed with low risk.
- Risk/stratification analysis of any asymptomatic OSD weighing potential outcomes of observation versus prophylactic surgery.
- Asymptomatic children with a thick, fatty filum and low conus might have a relatively low risk for future deterioration; however, surgery is straightforward with low risk and justifiable on grounds that waiting for a deficit to appear might compromise success of delayed intervention.
- Conversely, an asymptomatic child with a complex lipoma has a higher risk for deterioration yet surgery has a greater risk; thus, an observational policy might be prudent.[11,60]

(iii) Group III: In the presence of new or progressive deterioration (neurological, urological or orthopedic).
- Any OSD patient who develops symptoms or signs during surveillance that can be reasonably attributed to spinal cord tethering should be considered for surgery.

5 | SPECIAL CONSIDERATIONS ACCORDING TO OSD TYPE

5.1 | Thickened or fatty filum

MRI finding of a thickened or fatty filum (>3 mm) occurs in 1.5-5% of the population; it can be associated with TSC, alone or with other forms of OSD.[11,13,61] SCU for thickened or fatty fila carries a low surgical risk, but surgery for radiographic findings must be limited. Most neurosurgeons do not recommend prophylactic untethering for asymptomatic patients with isolated fatty or thickened fila found incidentally, except for the infant or child with an unequivocally low conus or an associated ARM.[62,63] 25-50% of children with ARMs have TSC with a thickened or fat filum, and are at risk for deterioration.[64] Close observation is recommended for isolated lesions, but surgery is reasonable in patients with the potential for continence and when detection of neurologic decline is difficult.[63,65] Informed consent outlining the risks and benefits is essential.

5.2 | Occult tethered cord syndrome (tight filum syndrome)

The presence of urinary incontinence with a normal MRI has been termed occult TCS or tight filum syndrome.

TABLE 2 Indications for spinal cord untethering

Group I: Natural history known, can be improved with SCU	Group II: Asymptomatic, natural history unpredictable, SCU can be performed with acceptable risk	Group III: Progressive deterioration
Neonatal myelomeningocele[a]	Limited dorsal myeloschisis	All symptomatic OSD with structural anomaly on MRI showing new/progressive symptomatology
Type 1 split cord malformation	LMM deemed high risk of deterioration (eg, transitional)	
DST with history of infection/ CSF leak	Terminal myelocystocele	

DST, dermal sinus tract; LMM, lipomyelomeningocele; OSD, occult spinal dysraphism.
[a]While not included under the term OSD, neonatal myelomeningocele repair is included for sake of completeness.

This scenario has been the source of considerable controversy.[63,65]

Advocates for intradural sectioning of the normal filum in these children with secondary incontinence, with or without other clinical findings of TSC (back pain), report improved continence.[51,53,66] Several reviews reported improvements in LUT symptoms after surgery, but limited urodynamic data before and/or after treatment raises questions about objectivity of results.[53] Nogueria et al noted approximately 50% improvement in clinical findings and UDS parameters after filum sectioning in OTCS.[67] UDS may help identify a subgroup that might benefit from surgery, by revealing external sphincter denervation or abnormal detrusor function not evident from clinical assessment. A multidisciplinary evaluation (urologists and neurosurgeons) is imperative to properly select patients who might benefit from surgery.[63,68] 50-75% of those with secondary incontinence from severe bladder dysfunction of unclear etiology improve daytime incontinence after surgery for OTCS.[66,67,69]

Given the controversy surrounding untethering in OTCS, a pilot randomized control trial recently showed no differences between surgical and non-surgical treatment, thus questioning the concept of OTCS.[70] Presently, there is insufficient evidence to support routine use of SCU in OTCS.

