Management of urological dysfunction in pediatric patients with spinal dysraphism: review of the literature

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An intact, fully functional spine is the result of a complex sequence of embryological events involving both nervous and musculoskeletal system precursors. Deviations from this highly ordered system can result in congenital abnormalities ranging from clinically insignificant cosmetic changes to CNS malformations that are incompatible with life. Closure of the neural tube, which is believed to be the embryological event gone awry in these cases, is complete by just 28 days’ gestation, often before pregnancy is detected. Although progress has been made to help prevent neural tube defects in the children of those attempting to conceive, these congenital deformities unfortunately continue to affect a startling number of infants worldwide each year. Furthermore, the precise mechanisms governing closure of the neural tube and how they might be interrupted remain elusive. What is known is that there are a large number of individuals who must deal with congenital spine dysraphism and the clinical sequelae on a daily basis. Bladder and urinary dysfunction are frequently encountered, and urological care is a critical, often neglected, component in the lifelong multidisciplinary approach to treatment. Although many treatment strategies have been devised, a need remains for evidence-based interventions, analysis of quality of life, and preemptive education of both caregivers and patients as they grow older. Pediatric neurosurgeons in particular have the unique opportunity to address these issues, often in the first few days of life and throughout pre- and postoperative evaluation. With proper management instituted at birth, many patients could potentially delay or avoid the potential urological complications resulting from congenital neurogenic bladder.

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KEY WORDS  •  spinal dysraphism  •  neural tube defect  •  neurogenic bladder  •  urologic dysfunction

Neural tube defects are among the most common congenital abnormalities that result in significant infant morbidity and mortality, accounting for over 130,000 deaths each year (43% of all neonatal deaths from congenital abnormalities) in regions with low income and high neonatal mortality. Anencephaly and spina bifida, the most prevalent NTDs, occur in approximately 1 in 1000 pregnancies in the US and 300,000 newborns each year worldwide. Myelomeningocele, a form of spina bifida, is one of the most severe CNS malformations compatible with survival. The overall prevalence of closed NTDs that are clinically significant is less clear, since many of these abnormalities are less obvious or hidden at birth. Recently, extensive medical and surgical care has allowed considerable numbers of infants born with myelomeningocele to survive into adulthood, creating a significant neurosurgical population requiring continuous multidisciplinary care. Urological dysfunction is a significant deficit in these children, and neurosurgeons play a significant role in both monitoring and management of these urological concerns.

Review of Spinal Dysraphisms

The clinical spectrum of NTDs includes anencephaly, the group of anomalies considered under the term “spina bifida,” encephalocele, craniorachischisis, and iniencephaly. Of those surviving after birth, and thus those disorders subject to clinical sequelae, the family of defects represent ed under the term spina bifida is by far the most prevalent. Traditionally spina bifida has been synonymous with “spinal dysraphism.” Although spinal dysraphism should in theory only refer to abnormalities of primary neurulation, the term has been used to encompass all congenital spine

Abbreviations used in this paper: CIC = clean intermittent catheterization; EMG = electromyography; NBSD = neurogenic bladder sphincter dysfunction; NTD = neural tube defect; UDS = urodynamic study.
disorders involving problems with differentiation and/or closure of dorsal midline structures. Spinal dysraphisms can be broken down into open or closed forms (Table 1). If neural elements and/or membranes are exposed to the external environment, the defect is classified as an open spinal dysraphism. The most common forms of open spinal dysraphism are myelomeningocele and myelocele, which share a common embryological pathogenesis and are associated with similar clinical implications. Closed spinal dysraphism, on the other hand, consists of spinal abnormalities covered by skin and representing a wide range of NTDs. Although many closed spinal dysraphisms go undetected or remain asymptomatic throughout the life of the patient, there are variants that may be associated with a subcutaneous mass, cord tethering, or more severe spinal cord abnormalities.

