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Surgical Treatment of Neural Tube Defects

Juraj Šutovský

Abstract

Neural tube defects (NTDs) are developmental pathologies associated with undesirable lifelong consequences. Incidence of these pathologies differs between countries and regions depending on socio-economic and healthcare quality. It is also influenced by folic acid and zinc supplementation. Genetic factors influence probability of NTD, increasing risk of defect in siblings up to 3–8%. Estimated incidence in United States is 3–4/10000 live births, and worldwide incidence increases on about 10/10000 live births. Despite various types and localizations of spina bifida, in all of them neural tissue is in danger. This can lead to various types of neurologic disorders. Not only due to direct damaging of spinal cord and nerve roots but also other parts of central nervous system are also endangered by disturbed prenatal development. Other consequences as orthopedic abnormalities, bladder, and bowel dysfunction influence quality of life. Surgical therapy is often the only possibility to preserve existing function of neural tissue, allows its further development and prevents complications. In this chapter surgical techniques with aim to restore spinal cord and nerve roots anatomy, preservation of its function and defect closures are presented. Also, treatment of possible comorbidities and complications is discussed. Spina bifida management requires multi-speciality cooperation and care to monitor, prevent and treat various potential complication that can negatively influence quality of life and even survival. Prenatal diagnosis is based on maternal screening of serum alpha fetoprotein (AFP) levels and prenatal ultrasonography examination. As the suspicion of neural tube defect arises, an amniocentesis is recommended to complete a genetic analysis and obtain amniotic fluid for more precise AFP and acetylcholinesterase examination. Some types of neural tube defects are diagnosed after delivery, some are symptomatic until adulthood and some are diagnosed incidentally. Each of them requires specific management, based on underlying pathology.

Keywords: neural tube defect, neural placode, spina bifida, hydrocephalus, tethered spinal cord

1. Introduction

Neural tube defect is with congenital heart defect the most serious birth anomaly compatible with life. Besides genetic influences, zinc and folic acid deficiency plays important role [1]. As neural tube closure occurs in fourth gestation week its failure should be diagnosed early during pregnancy. Despite spontaneous defect closure had been reported [2], once neural tube defect is diagnosed a pregnancy termination is an option. Up to 60% diagnosed NTS in Europe ends with planned termination of pregnancy [3].

Another option is in utero surgery, open or fetoscopic.

In the case of late diagnosis or gravidity does not match prenatal surgery criteria or just because in utero surgery is not accessible, postnatal surgery comes in role.

If spina bifida is diagnosed prenatally, delivery should be done in center with capabilities for taking care for both, mother, and new-born.

2. Spina bifida

Spina bifida aperta

Myelomeningocele

Myelomeningocele is the most common type of spina bifida aperta resulting in defective neural tube closure between 22nd and 26th days of gestation. Most occurred at lower lumbar segment, with neurological impairment below level of lesion. Not only distorted development but also exposure to amniotic fluid, toxic to neural tissue, is the cause of neural tissue malfunction.

In this type of spina bifida aperta, unclosed neural tube, neural placode is exposed directly to external environment and through arachnoid/connective tissue is attached to dysplastic skin (**Figure 1**).

Almost all cases are associated with some degree of Chiari II. malformation, and majority of them requires CSF derivation due to hydrocephalus.

Preoperative management

Because neural tissue in spina bifida aperta/myelomeningocele is exposed to external environment delivery is planned as cesarean delivery to minimize risk of infection. Nonlatex gloves usage is recommended while handling a baby due to possible allergy. Immediately after delivery, the open spina bifida must be covered with a sterile non-adherent saline-soaked dressing to prevent infection and dehydration due to CSF and interstitial fluid losses.

Postnatal thorough examination is recommended to rule out other congenital malformations and conditions that prevent early surgery, especially pulmonary and cardiac malformations. Cranial ultrasound is obligatory for cranial malformation identification. Preoperative MRI is helpful for anatomical orientation and associated pathology exclusion (**Figure 2**). If there are no contraindications, operation is indicated as soon as possible ideally up to 12 hours postnatally but not more than 24 hours. Any delay in spina bifida closure increases risk of infection and hemodynamic instability.

Besides early surgery, broad spectrum antibiotics are warranted administered, at least until the skin lesion is closed, to minimize the risk of infection of the central nervous system.

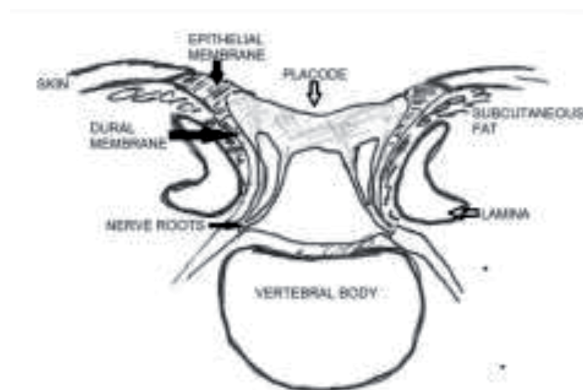


Figure 1.

Schematic picture of myelomeningocele. Neural placode is in direct contact with external environment.

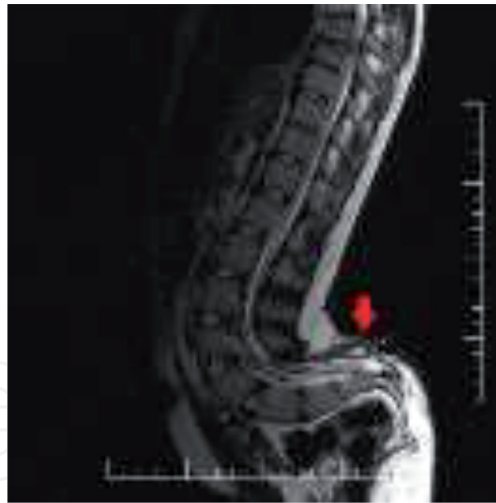


Figure 2.
Sacral myelomeningocele (red arrow) on T2 MRI.