5.3 | Complex spinal lipomas (lipomyelomeningocele)

Simple filar lipomas (fatty filum) should be considered separately from complex LMM as they are anatomically and prognostically more straightforward. Complex LMM occurring in 3-8/100 000 births, are a heterogeneous group, subcategorized as dorsal, caudal, transitional and chaotic.[71–73] Their surgical management is controversial.[9,11] Anatomic and phenotypic heterogeneity means classification is problematic and defining symptomatic versus asymptomatic status, particularly in pretoilet trained children presents difficulties making it challenging to draw conclusions from surgery.

Surgery for complex LMM patients, whether symptomatic or not, has been the treatment of choice for decades, based on the belief that inevitable neurological decline outweighs risks of intervention.[63,74–78] From a natural history perspective, the inference that urological decline is worse when the lesion is diagnosed and treated later compared to early management, has prompted early intervention.[56] Although some described improved long-term neurologic and urologic outcomes in children with early surgery compared to those operated later, these studies are limited by selection bias and retrospective methodology.[63,74–78]

Enthusiasm for surgery is often tempered by complexity of the defect because complications can occur intervening in complex abnormalities. Short term, manageable setbacks (CSF leak, delayed wound healing), occur in 5%, with significant neurological compromise that encompasses paralysis and life-changing urologic issues also estimated at 5%.[9,77,79–81] These risks must be balanced against declines without surgery.

SCU is recommended for symptomatic or asymptomatic complex LMM in the US and Asia, but Canadians and Europeans question the utility of prophylactic surgery. Even though no randomized controlled trials exist, retrospective studies have compared long-term outcomes following early surgery to those who underwent untethering only after clinical deterioration; they suggest surgery may not have a protective effect.[11,77]

Studies involving natural history of complex LMM report a risk of functional loss of 40% after 10 years.[11,60] Others report safe and efficacious outcomes with untethering for LMM in the short term but extended follow-up reveals poor outcomes and later deterioration due to re-tethering.[82] When comparing surgery with the natural history, outcomes suggest that conventional untethering has a worse prognosis than observation. Close surveillance for asymptomatic children is warranted if a non-surgical approach is taken.[11] However, once new deficits develop (particularly urological), these deficits are less likely to recover with subsequent surgical intervention.[56,83]

Pang et al reported good results with radical surgery. They hypothesize this lessens the chance of late deterioration.[72,73] In initially asymptomatic cases, 93% remained stable at 16 years.[72,73] This approach is gaining acceptance but the operation is technically demanding and carries greater neurological risk compared with conventional surgery. Time will tell if these results can be replicated to ensure improved late outcomes do not come at the expense of early, surgery-related morbidity.[72,73]

Making general recommendations for neurosurgical management is fraught with difficulty. Initially symptomatic patients or those who develop symptoms of tethering during surveillance should be offered surgery. Currently, no evidence exists that support prophylactic untethering for all lumbosacral lipomas using conventional techniques. Neurosurgeons should be aware of their complication rates and outcomes, and should apprise patients of controversies surrounding management of complex LMM, before advocating surgery in asymptomatic individuals.

Lipomas involving the conus and cauda equina (transitional and chaotic variants) have a greater risk for developing neurological deterioration compared with caudal and dorsal lipomas.[60] Transitional lipomas have high rates of symptomatic re-tethering after conventional surgery,[82] raising the prospect for risk profiling early in management, selecting patients at high risk for deterioration for early radical intervention. Given the complexity and controversies surrounding lumbosacral lipomas, these OSD patients should be managed by a multidisciplinary team.

5.4 | Split cord malformation (SCM) (aka Diastematomyelia)

Two types of SCMs exist: Type I—each hemicord has a separate dural sleeve surrounding it, separated by a central bony spur; and Type II—both hemicords are contained within a single dural sleeve.[4,84] SCMs are exceedingly rare, representing approximately 3.8-5% of all congenital spinal anomalies.[3] Type I is more severe and usually associated with vertebral malformation and deformity; it carries a higher risk of deterioration over time. Surgery for asymptomatic children or those with fixed deficits has been advocated due to progressive neurological decline without SCU. Surgery can be conducted with little risk of new injury.[3,85] Type II SCM is more straightforward, having a benign course; observational follow-up is appropriate in most.