Tethered cord is not strictly considered a malformation but rather a clinical syndrome that is usually a complication of myelomeningocele repair (acquired or secondary) or the symptomatic presentation of a form of closed spinal dysraphism (congenital). The position of the conus medullaris is abnormally low (below the L-3 vertebra) and the filum terminale is often thickened. The arterioles, venules, capillaries, and nerve fibers are all subjected to stretching, distortion, and kinking that result in a spectrum of motor and sensory nerve dysfunctions. Many cases of tethered cord syndrome are the result of retethering that occurs after the repair of a myelomeningocele (that is, in 2.8%–32.0% of patients usually between 5 and 9 years of age) or a lipomyelomeningocele, which is frequently associated with progressive neurological and subsequently urological deterioration.

Spinal dysraphisms are associated with a variety of structural abnormalities in addition to the primary defect.

**TABLE 1: Classification of spinal dysraphisms**

<table>
<thead>
<tr>
<th>Classification</th>
<th>Description</th>
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<tbody>
<tr>
<td>open spinal dysraphism</td>
<td></td>
</tr>
<tr>
<td>myelomeningocele</td>
<td>protrusion of spinal cord segment &amp; meninges through a bony defect in midline of the back</td>
</tr>
<tr>
<td>myelocele</td>
<td>myelomeningocele w/ expansion of underlying subarachnoid space</td>
</tr>
<tr>
<td>hemimyelomeningocele</td>
<td>myelomeningocele w/ associated SCM (defect of 1 hemicord)</td>
</tr>
<tr>
<td>hemimyelocele</td>
<td>myelocele w/ associated SCM (defect of 1 hemicord)</td>
</tr>
<tr>
<td>closed spinal dysraphism w/ subcutaneous mass</td>
<td></td>
</tr>
<tr>
<td>lipomyelomeningocele</td>
<td>large subcutaneous lipoma extending intraspinally; cord is tethered &amp; lipoma-cord interface lies outside the spinal canal</td>
</tr>
<tr>
<td>lipomyeloschisis</td>
<td>large subcutaneous lipoma extending intraspinally; cord is tethered and lipoma-cord interface lies w/in or at edge of spinal canal</td>
</tr>
<tr>
<td>terminal myelocystocele</td>
<td>ependyma-lined cyst (dilation of terminal ventricle) bulging through a posterior spina bifida, causing herniation of meninges</td>
</tr>
<tr>
<td>lumbosacral meningocoele</td>
<td>herniation of CSF-filled sac lined by dura &amp; arachnoid through a posterior spina bifida</td>
</tr>
<tr>
<td>cervical myelocystocele</td>
<td>epithelial-lined cavity; only dorsal wall protrudes into meningocele</td>
</tr>
<tr>
<td>cervical myelomeningocele</td>
<td>fibroneurovascular stalk containing neurons, glia, &amp; peripheral nerves traveling through a narrow dorsal dural opening; fans out into lining of meningeal sac; spinal cord remains in canal</td>
