



# The Indispensable Role of Intraoperative Neurophysiological Monitoring in Tethered Cord Release Surgeries

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## INTRODUCTION

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Untethering of the spinal cord is performed to surgically treat abnormal spinal cord fixation, which tends to arise from conditions such as spina bifida, spinal dysmorphisms (including diastematomyelia, lipomyelomeningocele, and myelomeningocele), trauma, or tumors. This fixation often leads to complications known as Tethered Cord Syndrome (TCS), where the spinal cord becomes abnormally anchored, resulting in restricted mobility and progressive neurological deficits [1]. Surgical untethering aims to prevent further damage or potentially reverse symptoms, with intraoperative neuromonitoring (IONM) playing a vital role in distinguishing functional neural tissue from fibrous tissue to reduce the risk of nerve damage [2].

Spina bifida encompasses a range of neural tube defects. Myelomeningocele, an open defect, involves both spinal cord and meningeal herniation through vertebral defects, leading to severe neurological impairment and potential TCS development. Spina bifida occulta (SBO), a closed defect, is less severe but may contribute to TCS when associated with other anomalies, such as a thickened or fatty filum terminale [3]. Tethered cord conditions can also develop due to trauma or tumors, requiring surgery to free the spinal cord from surrounding attachments. TCS leads to stretching nerve roots and the lower spinal cord, resulting in hypoxic-ischemic damage and progressively worsening motor, sensory, or sphincter function [4]. Surgical untethering is the primary intervention to halt further decline. Intraoperative neurophysiological neuromonitoring (IONM) is critical for preserving neural function, enabling safe dissection by identifying ambiguous neural tissue through mapping and assessing motor, sensory, and reflex pathways, such as the bulbocavernosus reflex (BCR) [5].

IONM provides real-time neuromonitoring modalities to distinguish nerve fibers from nonfunctional tissue. Mapping helps surgeons safely cut fibrous tissue that tethers the spinal cord. Given congenital malformations and chronic inflammation, which alter tissue anatomy and appearance, IONM is indispensable in guiding safe and precise untethering [6]. Magnified imaging may fail to differentiate

between fibrous and neural tissue and relying solely on observable gross features may profoundly mislead the neurosurgeon. Fibrosis and other pathological impacts distort tissues' morphological features and significantly shift anatomical positioning, emphasizing the critical value of neuromonitoring in untethering procedures [7].

Common complications of untethering surgery include worsening motor, sensory, or sphincter function, often due to nerve root manipulation. There is also a risk of retethering, where scar tissue reattaches the spinal cord, and CSF leaks, potentially leading to infections like meningitis. IONM helps minimize these risks by protecting functional nerve roots and guiding dissection [8].

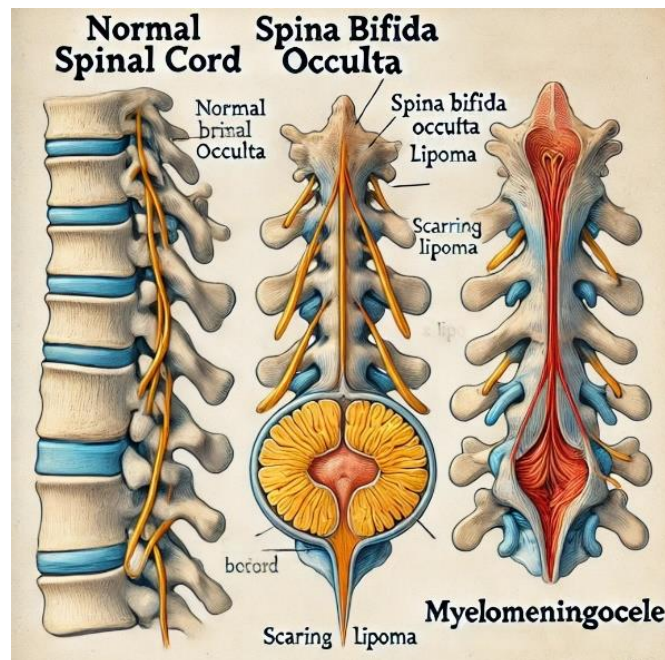
Neural tube defects can be categorized as open or closed, depending on the exposure of neural tissue. Open defects, such as meningocele and myelomeningocele, involve the protrusion of meninges or spinal cord and meninges, respectively, leading to significant impairment. On the other hand, closed defects, like lipomeningocele and lipomyelomeningocele, entail the presence of fatty masses and sometimes a herniated spinal cord, resulting in tethering. Additionally, spinal cord malformations like split cord malformation (SCM) involve dividing the spinal cord into two hemicords. In Type I SCM, the hemicords are enclosed in separate dural sheaths, separated by a bony spur, while in Type II, the hemicords are within a single dural tube split by fibrous bands [2]. SCM often leads to tethered cord syndrome (TCS) due to tethering by a bony or fibrous septum. Lipomyelomeningocele, another closed defect, involves the herniation of the meninges and spinal cord, with a fatty mass contributing to TCS. These malformations often present with symptoms such as scoliosis, leg asymmetry, and urinary dysfunction, necessitating untethering surgery. Furthermore, lipomas in various locations, such as the conus medullaris, tether the spinal cord to surrounding tissues, requiring precise surgical intervention [3].