5.5 | Limited dorsal myeloschisis (LDM)

The cutaneous signature of this entity can either be a saccular protuberance over the midline or a flat crater of abnormally epithelialized skin.[8] In both, a fibroneural stalk between the skin and underlying spinal cord exists. Symptoms from TSC can occur, but many have no or minimal symptoms at presentation. SCU and repair can be carried out safely and effectively; neurological and urological prognosis is good. Prophylactic untethering is recommended.

5.6 | Dermal sinus track (DST)

DST lies within the same group of incomplete dysjunction as LDM. A lumen exists within the fibroneural stalk that predisposes to spinal abscess, recurrent meningitis or an intra-spinal dermoid. Given such risks, and low complications from surgery, excision is recommended regardless of symptoms. Neurological and urological outcomes are good following surgery as long as repair is carried out before damage or scarring secondary to infection occurs.[7]

5.7 | Terminal myelocystocele (TMC)

TMC is a rare malformation that is frequently associated with major genitourinary malformations (cloacal exstrophy and OEIS complex).[10] While continence may not be a factor, lower limb neurological deterioration can occur early and precipitously, particularly when expansion of the terminal cyst or associated syringomyelia exists.[86] For these reasons, prophylactic surgery is usually recommended. Surgery is demanding and, as with complex LMM, should be performed by a multidisciplinary team familiar with OSD.

5.8 | Asymptomatic infant or child with urogenital and anorectal anomalies

Where OSD occurs in major urogenital and ARM there may be both structural and neurogenic factors that determine prognosis for bladder/bowel continence. Where there is little or no prospect for developing continence (cloacal exstrophy, high imperforate anal fistulas) neurosurgical objectives must be redefined. A conservative approach might be prudent, reserving untethering only for those with lower extremity motor deficit or pain. It is essential urologists and neurosurgeons collaborate so objectives and patient/family expectations are compatible.

6 | SHORT AND LONG TERM SURGICAL OUTCOMES

6.1 | Urologic outcome

Patients without urologic dysfunction prior to SCU for OSD rarely deteriorate with surgery, but patients with complex LMM are at risk for decline even with normal preoperative LUT function. Urologic recovery (continence and/or UDS improvement) is highly variable, depending on type of OSD, and duration and severity of urologic impairment prior to surgery. Close urologic surveillance is recommended.

In assessing outcomes 10 years after SCU for non-complex lipomas comprising fatty fila and low conus, Frainey et al found unchanged continence for all continent patients before surgery; whereas only 45% of those incontinent preoperatively regained continence.[61] This involved a heterogeneous population, but no factor independently predicted continence after surgery, including age, type of skin lesion, level of conus, presence or absence of hydronephrosis, or vesicoureteral reflux.[61]

Improvement in LUT function for those with neurogenic bladder diagnosed before surgery also vary widely, from 11.5% to 59%, depending on severity of LUT dysfunction before surgery and OSD type.[34,36,37,83,87–89] Outcomes are variable: Children with OTSC having a 50% rate of UDS improvement, SCM improving in 1/3 and complex LMM only 11% improvement.[57,89–92] Infants and young children tend to have better UDS improvement than those undergoing surgery later in childhood.[35,56,77,93]

6.2 | Neurologic outcome

Short-term neurological consequences of SCU also vary widely by OSD type, with sectioning a fatty or tight filum having minimal risk of decline.[88,94] SCU for LMM has a 5-10% chance of neurologic decline, but many deficits are reversible.[9] Symptomatic patients have a greater risk of neurologic decline with later surgery, an argument used to support early intervention in asymptomatic children.