</tr>
<tr>
<td>cervical meningocoele</td>
<td>herniation of CSF-filled sac lined by dura &amp; arachnoid through a posterior spina bifida</td>
</tr>
<tr>
<td>closed spinal dysraphism w/o subcutaneous mass</td>
<td></td>
</tr>
<tr>
<td>posterior spina bifida</td>
<td>simple failure of fusion of the posterior vertebra</td>
</tr>
<tr>
<td>intradural &amp; intramedullary lipoma</td>
<td>encapsulated mass w/ fibrous bundles residing in spinal canal</td>
</tr>
<tr>
<td>tight flum terminale</td>
<td>short, hypertrophic flum terminale resulting in tethering &amp; impaired ascent of conus medullaris</td>
</tr>
<tr>
<td>abnormally long spinal cord</td>
<td>absence of normally tapered conus medullaris</td>
</tr>
<tr>
<td>persistent terminal ventricle</td>
<td>cystic dilation of terminal ventricle; an ependyma-lined cavity in conus medullaris</td>
</tr>
<tr>
<td>dorsal enteric fistula</td>
<td>fistula connecting bowel w/ dorsal skin surface, traversing the spinal canal, spinal cord, neural arch, &amp; subcutaneous tissue; involved segment of vertebral column &amp; spinal cord is split around the fistula</td>
</tr>
<tr>
<td>neurenteric cyst</td>
<td>intradural cyst lined by mucin-secreting cuboidal or columnar epithelium that resembles gastrointestinal tract</td>
</tr>
<tr>
<td>split cord malformation</td>
<td>variations of splitting of spinal cord into 2 hemicords</td>
</tr>
<tr>
<td>dermal sinus</td>
<td>epithelium-lined fistula extending into the CNS; point of termination varies</td>
</tr>
<tr>
<td>caudal regression syndrome</td>
<td>family of anomalies representing total or partial agenesis of the spinal column, imperforate anus, genital anomalies, bilateral renal dysplasia/aplasia, &amp; pulmonary hypoplasia</td>
</tr>
<tr>
<td>segmental spinal dysgenesis</td>
<td>segmental agenesis/dysgenesis of lumbar/thoracolumbar spine, segmental abnormality of spinal cord &amp; nerve roots, congenital paraplegia or paraparesis, and congenital lower-limb deformities</td>
</tr>
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of neurulation. The mesoderm that forms the vertebrae is also the precursor for the mesonephros, an embryological structure that represents an important stage in the development of the mature genitourinary system. This association explains the frequency of coexisting anomalies of the urinary tract and congenital spine dysraphisms.33 In a study of infants with urogenital anomalies, it was noted that they had a higher prevalence of closed spinal dysraphism (23 per 1000 vs 1 per 1000).36 Infants in the abnormal group exhibited a consistently lower conus medullaris and thicker filum terminale when compared with healthy infants without anomalies.