At the surgical theater new-born should be positioned prone or laterally to avoid pressure on the placode prior to surgery. Besides dehydration attention must be paid to possible hypothermia.

Before operation field preparation swabs for aerobic and anaerobic cultures should be taken.

Because of exposed neural tissue, operation field is prepared only with sterile saline and surrounding skin can be prepared in standard manner. Draping must be adapted to planned type of surgery. If direct closure of skin defect is in question, sufficient visible skin must be left for flap reconstruction. Swabs for aerobic and anaerobic cultures should be taken from the skin and CSF samples as soon as CSF is encountered.

Neural tube closure

Spina bifida surgery must be done under microscopic control. Sharp dissection is mandatory to prevent traction injury to already damaged neural tissue. The primal incision starts in midline cranially to the lesion, best above the level of identified spinous process cranial to defect. Dissection continues caudally to the placode at the border between skin and arachnoid, which is not regular with possible extension of dystrophic skin reaching placode. Cranial view from upper part of the placode reveals spinal cord emerging from the spinal canal. Separation of neural placode with adjacent arachnoid continues in step by step manner. After circumferential neural placode dissection from skin, all arachnoid adhesions must be released, preventing spinal cord tethering. Dissection must be done with awareness of possible spinal nerves lateral to placode adherent to sac.

After deliberation thorough inspection of neural placode must be done to remove any non-nerve tissue reducing not only possibility of future spinal cord tethering but also preventing epidermoid/dermoid cyst or lipoma formation. In cases of lipomyelomeningocele, with indistinct boundaries between neural and adipose tissue, perioperative neuromonitoring is useful and should be used.

Dorsal nerve roots usually just below placode edge and ventral roots emerging more medially from the ventral side of placode are inspected and are released from adhesions allowing neural tube reconstruction without tension. Aberrant nerve roots ending blindly in dura must be divided.

At the caudal end of placode filum terminale should be identified. When it is clearly recognizable, its section is indicated to minimize risk of tethered spinal cord and prevent future neurological deterioration and need for another operation.

During surgery not only neural tissue but also vascular supply must be managed carefully. Vessels passing from formed spinal cord and nerve root vessels must be preserved. If any bleeding occurs, avoiding coagulation is recommended until inevitable. Primal management of bleeding can be done with haemostatic material (Gelaspon or Flo-seal) or cotton patches alone.

After placode deliberation inspection for any other fixation point is mandatory (**Figure 3**). The attempt to reconstruct neural tube is the next step. Approximation of lateral edges medially with arachnoid-pial sutures partially re-create tubular spinal cord with arachnoid-pial outer surface, what minimizes risk of adhesions and spinal cord fixation. Non-absorbable 7/0 – 8/0 monofilament sutures are used. Care must be taken not to “squeeze” neural tissue with sutures. If the placode is too bulky relinquish attempt to reconstruct neural tube not to damage liveable neural tissue at the cost of higher possibility of spinal cord tethering.

After the placode is released and neural tube is reconstructed eventually, dural sac closure is the next step.

Intact dura is identified at the cranial part of defect, at the site of lower lamina. After dura is identified at the upper part, epidural fat and space between vertebral structures and dura is the plane for sharp dissection and identification of dural lateral borders, where dura continuously passes to the thoracolumbar fascia. At this transition, dura must be divided by sharp dissection. If there is a sufficient dura for primary closure of dural sac without neural tissue compression, dura is closed with continuous suture with non-absorbable 5/0 monofilament thread. If there is no sufficient dural covering for closure or closure without compression of neural tissue, grafts must be used. As a first choice is autologous substitute harvested from thoracolumbar fascia or muscular flap from latissimus dorsi muscle. If there is no possibility acquiring sufficient autologous flap, nowadays artificial collagen substitute of dura (DuraGen) can be used. Attention must be paid to eliminate any compression of neural tissue, allowing free passage of CSF around deliberated placode and prevent later adhesion.

If there is any suspicion of possible CSF leakage, dural glue helps to prevent complication. Eventually another layer formed by thoracolumbar fascia can be added if there is enough material left. Vertical parallel fascial incisions help mobilization of thoracolumbar fascia (**Figure 4**).

Skin closure



Figure 3.
Thickened filum terminale (red arrow) fixing spinal cord after successful myelomeningocele reconstruction.

Direct suture in small defects is most often doable. Undermining skin laterally helps relax tension on suture. Absorbable adaptational subcutaneous sutures help to relieve strain on skin edges, eventually subcutaneous tissue can be fixed to medial thoracolumbar fascia. Skin is closed with non-absorbable 5/0 monofilament. If there is still tension on the skin suture, lateral longitudinal relaxing incisions in the similar manner as the thoracolumbar fascia relaxation, without reaching subcutaneous tissue.

If a direct suture is impossible, cutaneous flap can be used to release tension on the suture. Usage of flap type depends on defect localization and surgeon's familiarity with surgical technique. A single rotational flap is the simplest one. The universal rhomboid (Limberg) flap useful in closure bigger defects. Also, O-to-Z flap can be helpful in skin closure especially in more longitudinal than wide defects (**Figure 5**).

If neurosurgeon is not completely familiar with skin flap techniques, or skin defect is too large, plastic surgeon should be a part of surgical team.

Postoperative management

After skin closure operative field is disinfected in standard manner and wound is covered with sterile non-adhesive covering (Atrauman, Atrauman Ag) to prevent wound injury during redressing, and sterile gauze. To prevent wound contamination postoperatively with urine and stool, wound is covered with plastic adhesive drape.

Postoperatively prone position with pelvis little bit elevated, to release CSF pressure in lumbar region. Prone position is preferred up to 7th postoperative day.

