

## **Surgical treatment of the lipomeningocele in children (a review)**

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*Lipomeningocele is a variant of a latent form of spinal dysraphism, in which during embryogenesis there is no complete separation between the neuroectoderm and mesenchymal elements that form the spine and soft tissues around the neural tube. The frequency of occurrence is 1:4000 newborns, which is 60% of all spinal cord lipomas. Depending on the anatomical relationship of lipomas with the spinal cord, the following forms are distinguished: dorsal lipoma, caudal lipoma, transitional lipoma. In relation to the dura mater and the spinal canal, they are divided into intradural, intra, and extradural. Treatment strategies vary depending on the subtype, neurological deficit, and surgical techniques.*

*This article is a review of the literature on the evaluation and results of surgical treatment of lipomeningocele in children.*

**Key words:** lipomeningocele, children, surgical treatment, review.

## **Хирургическое лечение липоменингоцеле у детей (Обзор литературы)**

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*Липоменингоцеле — вариант скрытой формы спинального дизрафизма, при котором в процессе эмбриогенеза не происходит полного разделения между нейроэктодермой и мезенхимальными элементами, формирующими позвоночник и мягкие ткани вокруг нервной трубки. Частота встречаемости составляет 1:4000 новорождённых, что составляет 60% от всех липом спинного мозга. В зависимости от анатомических взаимоотношений липом со спинным мозгом выделяют следующие формы: дорзальная липома, каудальная липома, переходная липома. По отношению к твёрдой мозговой оболочке и позвоночного канала разделяют на интрадуральную, интра-,экстрадуральную. Стратегии лечения варьируются в зависимости от подтипа, неврологического дефицита и методов операции.*

*Данная статья представляет собой обзор литературы посвященной оценкам и результатам хирургического лечения липоменингоцеле у детей.*

**Ключевые слова:** липоменингоцеле, дети, хирургическое лечение, обзор.

Lipomeningocele is a congenital lesion associated with spina bifida. This lesion is associated with a caudally descended conus infiltrated with fat which continues through the dural, bony and fascial defects to emerge as a subcutaneous mass [1-3]. It can be associated with other congenital anomalies such as sacral agenesis, imperforate anus and ectrophy of the bladder [4, 5].

It represents a complex disorder that may present with neurological deficits secondary to the inherent tethered cord. Treatment strategies vary with subtype, neurological function, and goals of surgery. This paper presents a review of lipomeningocele including embryology, evaluation, treatments, and outcomes.

Lipomeningocele represents a rare but complex neurological disorder that may present with neurological deterioration secondary to an inherent tethered spinal cord. Radiological testing is beneficial in determining the morphology

of the malformation. Specialized testing such as urodynamic studies and neurophysiological testing may be beneficial in assessing for neurological dysfunction secondary to the lipomeningocele. Early surgical intervention may be beneficial in preventing further neurological decline.

### **Embryology**

The pathology of congenital spine and spinal cord defects is best understood through knowledge of embryological development. Central nervous system development initiates in the third week in a process known as neurulation. During primary neurulation, the ectoderm overlying the notochord proliferates, forming the neural plate. The lateral edges of the neural plate soon elevate to form the neural folds. As development continues, the neural folds continue to elevate and approach each other in the midline, fusing to form the neural tube. This fusion begins in the cervical region and proceeds in both the cephalic and caudal directions [6,7]. Secondary