**Diagnosis**

Urological dysfunction in children may be present from birth in individuals with open NTDs, or it may present later in life in individuals with closed NTDs, such as lipomyelomeningocele, or in those with a secondary tethered cord. Establishment of a diagnosis generally relies on the children and parents monitoring urinary function to assess for changes such as incontinence, changes in urinary frequency, urinary tract infections, or other alterations in the patient’s baseline urological function. Of importance for all children with spinal dysraphisms is the monitoring of urological function throughout their clinical course, as early detection of worsening urological function combined with timely surgical intervention is important for the prevention of progressive deterioration. A long-term follow-up study of 25 newborns after myelomeningocele and meningocele repair demonstrated the importance of regular follow-up of urinary function, as those individuals younger than 6 years were at higher risk of secondary tethered cord and subsequent urological deterioration.57 In an evaluation of 25 children with spina bifida, Kumar et al.38 demonstrated abnormal baseline urodynamics in 90% of children with open defects and 66.6% of children with closed defects. Because these children may have a baseline degree of urological dysfunction or subclinical urological dysfunction, the neurosurgeon must rely on colleagues in urology, as well as urodynamic function studies, to assess for changes in urological function. Further preoperative evaluation of these children will be discussed.

**Innervation of the Genitourinary System**

Central control of bladder function begins in the locus coeruleus of the pons, which is responsible for a reflex pathway that synchronizes bladder contraction with internal urethral sphincter relaxation during voiding (Fig. 1).40 With appropriate training, cortical signals can voluntarily suppress this reflex. Infants who have not yet acquired this control have an uninhibited pontine reflex. Thus, the detrusor muscle contracts and the internal sphincter relaxes when a critical urine capacity is reached.65 Parasympathetic fibers are the primary innervation of the urinary bladder. Presynaptic fibers arise from neurons in the S2–4 cord segments and travel via pelvic splanchnic nerves and the inferior hypogastric and vesical plexuses to the bladder. They form synapses with postsynaptic neurons that are found on or near the bladder wall. Parasympathetic fibers provide motor innervation to the detrusor muscle and inhibit the internal urethral sphincter. Sympathetic fibers arise from the T11–L3 cord segments, travel via lumbar splanchnic nerves, and synapse on the hypogastric system of plexuses.46 Sympathetic nerves have little role in bladder motor activity, but they do appear to heavily innervate the neck and trigone of the bladder. Sympathetic stimulation allows for bladder neck closure, which is crucial for bladder filling. Somatic fibers to the external urethral sphincter arise from motor neurons in the S2–4 cord segments and travel to the bladder via the pudendal nerve. Although the external sphincter can be contracted voluntarily, it relaxes reflexively when micturition is initiated via cortical signals and the internal sphincter opens.25

**Urological Sequelae**

Most closed spinal dysraphisms or tethered cord syndromes are diagnosed following manifestation of neuro-
logical symptoms or are discovered incidentally during workup for unrelated problems. These neurological sequelae can include muscular deformity, bowel and bladder incontinence, sexual dysfunction, lower-extremity sensory loss, and paraplegia in severe cases. Patients with closed spinal dysraphisms may present with signs and symptoms much more insidious in nature, with the exception of the cutaneous markers that may be associated with 43%–95% of cases, so urological symptoms in these patients may go unnoticed until later in life. New-onset urinary incontinence, changes in voiding patterns, or recurrent urinary tract infections are indications for additional workup. Clinically significant symptoms are usually the result of pathology or abnormalities that cause spinal cord tethering.

The urological sequelae of myelomeningocele, the most common open spinal dysraphism, have been well studied. Urinary symptoms were initially thought to be the result of paresis of the detrusor muscle and/or sphincter muscles. It was not until the development of pediatric urodynamics in the late 1970s and early 1980s that physicians realized that detrusor-sphincter dyssynergia, which creates a functional bladder outlet obstruction, may also be the primary issue. In fact, children with myelomeningocele may show any of 4 unique combinations of detrusor muscle and urethral sphincter activity (detrusor overactivity with sphincter overactivity; detrusor inactivity with sphincter overactivity; detrusor overactivity with sphincter inactivity; and detrusor inactivity with sphincter inactivity). The type of detrusor-sphincter dyssynergia tends to remain consistent during follow-up because it represents the manifestation of the primary neurological lesion. The clinical manifestations include incontinence that results from urethral sphincter inactivity and obstruction, which results from urethral sphincter overactivity. Regardless of the underlying mechanism, the consequences of untreated urological sequelae are severe. Renal failure within the 1st year of life is responsible for up to 20% of deaths. Renal damage begins as early as 6 months of age, and up to 100% of children with untreated dyssynergia have renal scarring. Complications of poor urological management occur in a progressive fashion. Urinary stasis, combined with vesicoureteral reflux, leads to an increased incidence of infection of the urinary tract, bladder, and kidney, as well as an increased risk of urinary tract calculi. Elevated bladder pressure can lead to hydronephrosis, renal deterioration, renal failure, and ultimately death. Furthermore, the incidence of bladder cancer is significantly elevated, sexual dysfunction is common, and there is often physical damage due to long-term catheterization. With proper treatment and patient compliance, physicians can now ensure preservation of renal function provided that interventions begin immediately after birth. Quality of life improvements continue to be explored to address the social implications of management strategies.