Wide spectrum intravenous prophylactic antibiotics are indicated at least 24 hours postoperatively. Better choice is extended prophylaxis until results from perioperatively samples are negative. If cultivation reveals infection, therapeutic regimen according results is indicated.

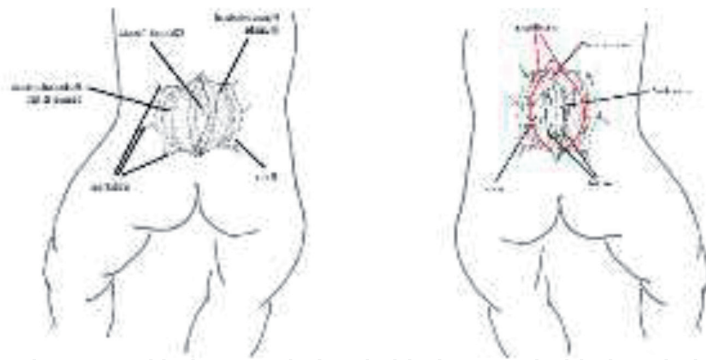


Figure 4. After dural closure, with, or without graft, curvilinear fascial incision is helpful in thoracolumbar fascia mobilization and addition another covering layer.



Figure 5. Skin closure usually possible with direct suture. In cases of larger defects, O-to-Z, or Limberg flaps are helpful in releasing suture tension.

Early postoperative complications

Due to exposure of neural tissue to outer environment, insufficient skin coverage, stress due to pain, surgical procedure and feeding problems, complication related to wound healing occurs.

Most common is skin healing problem. It is often due to insufficient blood supply because of high tension at the suture site or extended skin undermining with perforator vessel damaging. Usually skin necrosis is only superficial and besides daily check-up does not need any other action and reepithelization occurs. If the skin is affected in full thickness with deeper layer consisting of thoracolumbar fascia with underlying muscles are intact, debridement of necrotic tissue must be done until normal tissue with blood oozing from edges is apparent. With no other complication secondary healing with reepithelization from edges occurs, despite it takes longer time and meticulous wound management preventing secondary infection is necessary. Necrosis whole thickness of reconstructed dura covering requires revision surgery with plastic surgeon to recover defect with sufficiently vascularized flap.

After surgery wound must be inspected on daily basis to identify any problem with wound healing. Any collection under the skin worsen healing applying tension to the wound. The cause of tension and collection must be identified by aspiration. Hematoma or interstitial fluid collection must be evacuated. Not only internal, but also external compression from wound dressing must be avoided.

If there is CSF collection, just on 1st-3rd day after surgery with no apparent progression and tension, it may be due to fragile lumbar dura, CSF oozes around the stitches and collection resolves spontaneously up to 6th-7th day after surgery. Closure of dural defect can lead to changes in CSF flow pattern, with increasing pressure in subarachnoid and intraventricular spaces. In this case CSF may accumulate at the surgical site later, usually on 5th-7th day after surgery. If there are no sign of progressive hydrocephalus and closure is done in standard manner, collection may dissipate spontaneously after subarachnoid/intraventricular pressure comes to balance.

With any doubts or problems about the sufficient closure of all layers with progressive cumulation of subcutaneous CSF collection even if there is CSF leakage through the skin suture, CSF derivation and surgery site revision is indicated.

External ventricular drain, despite higher risk of neuroinfection, or subcutaneous reservoir (Rickham, Omayya) is the first option in cases without hydrocephalus. It is necessary keep in mind, that reservoir is associated with need for repeatedly evacuations of CSF what increases risk of infection and is also painful and stressful procedure.

Ventriculoperitoneal shunt in the case of CSF leak at the site of spina bifida closure is not ideal option, because valve opening pressure is usually higher as the pressure of sutured soft tissues at the site of spina bifida, thus not solve the problem. Possibility is the programmable valve with adjusted opening pressure on 0 mm Hg and after resolution wound problem readjusted to the desired level.

Surgical site infection is relatively rare. Reported incidence is about 1-12% [4].

If the wound infection is identified, without signs of intradural infection, and no presence of any subcutaneous collection, swabs for microbiological examination from surgical site should be done and local therapy applied. If there are signs of abscess formation, aspiration of purulent material for aerobic and anaerobic cultivation should be obtained and releasing of few sutures for effective drainage and antimicrobial rinsing abscess cavity. Wound defect is left for secondary intention.

If there are signs of neuraxis infection proved by clinical and laboratory examination, CNS penetrating wide spectrum antibiotics are administered. After cultivation proves infection, targeted antibiotics are indicated. In any suspicion of surgical site intradural infection, revision is indicated with removing of purulent content avoiding placode and nerve roots damage with surgical site irrigation with antibiotic solution. Wound reclosure is done in the same manner as in the primary surgery.

Meningocele

Meningocele is another open neural tube defect with protruding only meninges through the insufficient lamina and back soft tissue. Neural structures are not a part of the sac. Sometimes meninges are not fully visible from the outside but are covered with dysplastic or fully developed skin. As there is no damage to the neural structures, long time outcome is more favorable as is in myelomeningocele.

Prenatal, postnatal, and surgical management is identical to myelomeningocele with no necessity of neural tube reconstruction.

Because there is a malformation of dural development, during surgery is warranted close inspection of spinal cord with releasing it from any possible arachnoidal, fibrous adhesion to prevent from tethered spinal cord. Also, filum terminale should be inspected and cut if it is in the surgical field [5].

Prenatal surgery

Prenatal surgery is an option in OSB cases. According to MOMS study (Management of Myelomeningocele Study) foetal surgery [6] is promising treatment which reduces need for VPS approximately of 50% and lower incidence of severe hindbrain herniation and improves ability to walk independently (**Figure 6**).