neurulation is the process of development of the caudal cell mass that forms the caudal-most portion of the neural tube, forming the spinal segments below L2. Following neural tube closure, the epithelial ectoderm separates from the neural ectoderm, a process known as disjunction. The epithelial layers fuse to create skin covering the neural tube, and mesenchyme migrates between the neural tube and skin to form the meninges, neural arches of the vertebrae, and paraspinous muscles [8]. In the third month of development, the spinal cord extends the entire length of the embryo. However, as development continues, the vertebral column and dura lengthen more rapidly than the neural tube, and the terminal end of the spinal cord shifts to a higher vertebral level [7]. In a whole-spine imaging study by Kesler et al., [9] the conus medullaris in all 100 children studied terminated between the lower third of T12 and the middle of L2, with the mean level at the lower third of L1 and a mode at the L1-2 disc space. No child presented with the conus medullaris below the middle third of L2. Pinto et al. [10] observed that the inferior tip of the conus medullaris resides at or above the L2 level in 95,12% of the sample studied, with the greatest number (41,5%) at the L1 level. While there is normal variation in the vertebral level of termination, the “adult” level is reached approximately 2 months postnatally [11]. If there are abnormalities in any of these developmental stages, a spinal dysraphism, or defect in closure of the neural tube, can result.

Spinal dysraphisms can be classified as either open or closed dysraphisms. Open spinal dysraphisms include meningocele, myelomeningocele, myeloschisis, encephalocele, and anencephaly. All involve exposure of nervous tissue and/or meninges to the external environment. Closed spinal dysraphisms such as lipomeningocele, lipomyelomeningocele, diastematomyelia, and spina bifida occulta have no exposed neural tissue and are accompanied by cutaneous markers in 43%–95% of cases [12,13,14,15,16,17,18,19] and include lesions such as subcutaneous masses, capillary hemangioma, dimples, and hairy nevus [20,21]. These cutaneous markers may present with closed spinal dysraphisms because of the chronological association of neural tube closure with separation of neural and epithelial ectoderm during embryological development [20]. These cutaneous markers can be used to recognize cases in an asymptomatic neonate. Guggisberg et al. [14] suggested that a combination of 2 or more congenital midline skin lesions is the strongest marker of closed spinal dysraphism. Kriss and Desai [17] observed that only atypical dimples were found to be associated with a high risk for spinal dysraphism, characterized by high placement on the back (> 2,5 cm from the anus), large size (> 5 mm), and appearance in combination with other lesions. Other high-risk cutaneous markers were raised lesions such as tails, masses, hairy patches, hemangiomas, and the presence of multiple skin lesions.

Several types of closed spinal dysraphisms result from embryological abnormalities during primary neurulation. Those that arise from premature disjunction result in fusion of the spinal cord with fatty elements, the most common of which is a lipomeningocele [21]. When premature disjunction occurs, the epithelial ectoderm detaches prematurely from the neural ectoderm, allowing mesenchyme to contact

the inner portion of the developing neural tube [22]. The mesenchyme is induced by the dorsal surface of the closing neural tube to form fat, and this prevents proper neurulation. The extent of the fatty tissue is limited laterally by the neural ridge because the ventral surface of the neural plate induces the mesenchyme to form meninges. This results in a junction between meninges and fat at the neural ridge, and thus the lipoma extends posteriorly through the meningeal and bony defect and into subcutaneous tissues in the extradural space. The neural placode-lipoma interface, which is the connection between the spinal cord and the lipoma, can lie outside of, within, or at the edge of the spinal canal. In contrast to a lipomyelocele where the neural placode-lipoma interface is located within or at the edge of the spinal canal, lipomeningocele is characterized by a placode-lipoma interface located outside the spinal canal [19].

Tethered cord is inherently associated with lipomeningocele as the lipoma tethers the cord to the adjacent dura and soft tissue. Syringomyelia may occur in 20%–25% of patients with a tethered cord [23]. Lipomeningocele can also be associated with other abnormalities. In a study of 97 patients with lipomeningocele, Hoffman et al. [24] reported an association with genitourinary tract anomalies (4,1%), split cord malformations (3,1%), associated dermal sinuses (3,1%), dermoid or epidermoid cysts (3,1%), diastematomyelia (3,1%), terminal hydromyelia (3,1%), anal stenosis (1,0%), and Down syndrome (1,0%). Kanev et al. [15] also reported associated anomalies in a series of 80 patients with lipomeningocele, including scoliosis (8,75%), amniotic band extremity deformity (7,5%), sacral dysgenesis (5,0%), anterior anal displacement with stenosis (2,5%), and hydromyelia (2,5%). There is an increased incidence of Chiari malformation Type I in patients with lipomeningocele as compared with the general population, with 13% of a 54-patient series with Chiari malformation also having lipomeningocele [25].