**Urodynamic Testing**

Urodynamic studies provide a reliable way to evaluate the function of the lower urinary tract and can be adapted specifically for use in children. Significant controversy exists about the use of invasive urodynamic testing in children with nonneurogenic voiding disorders because these children may often be evaluated with noninvasive UDSs and managed expectantly with behavioral and pharmacological therapy. However, identification or confirmation of neurogenic bladder is an excellent use of UDSs, providing more specific and accurate information than imaging, neurological examination, and symptomatology alone. Noninvasive UDSs consist of uroflowmetry and postvoid residual measurement. Uroflowmetry can be used in isolation, simply measuring the child’s ability to empty the bladder, or as part of more comprehensive studies. In children, the flow pattern is more useful than the rate of flow, as it can provide information about detrusor function, amount of outlet resistance, and degree of external urethral sphincter activity. Postvoid residual measurements may be performed using ultrasonography or specialized bladder scanners, and abnormal values indicate a need for repeated measurements or further workup. Jansson et al. found that the median postvoid residual in 6-month old infants was 5 ml (r = 0–22), decreasing to a median of 0 ml at age 3 (r = 0–18).

If noninvasive UDSs show abnormal bladder function, either through postvoid residual measurement or bladder enlargement, invasive studies are warranted, including cystometrography, in which rectal and urethral catheters measure intraabdominal and intravesical pressures. Perineal EMG patches may be added to assess external urethral sphincter activity. To perform the test, the bladder is filled using a multilumen catheter until the child feels a strong urge to urinate, becomes uncomfortable, urination occurs, detrusor pressure exceeds 40 mm H2O, greater than 150% of expected capacity has been infused, or the leakage rate exceeds the infusion rate. Fluoroscopy can be used during filling-phase studies to visualize the shape and contours of the bladder while full or voiding, as well as intrinsic sphincter deficiency in patients with neurogenic bladder disease and dynamic dysfunction. Evaluation of the storage function of the bladder is typically described subjectively in terms of bladder sensation (as well as objective values such as compliance, activity, and capacity) and, therefore, is only of use in patients able to describe what they are feeling. In younger patients, however, signs such as toe curling or abdominal tensing may be indicative of a full bladder, signifying incipient micturation. Bladder capacity and compliance are then determined, although there are no standardized reference values available for compliance in children or adults; this is further complicated in that compliance varies according to capacity at the time of measurement and in that capacity does not change linearly with age. Finally, in children capable of spontaneous voiding, voiding-phase studies measure voiding pressure and flow rate, with attention paid to whether voiding pressure has been maintained throughout the duration of bladder contraction until completely empty, as well as any external urethral sphincter activity, monitored through EMG. As in filling studies, concurrent imaging of the bladder improves diagnostic interpretation and characterization of any pathology.
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Current Strategies for Management of Urological Dysfunction

Prompt detection of children with high-risk urological dysfunction and early intervention is key in protecting against hydronephrosis, preserving renal function and preventing the development of a poorly compliant bladder. Neurogenic bladder sphincter dysfunction can cause these problems long before continence becomes an issue. There are several types of NBSD seen clinically, consisting of the previously mentioned combinations of detrusor over- and underactivity and sphincter over- and underactivity. Management is determined by the consequences of the particular combination. Of all four types, only the combination of sphincter underactivity and detrusor underactivity is inherently “safe” in that, untreated, it will not lead to urinary tract damage, although it does result in incontinence and increased rates of urinary tract infection.62

All of the other forms of NBSD include CIC to empty the bladder. In the case of sphincter overactivity with detrusor underactivity, it is the only management needed to control urinary tract damage and leakage. With proper parental/caregiver education, and with the practice of proper basic technique, including clean and atraumatic catheter application, the specific techniques and materials used are unimportant with regard to efficacy.62 It should be noted, however, that Credé voiding is not recommended for noncatheterized patients because it can result in compression of the external urethral sphincter, causing high pressure voiding or reflux in some patients, which is detrimental to the upper urinary tract.14

Beginning CIC early in infancy conveys several advantages over expectant treatment, including easier caregiver and patient adaptation to the routine and less need for augmentation cystoplasty.20,30,66 As the children age and develop manual dexterity, they may be transitioned to clean intermittent self-catheterization.62