But according to meta-analysis study, prenatal spinal closure does not reduce the possibility of SCT, conversely higher incidence is reported [7] (**Figure 7**).

This result must be evaluated with caution because foetal surgery is a demanding procedure associated with risk of premature birth, potential morbidity for mother and can be a limitation for further pregnancies.

Spina bifida occulta

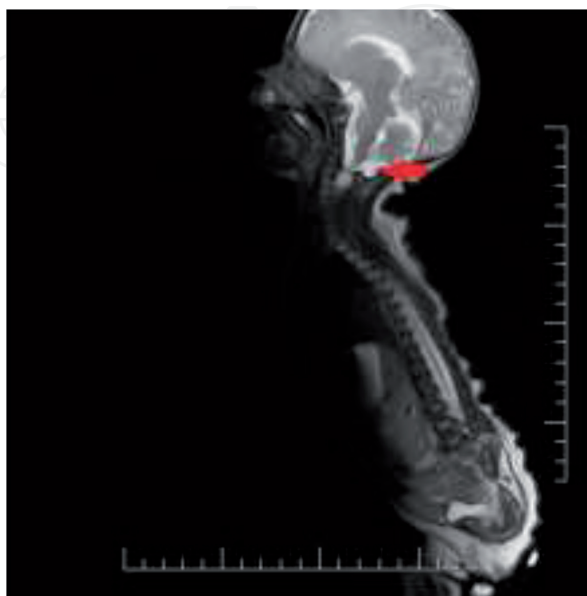


Figure 6.
Complete resolution (red arrow) of hindbrain herniation after in utero myelomeningocele closure.

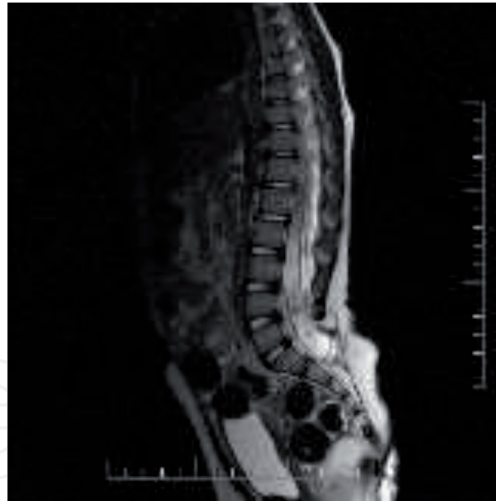


Figure 7.
Despite prenatal surgery, tethered spinal cord persist postnatally.

Many defects of neural tube closure are not apparent, so they are named “spina bifida occulta” (SBO) or “closed spina bifida”. As these defects are results of failed secondary neurulation, they are covered with continuous, sufficient skin.

There are several subtypes of SBO. Cystic lesion protruding through open spinal canal covered with sufficient skin can be formed only with meninges – meningocele, containing only fat tissue – lipomeningocele, or with added neural structures – lipomyelomeningocele. A rare type of cystic SBO is terminal myelocystocele. Another SBO variant without cystic component are the most common type of SBO. They comprise diastematomyelia, diplomyelia, dermal sinus (with or without epidermoid or dermoid cyst), lipoma, teratoma and tethered spinal cord due to various causes (adhesion, thickened filum terminale w/o fat tissue).

Typically, they are not only covered with the skin, but also initially are not presented with any neurological symptoms at least in the new-born period. No other neuraxis defects are usually associated with them.

At the level of covered NTD, typically in the midline or near midline, usually some cutaneous stigmata, like hairy patch, dimple, naevus, skin pigment changes or even subcutaneous mass can be detected. Any skin defect along the spinous processes or paraspinal skin is suspicious from occult spinal dysraphism and need further investigation. Ultrasound can be useful for initial screening but cannot be used stand-alone. MRI is a definitive tool for diagnosis and evaluation of the defect.

If even clinical symptoms of occult spinal dysraphism presents, they are usually symptoms of tethered spinal cord manifested during growing up, when spinal cord is fixed at the same level as at birth and does not adapt to osseous spinal canal growth.

Problems usually start with voiding and stooling problems that can be overlooked in new-borns. Continuous dripping may attract attention and should prompt bladder volume measurement and urge urological and neurosurgical consultations. Walking delay in children and contractures and deformities of lower extremities, back and legs pain, scoliosis, and onset of sphincter problems in childhood and adulthood justify the exclusion of OSD. Rarely recurrent neuroinfection can be the sign of OSD.

As usually no initial neurological deficit or other neuraxis malformation is present, question about indication for surgical repair arises. As many spina bifida occulta cases are diagnosed incidentally in adulthood, on graphical examination due to other problems, or even post-mortem, conservative management is a logical conclusion.

Against postponed surgery is fact, that surgery at the time of developed symptoms rarely reverse neurological deficit and intervention is successful if neurological deficit remains stable, with minimal possibility for improvement.

As sequelae of spina bifida occulta can be combination of symptoms including bladder dysfunction, orthopedic problems seriously limiting quality of life, preventive surgery or at least long-lasting meticulous patient's observation is recommended.

Cystic spina bifida occulta

As the surgery for SBO is indicated, there is no big difference with spina bifida aperta.

If meningocele covered with sufficient skin is indicated for surgery without signs of tethered spinal cord syndrome, e.g. for cosmetic reasons, after skin incision and identification and preparation of dural sac, just its resection and watertight dural suture without squeezing neural structure is the goal of surgery. Intradural inspection for any possible adhesions to spinal cord or thickened filum terminale is warranted. If there are clinical and radiographical signs of tethered spinal cord meticulous MRI evaluation must be done to rule out other sites of spinal cord fixation.