#### Epidemiology

According to WHO materials, an average of 4-5 children out of 10,000 babies are born with this congenital abnormality. The prevalence of spinal hernia is maximal in northern China, eastern Ireland, Norway and Egypt and minimal in Eastern Europe and the USA (0.1-0.6 observations per 1000 born). The first indications of tumors of the lumbar region similar to spinal hernias are found as early as 460–370 bc [26].

The prevalence of lipomeningocele has been found to range between 0.3 and 0.6 per 10,000 live births [27,28,29,30]. Agopian et al. [27] observed 14.4% of spina bifida cases were lipomeningoceles during examination of the prevalence of spina bifida subtypes.

In general, neural tube defects have a complex origin in which both environmental and genetic factors play a role. There is substantial evidence that maternal dietary folic acid supplementation can reduce the risk of neural tube defects among offspring [31]. However, studies [28,29,30] have reported no reduction in rates of lipomeningocele following folic acid fortification, suggesting that the pathogenesis of lipomeningocele is fundamentally different from that of other neural tube defects. Forrester and Merz [29] also reported higher lipomeningocele rates in infants born to mothers in younger and older age groups. Spinal dysraphisms have been found to

be more common among Hispanics, and less common among non-Hispanic blacks than non-Hispanic whites [27]. Compared with non-Hispanic whites, Hispanics had an even higher prevalence of lipomeningocele than of myelomeningocele, meningocele, and myelocele subtypes of spinal dysraphisms [27]. Maternal obesity has been associated with an increased risk of neural tube defects, suggesting that the embryonic environment for development may be metabolically different for obese women, [32,33] but no particular spinal dysraphism phenotype was observed to explain the overall increased risk [32]. Familial forms of lipomeningocele are rare, with only 2 previous reports [14]. A report of lumbar lipomeningocele and sacrococcygeal teratoma in siblings suggested an inherited regional tendency to developmental error affecting the caudal embryonic segments [34].

#### **Diagnosis and Presentation**

Before the first year of life in newborn infants, ultrasound diagnosis of lipomeningocele makes it possible to determine the state of the spinal cord (CDC, compression by lipomas), since the ossification of the vertebral arches is completed only by the end of 1 year of life. The research of the authors shows that knowledge of the normal echographic anatomy, as well as the use of modern ultrasound devices, allows us to timely diagnose various anomalies of the development of the spinal cord and spine [35]. The diagnosis of a lipomeningocele largely depends on an understanding of the pathology. Lipomeningocele is characterized by a subcutaneous lipoma that is generally located in the lumbar or sacral region [6]. The subcutaneous lipoma extends through a defect in the lumbodorsal fascia, vertebral neural arch, and dura, attaching to an elongated and tethered spinal cord [24]. The most common presenting symptom is a fatty mass positioned in the midline or just off the midline in the lumbosacral region. Additionally, the majority present with other skin lesions associated with the lipoma, including a hairy nevus, skin dimples, and cutaneous hemangiomas [24,15,36]. Because the fatty mass is clinically apparent at birth, those affected are generally diagnosed before neurological symptoms present, and as many as 48% have been found to be neurologically intact on initial diagnosis [6,8].

Three categories of lipomeningocele exist, based on the relative anatomy of the lipoma and neural components: dorsal, transitional, and caudal. The dorsal-type lipomas have an area of attachment to the dorsal spinal cord at the area of myeloschisis in the lower lumbar or lumbosacral levels of the spinal cord and are continuous with the subcutaneous tissue. The lipoma passes through a fascial defect, and may extend into and expand the central canal. A dural defect is present and the placode-lipoma interface may lie in the extradural space. Transitional lipomas have an attachment that extends beyond the area of myeloschisis down to the conus, with a less distinct lipoma-cord interface. The lipoma again extends through a dural defect. The caudal-type lipomas arise predominantly from the caudal end of the conus medullaris. These lipomas may extend through a dural defect or may be encased in the dura [37, 38].