In addition to CIC, pharmacological treatment in the form of the anticholinergic oxybutynin is used to treat both forms of NBSD involving detrusor overactivity, long-term studies having demonstrated safety in children and infants and efficacy in lowering filling pressure, increasing bladder capacity, and preventing renal damage.62 Pharmacological treatment can be administered orally or as an intravesical preparation in children with significant side effects or a poor response to the oral form.12 The combined therapy of CIC and oxybutynin is successful in treating 90% of patients with NBSD, but for those patients in whom this therapy is unsuccessful, other drugs such as propiverine or tolterodine are available, but there is relatively little published information on efficacy in children.62 Recent work has indicated that injection of botulinum A toxin may be a viable treatment in children unresponsive to CIC and oxybutynin.39

After a treatment regimen is chosen, follow-up to assess for efficacy is needed. Assessment can include ultrasound, US, cystography, urinalysis, or other tests. The frequency of visits can be tapered from 3 per year in children up to age 3, 2 per year in school-age children, and annually in adults.62

Several urological procedures exist for the purpose of increasing bladder capacity and for urinary diversion. These procedures include ureterostomy, vesicostomy, ileal conduit, and ileovesicostomy. Although these procedures are outside the scope of the present review, they are well described in the urology literature.20,63,64

Neurosurgical Intervention

The particular role of the neurosurgeon in the management of urological dysfunction revolves around the preoperative evaluation of children with urological complaints, intraoperative techniques to release a tethered cord and limit neural injury during surgery, and postoperative monitoring for further deterioration. Newer techniques are emerging to reinnervate the bladder. A strategy for management during each of these stages follows.

Preoperative evaluation of any child with a primary urological complaint of possible neurogenic origin should include formal urological assessment, which may include a bladder ultrasound or invasive urodynamic testing as previously described. Additionally, imaging tests such as an MRI may reveal evidence of a closed defect or tethered cord as the likely cause. As many children may be referred to a neurosurgeon after advanced imaging studies have already been completed, the referral to a urologist and correlation of symptoms with results of a urological assessment should follow. This becomes especially important in children with secondary tethered cord in whom there are radiographic concerns of tethering even in the absence of clear symptoms.4,43 Often in these patients the presence of symptoms rather than radiographic findings should take precedence. Suspicion of a symptomatic tethered cord should be raised when there changes in a child’s baseline urinary pattern because these changes may represent secondary tethered cord or development of syringomyelia.

Additional testing such as neurophysiological evaluation may be of assistance in confirming neurological dysfunction. Torre et al.39 undertook electrophysiological testing in 28 children with spinal dysraphisms and found that a combination of electrophysiological tests yielded good predictive results. Combined results from EMG and perineal evoked potentials monitoring predicted vesico-sphincter dyssynergia with 100% sensitivity and bladder dysmotility with 86% sensitivity. Perineal evoked potentials alone detected urodynamic dysfunction with 90% specificity, and EMG combined with perineal evoked potentials confirmed urological dysfunction with 79% sensitivity, yet a negative predictive value of 90%.

Surgical intervention can be divided into 2 categories—primary surgery and secondary release of tethered cord—with an overarching surgical goal of releasing the tethered cord to prevent further deterioration in urological function. For open NTDs, the surgical technique involves releasing the edges of the neural placode, reapproximating the pial edges of the placode to reform the neural tube, and creating a CSF-filled space within the dural closure. Additionally, any existing filum may be transected at the time of initial surgery. For closed NTDs such as lipomyelomeningoceles, the area of tether,
nervation (Fig. 2).\(^2\) Assessing bladder function integrity because of similar innervation of the anal sphincter by muscle groups and sensory distributions as well as the rectal size may be prohibitive to this monitoring strategy.