During untethering surgery, multimodal IONM involves the use of electromyography (EMG), somatosensory evoked potentials (SEP), BCR, and transcranial motor evoked potentials (TCeMEP). EMG monitors muscle activity in the lower limbs and sphincters, providing real-time information on neural integrity. SEPs assess sensory pathways by stimulating nerves (ulnar, tibial, pudendal), while BCR monitors sacral reflexes essential for sphincter control [1]. TCeMEP evaluates motor pathway integrity, aiding in protecting neural pathways during surgery.

It is essential to manage anesthesia carefully to ensure the effectiveness of IONM. Muscle relaxants should be limited to intubation to avoid interference with motor mapping. For adults, total intravenous anesthesia (TIVA) using propofol and fentanyl is preferred over inhalational agents such as isoflurane and sevoflurane, as the latter can disrupt SEP and MEP signals [6]. In pediatric patients, especially those under six years old, reducing propofol and incorporating adjuncts like ketamine can improve MEP monitoring, particularly since the corticospinal pathways are not fully developed in this age group. This approach enhances the success of neuromonitoring in pediatric patients [2].

EMG monitoring is crucial in untethering surgeries, evaluating neural integrity, and guiding precise dissection. Both free-running (s-EMG) and triggered EMG (t-EMG) provide real-time feedback on muscle

activity, ensuring optimal monitoring of neural function. Spontaneous EMG activity is characterized by patterns such as motor unit potentials (10-15 Hz), neurotonic discharges (50-200 Hz), and fibrillations (1-5 Hz), each indicating specific nerve activity. At the same time, movement artifacts remain distinguishable due to their irregularity [8]. Triggered EMG is critical for identifying motor nerve roots during untethering, distinguishing these from fibrous tissue to facilitate accurate and safe tethered cord release. Gradual increases in stimulation—ranging from 0.01 mA to 3 mA at a frequency of 1 Hz and a pulse duration of 200  $\mu$ s are employed to evoke compound muscle action potentials (CMAP) [8]. A stimulation threshold below 15 mA typically indicates direct nerve activation, whereas higher thresholds point to non-functional structures.



**Figure:** Diagram of a normal cord, spina bifida occulta, and myelomeningocele.

The bulbocavernosus reflex (BCR) is an essential aspect of untethering procedures to assess sacral nerve function and sphincter control. This reflex is triggered by stimulating the dorsal penile or clitoral nerve with a 0.5 ms pulse at 40 mA and recording the reflex from the anal sphincter [6]. Temporal summation can amplify the reflex response by using a series of 2-5 stimuli with an interstimulus interval of 3 ms or a double train with an intertrain interval of 75-250 ms. It is vital to avoid inhalational anesthetics during BCR monitoring to maintain accuracy, as they can diminish reflex responses. When stimulating the filum terminale, a significantly higher stimulus intensity is required than nerve roots due to its lack of neural elements. Care must be taken when applying a supramaximal stimulus to prevent activation of surrounding structures and ensure accurate testing of the filum [4].

Post-untethering outcomes show that most patients exhibit neurological function improvement or stabilization, particularly pain relief [4]. Retethering rates are generally lower in split cord malformations (SCM) than in lipomyelomeningocele [7]. Scoliosis over 40 degrees necessitates spinal stabilization in most cases, and urinary symptoms stabilize in the majority post-surgery [7]. IONM proves highly effective in untethering surgeries, with TCeMEP showing a sensitivity of 75-100% and free-running EMG demonstrating 87.5% sensitivity and 83.3% specificity [2]. SEPs are more challenging to monitor in young children but retain high specificity (94.7-100%) [1].

## Conclusion

Effective collaboration among the neurosurgeon, anesthesiologist, and IONM team is crucial for successful surgical outcomes. Clear surgical goals, proper anesthetic protocols, and proactive responses to neurophysiological signal changes are critical. The use of multimodal IONM unquestionably assists the neurosurgeon in accurately identifying nerve roots during untethering, resulting in a remarkable reduction in neurological deficits by up to 50%. While the benefits are substantial, applying IONM is not standard for all untethering surgeries; instead, its use is selective, with each case undergoing a comprehensive risk-benefit assessment to enhance surgical precision and patient outcomes.

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