As the subcutaneous lipoma is restricted by the defect in the lumbodorsal fascia, the upward movement of the conus medullaris during axial growth may be limited and

thus may lead to progressive neurological and urinary deficits, the sequelae of a tethered cord [39]. Loss of neurological function has been found to increase with age because of progressive conus tethering and injury to nervous tissue. Loss of neurological function has also been demonstrated to have a logarithmic association with increasing patient age and is believed to be secondary to increased stretch on the spinal cord with axial growth spurts [40,24,41,42]. Other theories regarding the mechanism of progression of symptoms include decreased perfusion secondary to stretch on the spinal cord, increased mass effect from progressive deposition of fat, and stretching effects on the spinal cord [43,42,44]. Hoffman et al. [24] observed that 62,5% of patients were neurologically asymptomatic prior to 6 months of age, while only 29,3% were asymptomatic after 6 months of age. Furthermore, Koyanagi et al. [45] reported progressive neurological symptoms with age in patients with tethered cord, with no children remaining asymptomatic after age 5. A series of 80 patients reported by Kanev et al. [15] demonstrated that bowel and bladder function deteriorates prior to motor function or sensation. Patients in their series demonstrated complete paralysis of bowel and bladder prior to the appearance of motor or sensory loss on physical examination. The disease progression can result in frequent urinary tract infections and neurogenic bladder and bowel incontinence or constipation, as well as leg length discrepancy, foot deformities, gait abnormalities, scoliosis, spasticity, and back and leg pain [24,15].

Urinary complaints in these children are secondary to the impaired innervation of the urinary system, either from malformation during embryogenesis, or a tethered cord as a result of the lipomeningocele. Urinary dysfunction can be due to detrusor paresis, external sphincter dysfunction, or most commonly, detrusor-sphincter dyssynergy [46]. Urinary dysfunction may cause symptoms such as urinary incontinence, frequent urinary tract infections, urinary urgency, and in severe cases of urological dysfunction, hydronephrosis or pyelonephritis may cause upper urinary tract damage. The initial symptom of a neurogenic bladder is frequently a change in micturition pattern [47]. Urodynamic testing aimed at evaluation of urological dysfunction may help with evaluation of the severity of dysfunction. Additionally, abnormality of bladder function may be the only evidence of neurological compromise in these children [48].

#### **Radiological Assessment**

Recent advances in both ultrasonography and MRI have substantially aided the diagnosis and treatment of spinal dysraphism, both prenatally and postnatally. Prenatal diagnosis of lipomeningocele can be very challenging. A detailed examination of the fetal spine requires diligent scanning in various planes, with results that are very dependent on the position of the fetus. This examination has been aided by the utilization of 3D ultrasonography. The integrity of the neural canal is inferred by the regularity of the 3 ossification centers of the spine and the presence of soft tissue covering the spine; visualizing the conus medullaris in its normal location strengthens the likelihood of no abnormalities [49]. With higher frequency transducers, placode contents and cord tethering can be discerned [50].

It may be difficult to detect lipomeningocele by ultrasonography if the spine lies adjacent to the uterus, resulting in limited visualization of the subcutaneous mass. Magnetic resonance imaging is useful in demonstrating the presence of a fatty mass and cord tethering. Additionally, axial images are important in identifying splayed pedicles [50].

While almost all open spinal dysraphisms are associated with an abnormal appearance of the posterior fossa on obstetric ultrasonography [51], in a case of lipomeningocele reported by Kim et al., [52] the posterior fossa was completely normal. On ultrasonography, a well-demarcated subcutaneous mass was detected in the lower sacral area at 36 weeks. The spinal cord was observed to extend into the sacral area instead of being located in the upper lumbar spine, and an additional echogenic intraspinal mass contiguous with the lower spinal cord was identified. Kim et al. [52] reported that MRI revealed similar findings, but did not add new findings to the ultrasonography study.