Recently, prenatal intrauterine repair has been receiving increased attention. The Management of Myelomeningocele Study (MOMS) was conducted to specifically address whether this strategy had improved outcomes compared with traditional postnatal surgery, but urological function in these patients was not assessed.\(^3\) Other studies with long-term outcomes of intrauterine myelomeningocele repair have shown a range of urological outcomes. A long-term follow-up study of 28 patients who underwent fetal myelomeningocele closure demonstrated a range of urological dysfunctions in this subset (decreased bladder capacity, detrusor overactivity, and increased detrusor pressure) and no significant difference compared with individuals who underwent traditional postnatal repair.\(^3\) However, a limited number of studies have also shown an improvement in urodynamic outcome, but there are minimal data to substantiate this.\(^3\) Given the evolving expertise in fetal myelomeningocele closure, further investigation into the urological outcomes after intrauterine repair should be forthcoming.

Neurophysiological monitoring may be used to assist the surgeon with intraoperative planning by alerting the surgeon to changes in monitoring parameters during surgery to reduce the risk of neurological injury and iatrogenic worsening of urological function.\(^3\) Total intravenous anesthesia and minimal use of muscle relaxants facilitate the use of somatosensory evoked potentials, motor evoked potentials, and EMG, both free run and stimulated.\(^4,28\) Monitoring frequently includes lower-extremity muscle groups and sensory distributions as well as the anal sphincter. The innervation of the anal sphincter by the lower sacral nerves (S2–5) is a good proxy for assessing bladder function integrity because of similar innervation (Fig. 2).\(^2\) It is possible to monitor the external urethral sphincter by way of a special ring electrode on a Foley catheter.\(^2\) However, in a pediatric population, urethral size may be prohibitive to this monitoring strategy. The combination of free-run and stimulated EMG allows for continuous EMG monitoring and assessment of structures during dissection, which may be beneficial when encountering dysmorphic neural elements.\(^3,4,37\)

The risk of a retethering in patients with spinal dysraphism may be as low as 3% or as high as 40%, occurring most frequently in patients who underwent primary repair of myelomeningocele and resection of lipomyelomeningocele.\(^3\) Given this elevated risk for future neurological decline, postoperative monitoring of urological function is critical. This includes monitoring of urological function by patients and their family, most importantly noting changes in urological function, especially in the population in which a baseline degree of function requires catheterization. Symptoms may include episodes of incontinence or changes in urinary habits. These patients should be followed up by a urologist for urodynamic evaluation, especially during periods of growth when a patient’s cord is at greatest risk of injury secondary to a tether. Similar to preoperative assessment, any concern about a change in urinary function should prompt urgent evaluation, either urodynamic testing or neurophysiological testing.

**Neurosurgical Outcomes**

The initial goals for neurosurgical repair in open spinal dysraphism include elimination of CSF leakage, prevention of infection, preservation of neural function, and prevention of secondary tethering postoperatively. Once definitive repair is completed, the focus generally shifts to lifestyle improvements. Bowel and bladder dysfunction remains a primary morbidity concern as bladder dysfunction predisposes this group to the risks of recurrent urinary tract infections, pyelonephritis, hydronephrosis, and, in severe cases, renal failure. Given that the primary repair of myelomeningocele often occurs within the first few days of life, when no baseline urological function is known, the outcomes of interest generally arise after a patient presents with worsening urological function due to a tethered cord.

Initial surgical intervention for spina bifida has been shown to result in some improvement in urological function. In the previously mentioned study by Kumar et al.,\(^3\) 42.8% of patients with open defects and 46.2% of those with closed defects experienced improvement in urological function after an initial surgical intervention. Wu and colleagues\(^6\) reviewed data obtained in 43 patients who underwent early surgery for lipomyelomeningocele prior to manifestation of urological dysfunction; they found that 84% of the patients maintained stable function. Macejko et al.,\(^4\) however, reviewed the cases of 79 patients who underwent primary release of a tethered cord inclusive of closed defects and demonstrated that those with lipomyelomeningocele were at risk for the poorest urological outcomes after surgery. In contrast, evaluation of 36 patients with occult tethered spinal cord who underwent sectioning of the filum terminale demonstrated improvement in urinary symptoms in 72% and resolution of incontinence in 42%.\(^4\)