Lipomeningocele is another subtype of cystic SBO. The cyst is formed by dural protrusion filled with fat tissue which part of spinal cord within spinal canal. If the neural tissue is a part of the cyst, lesion is named as lipomyelomeningocele. Distinction of this two lesion is only didactic, because neural tissue is always an integral part of lesion and both are the results of the same pathology when premature disjunction of epithelial ectoderm from neural ectoderm occurs, allowing mesenchymal tissue come to contact with neural ectoderm, which stimulates mesenchyme to develop to fatty tissue. Ventrally is mesenchyme stimulated by neural plate to transform to meninges and interface between meninges and fat tissue is at neural ridge. Skin covering lesion is completely differentiated with possible skin marks. As the tethering of spinal cord is inherent to this lesion, surgery is indicated to prevent deterioration of preserved neural functions. If there is apparent soft tumor visible or palpable under the skin or if there are cutaneous stigmata suspicious from SBO or even suspicious tethered spinal cord syndrome based on clinical presentation HR MRI proves diagnosis (**Figure 8**) Urologic and urodynamic testing proves impairment of bladder function. Preoperative electrophysiological examination assesses lower sacral and coccygeal nerve roots functions. Electromyography of anal sphincter demonstrate 96% sensitivity of sphincter dyssynergia and 78% sensitivity in



Figure 8.
Thoracic and lumbar lipomyelomeningocele (red arrows) on MRI.

bladder dysmotility detection. With perineal evoked potential added, sensitivity for sphincter dyssynergia increase to 100% and for bladder dysmotility to 86% [8].

Interface between neural placode and lipoma is indistinct. So, the aim of surgery is not to remove fat tissue intimately adherent to neural tissue, but deliberate spinal cord and reducing fatty mass, with preservation of neural structures, filum terminale resection and prevention of re-tethering. Lipoma may be resected using ultrasonic aspirator, cautery, even laser, while preserving neural structures intact. It is better left more fatty tissue on neural placode as do irreversible damage, while distinction between neural placode and lipoma is impossible even under microscopic control. For maximizing surgery safety, intraoperative electrophysiological monitoring using somatosensory evoked potential, motor evoked potential and electromyography including anal sphincter monitoring is necessary.

As the lipomyelomeningocele and lipomeningocele are of all spina bifida malformations most prone for retethering, generous duroplasty must be done to create copious space filled with CSF preventing postoperative adhesions. After watertight duroplasty, upper layers are closed in standard manner.

Postoperative complications and their management are like open spina bifida.

Terminal myelocystocele is a rarest form of cystic SBO, accounting about 4–8% of all OSD cases [9–11], which is defined as cystic dilatation of central canal protruding dorsally to subcutaneous tissue in lumbosacral region, usually coupled with another spinal, anorectal and genitourinal malformations. This malformation is caused by disruption of secondary neurulation resulting in persistent terminal vesicle and its expansion through dorsal mesenchyme but not epithelial ectoderm. Cystic dilatation is lined with ependymal cells and is continuous with central canal. Transition zone is funnel-like on MRI. At the basal site of the cyst is formed by placode, cranially continuous with spinal cord. Usually this malformation, besides subcutaneous mass and cutaneous stigmata is presented with neurological deficit, but cases without no neurological disturbance had been referred [9].

Spinal cord fixation is always associated with this lesion, so detethering is indicated. There is no imminent threat of neurological deterioration and surgery can be done electively. The principle of intervention is releasing spinal cord and placode and if possible, neural tube reconstruction. Starting with midline incision above lesion and resection of fat tissue/lipoma to identify dural sac. If necessary, cranial laminectomy allows better identification of dura. After dural opening cyst is in view, with possible adhesions. Any fixations also of spinal cord in operative field must be released. After cyst resection at the bottom placode is visible. After neural placode, spinal cord and nerve roots identification, any fixation points of neural structure responsible for persistent or newly developed fixation must be unloosed. Reconstruction of placode using 5/0 monofilament nonabsorbable pial sutures prevents retethering. Dural reconstruction is done in standard manner, bearing in mind not to compress or squeeze underlying neural structures. Flap from adjacent paravertebral muscles fortify dural suture and prevent CSF oozing. Subcutaneous tissue and skin are closed in standard way. After surgery prone position with slightly elevated hips helps wound healing. Due to surgical site localization nearby anus, infection prevention is particularly important.

In addition to the caudal end of spinal canal, myelocystocele can be found wherever along the spine. Neurological impairment is defined by the malformation level. Surgery is performed in the standard manner described above.

Spinal lipoma

Spinal lipomas are the most frequent varieties of non-cystic SBO. They are mature teratomas, consisting of mature adipocytes, which react to metabolic stimuli like fat tissue anywhere in the body, what may influence their clinical presentation.

The filar lipoma, lipoma as a part of filum terminale, is the most common type of intradural lipoma. Its estimated incidence is about 1–5% and symptomatic are about 5% of them [12].

Filum terminale is infiltrated with fat in various extent. Commonly caudal nerve roots are not involved or compressed. If the filum terminale lipoma is symptomatic, it is usually associated with low lying conus medullaris and tethered cord syndrome. As asymptomatic cases are not usually diagnosed and diagnosis is made incidentally, there is not strict indication for surgery. On the other hand, filum terminale resection is surgery associated with low risk and can prevent later deterioration.

In symptomatic cases, surgery of filum terminale resection at the level of lipoma, through midline incision, laminectomy and durotomy is indicated.

Conus medullaris lipomas are distinct category of lipomas in lumbar spine, which are broadly adherent to lumbar spinal cord. Usually are associated with cystic spina bifida. As the fat tissue is intimately connected with neural tissue, complete resection is rarely possible. Intraoperative neuromonitoring is necessary for surgery safety.

Classical intradural lipomas occur along whole spinal canal, with predominant location in lumbosacral region. They are enclosed in normally formed dural sac. Their localization defines their clinical presentation, caused by spinal cord compression. As mentioned before, their separation from neural tissue is difficult, so partial resection with duroplasty is principle of surgery.