Postnatally, MRI has aided in both the diagnosis and treatment of lipomeningoceles. Lipomeningocele features can vary substantially depending on the relative size of the lipoma and meningocele, along with the orientation of the neural placode [19]. Characteristically, imaging of lipomeningocele reveals expansion of the spinal canal and subarachnoid space. The cord and the dura extend dorsally through the spinal dysraphism [53]. Most cases present with a deformed and stretched neural placode that is rotated toward the lipoma on 1 side. The meninges herniate on the opposite side. Spinal roots on the side of the lipoma emerge nearer to the neural foramina. These roots are shorter than the roots that emerge from the side where the meninges herniate, and these short roots serve to tether the spinal cord. The neural placode is frequently segmental [19]. A lipomatous dura mater can result if the lipoma surrounds the spinal cord or infiltrates the extradural space.

After operation for lipomeningocele, the cord may not be completely untethered, or after a short period may retether. Often this population will show imaging evidence of a tethered cord or low-lying conus despite the absence of symptoms [55,56]. As such, routine imaging may not be useful in evaluating patients after lipomeningocele resection without clinical indication, unless the MRI is obtained in the immediate postoperative period to delineate a baseline. A study of 140 cases of tethered cord consisting of 48 cases of lipomeningocele demonstrated no advantage in obtaining routine postoperative follow-up MRI scans [57]. In this study, only a single reoperation was performed on lipomeningocele, and that was prompted by clinical evaluation rather than imaging. Determining the need for a tethered cord release after primary repair of a lipomeningocele therein remains largely a clinical decision based on neurological function rather than on radiological findings. Additionally, because many pediatric patients may require sedation for MRI, the consideration for obtaining MRI must also include the risk and cost associated with this routine imaging. Tethered cord has been associated with progressive spinal deformities, and a series of 9 patients reported by Tubbs et al. [58] noted that tethered cord should be suspected in the presence of

symptoms of tethered cord with an increasing lumbosacral angle.

### **Specialized Testing**

Additional specialized testing such as urodynamic function studies and neurophysiological monitoring may be beneficial in evaluation of these patients to assist with timing of surgical intervention. Neurophysiological monitoring is also beneficial intraoperatively to assist with preservation of functional spinal cord and nerve roots during initial surgery and during repeat detethering procedures. Specific nuances of intraoperative neurophysiological testing will be discussed in a later section.

Urodynamic testing includes invasive and noninvasive testing, both aimed at determining the functional status of the bladder. This testing includes assessment of detrusor function and external urethral sphincter activity, both of which can be compromised in patients with lipomeningocele. Noninvasive urodynamic studies include uroflowmetry, which evaluates the flow pattern during urination, and bladder ultrasonography, which can evaluate the shape and size of the bladder, as well as the postvoid residual, which should be zero if there is no urological dysfunction. Invasive testing includes a cystometrogram, which requires catheterization with or without concurrent electromyography of the external sphincter. This test evaluates bladder capacity, compliance, continence, and emptying [46]. The assignment of a urodynamic score that incorporates the parameters of volume, compliance, detrusor activity, and bladder-sphincter synergy allows for a preoperative measure of overall urinary function, as well as the opportunity to compare overall function both pre- and postoperatively [59].

Neurophysiological testing may be used preoperatively to assess neurological and urological function by way of assessing innervations, and may include somatosensory evoked potentials, motor evoked potentials, and electromyography. Anal sphincter electromyography has demonstrated 96% sensitivity in detecting patients with sphincter dyssynergia and 78% sensitivity in detecting bladder dysmotility. The addition of perineal evoked potentials increased the sensitivity of the combined testing for sphincter dyssynergia to 100% and bladder dysmotility to 86% [54].