Surgical intervention for tethered cord manifesting with worsening urological function also demonstrates a wide range of improvement in urological function after surgery. Tarcan and associates\(^8\) reported on 36 patients with secondary tethering after repair of myelomeningocele and evaluated outcomes after a detethering procedure. They tiered the outcomes according to the initial grade of urinary tract dilation. Individuals with Grade 1–2 dilation had an improvement in grade in 40% of the cases and complete resolution of the dilation in 33.3%. Those with Grade 3–4 dilation had an improvement in 50% of the cases and resolution in none. However, more profound improvement was noted in the degree of vesico-
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Fig. 2. Intraoperative EMG tracing obtained during a tethered cord release demonstrating train activity of the anal sphincter that can be noted with manipulation of the lumbosacral nerve roots. From top to bottom: left iliopsoas/adductor, left quadriceps, left anterior tibialis, left gastrocnemius, right iliopsoas/adductor, right quadriceps, right anterior tibialis, right gastrocnemius, left anal sphincter, right anal sphincter.

ureteral reflux, which resolved in 62.5% of patients with Grade 1–3 reflux and improved in 37.5%. Patients with Grade 4–5 reflux demonstrated resolution in 33.3% and improvement in 40%. These results were most significant in children treated before the age of 7 years. In a review of 20 patients who underwent surgery for various manifestations of tethered cord, Abrahamsen et al. demonstrated that in all patients with deterioration in urodynamic function prior to untethering function improved after surgery, and in 90% with stable urological function prior to surgery function, remained stable. Dushi and coworkers introduced a urodynamic score as a means of assessing outcome and demonstrated that the postoperative score improved in 55% of patients who presented with urological symptoms and in 86% of those who did not present with urological symptoms. As previously mentioned, these positive outcomes may be subclinical. Palmer et al. reviewed the cases of 20 children who initially presented with worsening scoliosis or lower-extremity function and no discernible change in their urological function; they found that even within this group, 75% of the patients had improved urodynamic function after tethered cord release. These results are not limited to the first untethering procedure, as Maher et al. demonstrated improvement in urological function in 71% of patients who underwent multiple repeat untethering operations.

Conclusions

Spinal dysraphism and other forms of NTDs continue to be significant sources of morbidity and mortality worldwide. Until the 1950s, the probability that an infant with a severe abnormality, such as myelomeningocele, would survive to adulthood was low. Therapeutic advancements and improvement in neurosurgical technique have drastically changed the nature of these congenital malformations. In fact, adults with myelomeningocele now account for greater health care expenditures and more hospital admissions than children with this condition. Thus, the focus of treatment is rapidly shifting toward early recognition and surgical intervention, postoperative monitoring and management, and improvements in patient quality of life.

Perhaps the most critical consideration is the lifelong bladder and urinary dysfunction encountered by these patients. A wide range of clinical pathology can lead to damage of nerves of the genitourinary system, often as a result of spinal cord tethering. Without treatment, the urological sequelae are debilitating and often fatal. For the neurosurgeon involved in the care of such patients, early and frequent urodynamic testing, combined with a multidisciplinary approach involving urologists to guide management, is crucial to improve outcomes. Early detection and surgery for worsening urological dysfunction remains the mainstay for treatment of primary and secondary tethered cord to minimize the long-term effects of irreversible urological dysfunction. Although recent trials have shown promising results with prenatal neurosurgical intervention, it remains to be seen whether these techniques result in consistent favorable urological outcomes. Increasing awareness among neurosurgeons who encounter spinal dysraphisms is one of the first steps in lifelong management that can both drastically improve quality of life and lower future health expenditures.
Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following: Conception and design: Tomei. Acquisition of data: Amarante, Shremel. Drafting the article: Amarante, Shremel, Tomei. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Study supervision: Gandhi, Tomei, Carmel.

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