Split cord malformation

Split cord malformation (SCM) is category of OSD which includes two subtypes – diastematomyelia and diplomyelia. According generally accepted theory, these two types are variations from opposite sides of one developmental failure as proposed by Pang [13].

In diastematomyelia, type I., there are two hemicords, each with its own dural covering, separated by septum. Septum is composed of connective tissue, usually bone. Also, cartilaginous septum or spur can separate this hemicords. Septum is in general composed also of other types of connective tissue, including blood vessels. Arteries can be huge and making surgery difficult. Septum can be localized more laterally to either side or can be oblique in relation to spinal canal axis. This type is usually associated with vertebral anomalies.

In type II., diplomyelia, there are two cords enclosed in common dural sleeve, usually separated with fibrous septum, attached to ventral and dorsal dura. There are identified three types of septa according its attachment to dura. One type is only septum attached to the dorsal circumference, another with attachment to ventral dura and the last one traversing all internal diameter of dural sac. Septal attachment to the dura is usually localized more caudally as its attachment to the cleft between hemicords, as a sign of spinal cord tethering.

The embryologic basis of pathology is still in question.

Although some SCM are diagnosed prenatally, most of them are diagnosed after delivery or even in adulthood. At the birth neurological status is normal or almost normal. SCM is associated with cutaneous stigmata. CT and MRI are diagnostic tools capable to identify not only two hemicords, but also character of septum and identification vessels in it.

Clinical presentation is associated with spinal cord fixation, presenting with tethered cord syndrome, usually in type I., while the diplomyelia can be asymptomatic, but as a possible source of spinal cord tethering with insidious neurological deficit progression, when diagnosed, is also indication for surgical treatment.

In the type I. SCM (diastematomyelia), after midline skin incision above lesion and posterior bony structures are skeletonized, laminectomy above and below

lesion is performed. It is sometimes tricky, because in this type of SCM laminae used to be hypertrophic and fused. After identification of dura at the sites of laminectomies, laminectomy on the top of lesion, from lateral to medial is done up to septum. Next step is removing intervening septum, what may be challenging step because of vessels in it. In this step bone wax or haemostatic material is helpful. Dura above and below split cord is opened, and unified spinal cord is identified. Dural opening continues above both hemicords. Medial adhesions between hemicords and dura are sharply cut. Some aberrant nerve roots, ending in medially located dura, can be identified, and interrupted. Medial dura is resected. There is no need to close ventral dura. Dorsal dura is closed in standard watertight manner, and if needed, with graft. Muscles, fascia, subcutaneous tissue, and skin is closed standardly.

The type II laminectomy is usually easier to do because laminae used to be more delicate. After exposure of dura, there are usually no signs of intradural septa visible from outside. Sometimes this pathology is associated with meningocele manqué, which must be distinguished from septum. Paramedial vertical durotomy is done. After elevation of dorsal dura, posterior circumference of spinal cord is inspected. If there is septum, must be sharply bisected. Along the septum are sometimes vessels resembling arteriovenous malformation or aberrant nerve roots. If possible, vessel should be preserved. In this type of SCM both hemicords are in intimate relation, so ventral inspection through the medial cleft is not recommended. Gentle rotation of spinal cord usually allows visualization of ventral dura and safe section of septum. Watertight dural closure and wound suture is done in standard manner.

Dermal sinus tract

Dermal sinus tract (DST) is a form of occult spinal dysraphism. Incidence of this malformation is estimated to 1 in 2500 live births [14].

This type of OSD is characterized by tract between skin and deeper structures. It is result of impaired disjunction between dermal and neural ectoderm at the 3rd to 8th gestation week. Majority of referred cases are in lumbar and sacral region. Epithelial-lined tract can end in variable depths ranging from subcutaneous tissue to dural sac, which is the most frequent its termination and used to be associated with intradural inclusion tumor, i.e. dermoid/epidermoid tumor, even teratoma. Cutaneous stigmata are associated with DST in 77.1%, what can help differentiate DST from benign sacrococcygeal pits [15].

In the case of transdural penetration DST is associated with tethered cord syndrome. Dermal sinus tract can be reason of repetitive neuroinfection. Source of infections is usually external, passing through the tract, but inflammation can also be caused by chemical irritation from leakage of dermoid cyst content.

Different incidence of clinical symptoms is referred in literature ranging from up to 1% [16] to 75% [14].

If there is clinical suspicion of DST, its probing is not recommended. Primary diagnostic tool is ultrasound with its specificity 98% [17].

Definitive diagnosis is confirmed by MRI, which can reveal not only tract, but also its relation to dural sac and any intradural pathology (**Figure 9**).

Although MRI does not identify tract traversing through dural sac, surgical treatment is indicated. Despite high sensitivity of MRI, narrow tract ending in dural sac can be unidentified on MRI.

Surgery is performed with midline skin with elliptical incision encircling external ostium. Tract is followed until its end is identified, even if unplanned laminectomy must be done. If there is a contact with dura mater, part of dura encircling tract is excised with intradural revision. Intradural pathology, if identified accidentally, or identified preoperatively, are dealt according its nature.

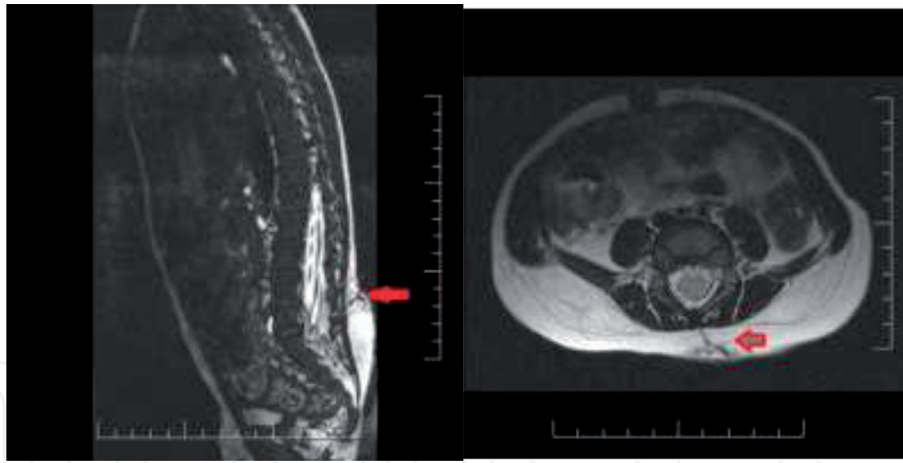


Figure 9.
Dermal sinus tract extending to the dura mater (red arrows) on sagittal and coronal MRI.