### **Surgical Intervention**

Surgical objectives in a lipomeningocele repair include removal of the adipose mass, identification of the defect in the lumbosacral fascia for release of the tether, possible release of the filum terminale, preservation of neural elements, and prevention of retethering of the spinal cord. The lipomatous component is intimately associated with the neural placode, which will preclude complete resection of the lipomatous component without neurological injury [43]. Lipoma resection may be achieved utilizing cautery, laser, or ultrasonic aspiration. Additionally, if primary repair of the dura cannot be achieved, duraplasty may be warranted. There have been minimal associations between the types of dural substitutes used for duraplasty and their relation to retether of the cord [60]. Many of the studies evaluating efficacy of various dural substitutes in duraplasty for lipomeningocele include surgery for myelomeningocele as well. Lipomeningocele is considered among the complex pathologies prone

to secondary tether. A study comparing complex (myelomeningocele, lipomeningocele) to noncomplex (fatty filum, split cord malformation) pathologies demonstrated a higher incidence of retether in complex pathologies with primary dural closure, but no statistical difference in retether with duraplasty [61]. A generous duraplasty, therefore, may in fact be protective against retethering by creation of a larger CSF space around the neural placode [61]. It should be noted that some late complications have been noted with Silastic duraplasty including neomembrane formation, which may predispose the patient to hemorrhage near the operative bed, and low virulence infections [62].

Timing of surgical intervention has remained a rather controversial topic, with some advocating for intervention prior to presentation of neurological dysfunction, and some advocating for waiting to intervene until evidence of dysfunction exists. In the absence of good natural history data on the rate of neurological deterioration secondary to lipomeningocele, it is unclear how many individuals would remain asymptomatic without intervention. However, attempts to determine the natural history of lipomeningocele through extrapolation from initial presentation in various studies would suggest that the condition in a majority of patients will deteriorate over time [63,64]. Up to 40% of infants will display abnormal neurological, orthopedic, or urological dysfunction around birth [41,65,18]. Much of the literature would suggest that earlier intervention leads to better outcomes, yet it is not clear whether this indicates surgery should always occur prior to the onset of neurological symptoms. Regardless of the timing, the goals of the surgery are consistent: prevent future or further neurological deterioration and preserve or improve current neurological function.

In considering the timing of surgical intervention, one must consider the nuances of surgery as they relate to the morphology of the lipomeningocele. Some correlation exists between the morphology of the malformation and subsequent postoperative deterioration. Cochrane [66] attempted to discern which morphologies of lipomeningocele were most appropriate for early versus late operative intervention, by evaluating outcomes in surgery for transitional lipomeningocele. He considered symmetrical versus asymmetrical malformations: asymmetrical malformations tend to be associated with unilateral deterioration, and symmetrical malformations tend to be associated with bilateral and bladder deterioration. Those patients with symmetrical malformations were less likely than those with asymmetrical malformations to exhibit early postoperative deterioration. As surgery involves identification of the subarachnoid space adjacent to the dorsal roots to facilitate detethering of the lipoma from the dura, this is more easily achieved when symmetry is present. In asymmetrical lesions, the rotation of the lipoma-placode interface makes identification of this safe zone more difficult. The difficulty of the case and likelihood for complication may suggest that those cases more prone to postoperative complications may be more optimal for intervention after the first signs of neurological dysfunction appear.

Because one of the highest morbidities of lipomeningocele is bowel and bladder dysfunction, the appearance of these symptoms, or a change in bowel or bladder dysfunction,

should prompt urgent surgical intervention. Longer times to surgery have been associated with worse outcomes [67]. Additionally, urological dysfunction appears to be more reversible in a younger population, namely infants [68]. However, the risk of a major urological problem after surgery such as frequent urinary tract infections, hydronephrosis, need for clean intermittent catheterization, or vesicoureteral reflux appears to be higher in those patients with lipomeningocele as opposed to patients with other closed spinal dysraphisms, and would suggest that a somewhat conservative approach may be indicated in completely asymptomatic patients [69]. Despite the favorable outcomes for early surgery in some series, one must consider both the risk of conservative management and risk of surgery in planning the timing for intervention. From the standpoint of patients who complain primarily of motor or sensory deficits, recovery of motor and sensory deficits occurs more frequently than recovery of normal bowel and bladder function [15].