Back pain, lower limb pain and fatigue with ambulation often improve with SCU;[95] long-standing motor and sensory deficits are less likely to show benefit. While the goal of surgery is maintaining function, neurologic improvement does occur when symptoms are of short duration.[96]

6.3 | Spine and deformity outcome

Scoliosis may result from vertebral anomalies associated with OSD or as manifestations of progressive spinal cord dysfunction related to TSC.[5,97] Progressive atypical scoliosis in older children, without a cutaneous lesion, is a common presentation that leads to the diagnosis of OSD. SCU performed before significant curvature development may result in improvement in spinal deformity, particularly before full skeletal maturation.[98] Scoliosis is less likely to progress when SCU is performed before the scoliosis is severe, particularly before the Cobb angle reaches 40 degrees.[99]

6.4 | Need for long-term follow-up

While the short-term outcomes of untethering for complex LMM appear favorable, this group is at risk of late deterioration due to re-tethering. Kulkarni et al noted late deterioration in asymptomatic children, untethered by conventional surgery, more commonly than in those managed expectantly.[100] This underscores the importance of accurate follow up of children with complex LMM and the imperative to develop techniques with more durable long-term benefit.

7 | CLINICAL FOLLOW-UP, WITH OR WITHOUT TETHERED CORD RELEASE

7.1 | Rationale for close monitoring

OSD covers a range of congenital malformations resulting from dysembryogenesis. The anatomical complexity, response to surgery and prognosis varies significantly among these malformations. All have variable potential to cause neurologic or urologic deterioration during growth, compression, syrinx formation or simply the inherent dysplasia of the terminal spinal cord.

Accurate diagnosis and evaluation, acknowledging limitations of defining normal urologic status in the pre-continent child, is an essential starting point in the multidisciplinary management of OSD.

Children with TSC related to OSD should be monitored closely, particularly during periods of rapid and significant changes in height and weight (puberty). All TSC, regardless of whether they have undergone SCU, are at risk for clinical decline. SCU does not eliminate the possibility that deterioration may occur later, even for the simplest form of TSC (fatty filum).[101,102]

Clinical progression after surgery may require further surgery for many OSDs and for variable reasons. Complex LMM are most likely to develop symptomatic re-tethering, recently estimated at 3% per year.[82] This risk underscores the importance of long-term follow-up of complex OSDs.

Although unusual, simple filum sectioning may result in re-tethering. Thickened filum, LDM and DST can be considered neurosurgically stable once continence is established and without any confirmed neurological deficit. Everyone needs to remain vigilant for signs and symptoms indicative of re-tethering.[101,102] It is critical clinicians provide counseling regarding these potential signs and symptoms.

7.2 | Recommended multidisciplinary follow-up

Periodic multidisciplinary evaluations are recommended because manifestations of spinal cord dysfunction can take many forms, including neurologic, urologic, and/or orthopedic compromise. Urologic deterioration is common, particularly in complex LMM.

We recommend monitoring pre-toilet trained children with neurologic and urologic evaluation semiannually. Once continence is established and for older children and adolescents, surveillance is reduced to annual evaluations. Parents should be alert for symptoms and seek medical attention when new onset incontinence, UTI, gait changes, foot or spine deformity, or spinal and lower limb pain occurs.[15,79] A multidisciplinary clinical examination is the most important aspect.

After initial assessment, UDS is warranted in suspicious cases or whenever clinical symptoms support investigation. Follow-up MRI is necessary only when deterioration occurs. Some perform surveillance MRI on clinically stable patients when a syrinx is present, but this is of questionable utility if clinical findings do not suggest deterioration.

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DEFINITION OF COMMON TERMS

OSD (Occult spinal dysraphism): a group of development anomalies of the terminal spinal cord that, although skin covered, that skin may have abnormal characteristics, or stigmata of spinal dysraphism. The OSD disorders are distinct from open spinal dysraphism (spina bifda aperta) where neural tissue is exposed and not skin covered.

TCS (Tethered cord syndrome): a constellation of clinical symptoms or signs, neurological, orthopedic or urological, that may result from spinal dysraphism; sometimes known as the neuro-orthopedic syndrome.

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