Dermoid/epidermoid, teratoma tumors are excised. Internal decompression of dermoid/epidermoid tumors increases the safety of the operation and aids in complete excision. Any part of the left tumor is a potential source of recurrence. Any adhesions to spinal cord must be released and filum terminal, if is accessible is indicated for resection. Wound closure is done in standard manner. Prone position after surgery is preferred.

Meningocele manqué

Special type of neural tube defect is so called meningocele manqué. In this type of malformation spinal cord, nerve roots or filum terminal are fixed by fibrous band formed by atrophic or incomplete meningoceles. These fibrous bands can pass through all soft back tissues ending at the skin or can end on lamina or be fixed to dura mater. This is like reverse dermal sinus tract.

Clinical presentation is identical to tethered spinal cord syndrome, consisting of reflex changes on lower extremities, progressive weakness, muscle dystonia, pain and/or bladder dysfunction.

Suspicion arises with cutaneous stigmata as described before.

Even with evident clinical presentation, there might no sign on MRI of spinal cord fixation with conus localized at the typical L1/L2 disc level. Thorough MRI scans examination, especially high resolution CISS MRI can be helpful in diagnostic hesitation.

If there is a proved, or suspect intradural spinal cord fixation correlating with clinical presentation, surgery, even exploratory, aimed to releasing any adhesions, is recommended. To minimize risk of perioperative neurological injury, perioperative monitoring is inevitable.

Surgery of accompanied pathologies

Hydrocephalus

Hydrocephalus is often, up to 85% of new-born with SB, associated disease. It may be identified prenatally on ultrasound or may be identified or develop after delivery. According to multiple observations, the higher is the spinal lesion localized, shunt dependent hydrocephalus is more probable [18].

Pathophysiology of hydrocephalus associated with spinal dysraphism is still in question. CSF leakage through spinal canal defect causes lowering CSF pressure intracranially resulting in circulation and resorption pathways non-development. After open spina bifida repair missing CSF resorption mechanism results in its accumulation and hydrocephalus.

Obstruction of CSF outflow due to hindbrain protrusion to the spinal canal may be the source of hydrocephalus development.

In the case of apparent hydrocephalus, MRI is indicated to exclude other reasons of obstruction of CSF pathways.

Timing of surgery is still in question. In the case of apparent hydrocephalus identified in utero or at birth, hydrocephalus surgery together with spina bifida aperta closure is recommended. One stage surgery of hydrocephalus and open spina bifida helps wound healing by reducing CSF pressure, reduce risk of CSF leakage and infection.

Doubts arise in the case of no clinical/graphical signs of postnatal hydrocephalus. Ventricular volume may be reduced because of CSF loss through open spinal defect. After defect closure, intraventricular CSF accumulation occurs and head circumference rises, but not necessarily due to pathologically elevated ICP and hydrocephalus. In new-borns calvarium bones are cartilaginous with open sutures, intracranial pressure reflects skin elasticity. Tendency is to avoid invasive procedures unless they are necessary, so MRI and CT are not useful in ventricular size monitoring because of radiation and/or need for anesthesia. Also, invasive ICP monitoring is not the solution. The best option for monitoring, except for direct biparietal, fronto-occipital and circumferential measurement on daily basis, ultrasound monitoring, also with Doppler sonography, is best practical option [19].

In surgery indication is necessary bear in mind that the young age and the small blood mass of the patients is not in favor of a long surgical intervention.

There are two main options for surgery. Ventriculoperitoneal shunt insertion is a standard hydrocephalus therapy, bearing higher risk of failure due to obstruction, infection, mechanical failure. As surgery is done on new-borns, up to adulthood, with exclusion shunt failure or infection, revision will be necessary at least due to patient's growth.

If ventriculoperitoneal drainage is indicated, ventricular catheter may be inserted into the frontal, occipital horn or trigonum in supposed non-dominant hemisphere. Site of insertion depends on surgeons' preferences. The shorter is extracranial segment of drainage, the smaller is probability of ventricular catheter dislocation outside ventricle with the head growth. Occipital horn insertion is associated with less probability of catheter obstruction by chorioid plexus with sufficient length of catheter placed intraventricularly. Frontal horn allows efficient portion of catheter to be inserted in ventricle with easy positioning for ventriculoperitoneal shunt, without need of patient repositioning during surgery. A little higher incidence of obstruction by chorioid plexus is associated with frontal horn drainage. Ventricular trigone allows shorter intraventricular length of catheter, but also shortest extracranial portion of ventricular catheter.

Ventriculoperitoneal shunt is completed in standard manner. Curvilinear incisions are preferred for exclusion contact points between parts of shunt system (catheters, valve, antisiphon unit) and edges of wound to minimize risk of infection and relieve pressure on the skin suture. In new-born non-toothed forceps for skin manipulation is preferred, and reduced coagulation prevents necrosis and wound healing problems including infection. Ventricular catheter insertion should be done after distal part is prepared for insertion. That means, valve with antisiphon unit are in retroauricular subcutaneous pocket connected with connected peritoneal catheter, which is tunneled to the paraumbilical incision, where intraperitoneal approach is prepared at least, up to dorsal fascia. After ventricular catheter is inserted and tunneled with shunt passer into the retroauricular pouch, then connected with valve and all connection are secured with silk tie. Before intraperitoneal insertion verification of functional system must be done. Both cranial wounds are closed in layers.