#### **Intraoperative Monitoring**

Lipomeningocele may be considered a high-risk group within spinal dysraphisms amenable to resection given the absence of a discrete plane between the lipoma and neural placode, and the rotational component that may be present as a result of the laterality of the lipoma [70]. To facilitate safe resection of lipomatous components and detethering of the spinal cord via sectioning of the filum terminale, intraoperative neurophysiological monitoring may be used, and may alter the intraoperative surgical plan [71]. Monitoring may include somatosensory evoked potentials, motor evoked potentials, and both stimulated and free-run electromyograms. This monitoring requires the use of total intravenous anesthesia, generally propofol and fentanyl or remifentanyl, with only short-acting muscle relaxants used during induction so as not to interfere with monitoring ability [72,70,73]. Somatosensory evoked potentials monitor the integrity of the dorsal column pathway, and frequently the tibial nerve is monitored for evaluation of the L4–S3 nerve roots, although this method is limited by long averaging times and fluctuations in response [73,74]. Motor evoked potentials may be obtained for the quadriceps, anterior tibial, and gastrocnemius muscles, as well as bilateral external anal sphincters using needle electrodes to monitor the lumbosacral nerve roots [70]. Because younger children have more immature myelinated fibers, monitoring of motor evoked potentials may require double-train stimulation to obtain useful motor evoked potential amplitudes [75,76]. The difficulty of obtaining free-run electromyograms should be noted when electrocautery is used [72]. Stimulated electromyography, however, allows for identification of functional nerve roots and delineation of nervous tissue from filum and scar elements [73,74]. In particular, external anal sphincter monitoring allows assessment of the pudendal nerve comprised by the S2–4 nerve roots and provides a good approximation of external urethral sphincter injury as well, although it may not well approximate injury to the parasympathetic nerves that supply the detrusor muscle [74].

#### **Outcomes**

Risk of surgery include CFS leakage; neurological deterioration either secondary to nerve injury during surgery or as a result of tethered cord; and incomplete wound healing

or wound breakdown, infection, and meningitis. The overall complication rate of surgery is between 10% and 30% [55,15]. In a series of 120 patients, worse neurological function was found in 5.8% of patients after primary surgery [55]. The incidence of spinal cord retethering following lipomeningocele resection has been found to be between 10% and 20% [77,24,15,18]. Retethering may present first with back pain and deterioration of lower-extremity function followed by worsening urological and bowel function, generally occurring 3–8 years after the initial surgery [55,63].

Outcomes after surgery for lipomeningocele are dependent on the preoperative function of the patient. A series of 80 patients demonstrated that 92.1% of children with a normal preoperative examination had no neurological deficits or bladder dysfunction at long-term follow-up, and all had normal bladder function. However, none of the children with preoperative bowel and bladder paralysis recovered normal function, despite improvement in sensory and motor deficits in this subgroup. Bowel and bladder paralysis remained the greatest morbidity of this series, with complications of recurrent urinary tract infection and pyelonephritis [15].

Another series of 120 patients who underwent surgery for lipomeningocele demonstrated improvement in

functional grade (a grading system incorporating neurological, urological, and orthopedic deficits) in 10% of patients and deterioration in 5.8%, with the remainder maintaining the same functional grade. Those patients whose conditions deteriorated the most commonly displayed worsening bowel and bladder function [55]. A series of 43 patients demonstrated that 84% of patients maintained stable urodynamic and neurological function after surgery and noted the primary predictor of normal bladder function in the long term was normal preoperative urodynamic status [78].

### Conclusions

Lipomeningocele is a form of closed neural tube defect with unclear predisposing factors. Due to the risk of worsening neurological and urological function secondary to a tethered spinal cord, it continues to be important to identify this condition for timely intervention. Magnetic resonance imaging and neurophysiological testing are useful tools for identifying the spinal cord pathology and assisting with surgical planning. Because bowel and bladder paresis remains the primary morbidity of this disease, early intervention either prior to symptom development, or at the first onset of symptoms, is recommended to optimize postoperative outcome.

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