There are others, less used, insertions of distal catheter. It can be inserted into the venous system through tributaries of external jugular vein. In this approach, any shunt infection is associated with sepsis.

Prophylactic antibiotics are indicated.

In the case of contraindication of longer surgery, or any pathology, which prevents intraperitoneal insertion, external ventricular drainage is an option or subcutaneous reservoir (Omay, Rickham).

Endoscopic third ventriculostomy, because of higher risk of failure before 6 months of age, is an option if revision due to previously inserted drainage is indicated [20].

Chiari II. malformation

Chiari malformation type II. is herniation of hindbrain into the cervical spinal canal and is exclusively associated with spina bifida.

While Chiari II. malformation was not observed during embryonal development and is detectable during foetal stage of development, it is believed to be not primary malformation, but reflexes changed environment due to different CSF pressure gradients in case of open spina bifida [21, 22].

With lower pressure in CSF spaces intracranially, stimulation for posterior cranial fossa enlargement is reduced and posterior fossa neural tissue grows in standard manner. This theory is supported by MOMS study when in utero myelomeningocele closure significantly reduces Chiari II. Malformation incidence [23].

Despite, it is usually asymptomatic (**Figure 10**), when symptomatic, its manifestation is life threatening. Clinical presentations, such as trunk and limbs muscle tonus impairment, dysphagia, stridor and vocal cord palsy, weak cry, central apnea manifest after delivery, during childhood and adulthood or never. According to present information, there is no correlation between presenting clinical symptoms and severity of anatomical severity compression on MRI.

Most accepted theory, why herniation occurs is disturbance of CSF pressure gradient between intracranial and spinal part of CSF compartment due to CSF leakage through neural tube defect.

Applying this theory in symptomatic Chiari malformation, type 2, the reasonable treatment is by lowering ICP.

As a primal type of surgery ventriculo-peritoneal shunt is usually applied in older children and adults.

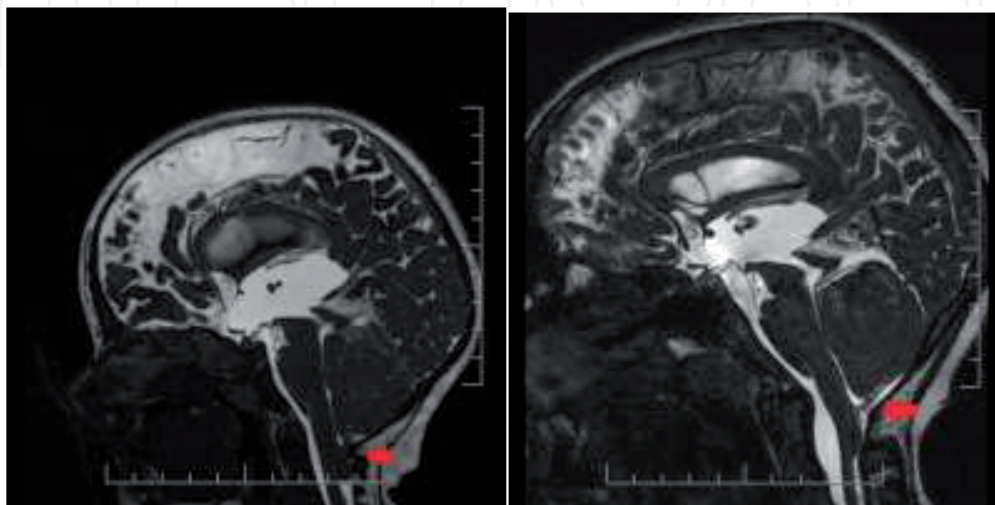


Figure 10.
Chiari II. Malformation identified at the time of myelomeningocele closure on left picture (red arrow points to herniation) resolves completely four years after surgery – Right picture.

New-born and infants are usually treated primarily with external ventricular drainage with later internalization when effectiveness of ICP lowering on clinical symptoms improvement was proved.

In the cases of persisted clinical symptoms cervico-cranial decompression comes to role. Despite foramen magnum is enlarged, suboccipital craniectomy with preferred laminoplasty over laminectomy on upper cervical vertebrae relieve compression on neural structures. Extend of cervical decompression is planned according preoperative MRI. Torcular identification on MRI and bearing in mind its localization prevents massive intraoperative bleeding with possible menacing consequences, when dura is opened, despite there no consensus about necessity of duroplasty [24].

Because spinal and cranial dura lacks elasticity consisting mostly of collagen fibers duroplasty is recommended.

Because of distorted anatomy associated with high risk of neural or vascular damage, arachnoideal dissection to explore the fourth ventricle and normal spinal cord identification is reserved only for cases with syringomyelia [25].

Artificial dura is used to complete duroplasty with sufficient space for neural content and CSF. Dural sealants reduce CSF leakage. Wound is closed in layers.

Approximately one tenths of patients require reoperation. Reason for recurrent compression can be extensive epidural scaring or new bone formation.

3. Conclusions

The treatment of spina bifida is a complex problem that requires multidisciplinary treatment not only at the time of surgery, but also often requires lifelong medical follow-up and therapy.

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Conflict of interest

Author has no conflict of interest.

Acronyms and Abbreviations

CSF	cerebrospinal fluid
ICP	intracranial pressure
VPS	ventriculoperitoneal shunt (VPS)
DST	dermal sinus tract (DST)
SCT	spinal cord tethering
SB	spina bifida
SBO	spina bifida occulta
SCM	split cord malformation
AFP	alpha fetoprotein (AFP)
OSD	occult spinal dysraphism

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