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Current Concept Review

Absent Baseline Intraoperative Neuromonitoring Signals Part 2: Neuromuscular Scoliosis

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Abstract:

Transcranial motor evoked potentials (TcMEPs) increase sensitivity and reliability of signals in neuromuscular scoliosis (NMS) patients and are safe to perform even in patients with epilepsy. Baseline neurologic and functional status, including weight-bearing ability and motor function, are associated with ability to achieve baseline signals in NMS. Supratentorial brain lesions such as focal periventricular leukomalacia, hydrocephalus, and/or encephalomalacia may affect IONM signal acquisition. When baseline signals are absent, troubleshooting can identify potentially modifiable anesthetic or technical neuromonitoring factors. In patients with hydrocephalus, alternative wider electrode placement may facilitate successful TcMEP acquisition.

Families need to be counseled preoperatively that without IONM signals, risk of neurologic deficit increases to 5%. This can compromise quality of life for patients and caregivers, as loss of protective sensory function increases risk of pressure ulcers. Loss of bowel and bladder function, including development of a neurogenic bladder or bowel, can require use of catheterization. A clear plan should be formulated preoperatively to determine whether to proceed with surgery if IONM signals are absent or unreliable.

Key Concepts:

- Intraoperative neuromonitoring with TcMEPs and SSEPs is recommended during spinal fusion for neuromuscular patients, including severely affected GMFCS IV and V spastic quadriplegic CP patients with epilepsy.

- Presence of supratentorial brain lesions, antiepileptic use, and baseline neurologic and functional status, especially weight-bearing ability and baseline motor function, are associated with ability to achieve signals.
- Preoperative evaluation and discussion with family is imperative for decision-making when IONM signals are absent and include anticipated risks of curvature progression without surgery versus 5% risk of neurologic deficit from surgery.
- Loss of protective sensory function increases risk of pressure ulcers, while loss of bowel and bladder function can cause development of a neurogenic bladder or bowel, which can be detrimental to quality of life.

Introduction

There is robust data supporting the use of multimodal intraoperative neurologic monitoring (IONM) for adolescent idiopathic scoliosis (AIS). Although the literature is somewhat more limited in neuromuscular scoliosis (NMS) patients, the data does support use of IONM for these patients, including severely affected spastic quadriplegic cerebral palsy (CP) patients with Gross Motor Function Classification System (GMFCS) scores of IV and V.

Previously, there was theoretic risk that TcMEPs could potentially stimulate seizures in patients with epilepsy, and therefore TcMEPs were generally avoided in NMS patients. However, recent studies demonstrate that TcMEPs do not trigger seizures and can be used safely in patients with epilepsy.[1-3] Salem et al. found that IONM with TcMEPs did not trigger intraoperative or postoperative seizures in epilepsy patients and were not associated with deterioration in seizure control.[3] The American Clinical Neurophysiology Society has recommended that a history of epilepsy should not be considered a contraindication to MEP stimulation.[4]

Presence of Signals

Despite the fact that NMS carries the highest risk of significant intraoperative IONM changes amongst all spinal deformity patients approaching 10%,[5] there remains some controversy regarding use of IONM in NMS, as presence of signals is variable. Previously, only somatosensory evoked potentials (SSEPs) were utilized with responses often absent at baseline or potentially unreliable.[6,7] In a review of 101 patients with NMS, Ashkenaze et al. determined that reliable tracings could not be obtained in 28% of neuromuscular patients.[6] This is likely due to poor cortical bone, intracranial abnormalities such as hydrocephalus, or anesthetic agents suppressing cortical signals. Furthermore, some antiepileptic medications, such as carbamazepine, phenytoin, and valproate, have been shown to affect sensory evoked potentials.[8,9]

Although Owen et al. stated that a single channel cortically based SSEP is unreliable in 27% of patients, the reliability improves to 92% of 187 NMS patients when using both cortical and subcortical recordings of SSEPs.[10] Subcortical SSEPs allow faster feedback and are significantly less sensitive to anesthetic depression.[11]

The use of TcMEPs has become more widespread in NMS[12-14] after it was shown that TcMEPs do not trigger seizures in epilepsy patients.[1-3] DiCindio et al. performed multimodal monitoring in 68 NMS patients and found that 100% of patients with mild or moderate CP had recordable SSEPs and TcMEPs, versus 70% SSEP and 90% TcMEP rate in severe CP, and 86% in non-CP etiologies of NMS.[1]

Baseline neurologic function and functional status, especially weight-bearing ability and motor function, are associated with ability to achieve baseline signals in NMS.[1,5,15] Additionally, Mo et al. reviewed cerebral anatomic factors that could predict success of obtaining IONM signals in CP patients. They

found that supratentorial brain lesions may affect IONM signal acquisition in patients with CP; focal periventricular leukomalacia (PVL), hydrocephalus, and encephalomalacia are significant predictors of lack of interpretable signals on multivariate analysis, especially lack of TcMEP.[14]

Preoperative Considerations

Preoperative clinical evaluation and communication are essential in the neuromuscular patient population. Since baseline neurologic and functional status correlate with ability to obtain IONM signals, these factors need to be carefully evaluated preoperatively in all NMS patients. A careful history should be conducted with the primary caretaker since the physical exam can wax and wane in neuromuscular patients. Weight-bearing ability, both with and without a gait aid and usable motor and sensory function, should be recorded. Clear documentation of baseline bowel and bladder function is essential. Many nonambulatory neuromuscular patients are incontinent at baseline with bladder spasticity, typically voiding spontaneously with need for diapering. Continence correlates with gross motor involvement, while nearly all GMFCS I and II patients are continent, this ability decreases with more significant gross motor involvement, as only 11%–16% of GMFCS V patients are continent.[16,17]

Any known supratentorial brain lesions that could affect signal acquisition should be identified on any prior images, including PVL, hydrocephalus, and/or encephalomalacia. Use of Antiepileptic medications that can affect signals should be reviewed, such as carbamazepine, phenytoin, and valproate.[8,9]

During the preoperative visit, it is essential for the surgeon and family to formulate and agree upon a clear plan to determine whether to proceed with surgery or not if IONM signals are absent, unreliable, or are lost during the case. In contrast to neurologically normal children with adolescent idiopathic scoliosis, further investigation into the cause of IONM signals that are unobtainable will usually be fruitless. Families should understand that it is typically impossible to further optimize IONM signals.

Troubleshooting Absent Signals

Neuromuscular scoliosis patients should have baseline SSEP and TcMEP neuromonitoring responses attempted prior to positioning patients prone for surgery. These signals should also be obtained before applying any traction. When baseline IONM signals disappear with traction, then weight should be removed, and signals reassessed. If signals return, then traction should be avoided or carefully titrated intraoperatively to allow for signal capture, as IONM signals are prioritized over traction.

Multimodal IONM requires coordination between the spine surgeon, neuromonitoring technician, electrophysiologist, and anesthesiologist. Walker et al. recently published neuroanesthesia consensus guidelines for optimizing TcMEPs,[18] which can be applied to NMS patients.

In patients with hydrocephalus, alternative electrode placement may facilitate successful TcMEP acquisition. Mo et al. stated that enlargement of the ventricles may displace cortical and subcortical pathways and adversely affect motor pathways after TcMEP voltage stimulation. Furthermore, conduction through the cerebrospinal fluid in the enlarged ventricles may be ineffective. Wider electrode spacing may allow the signal from TcMEP stimulation to reach the displaced pathways.[14] Additionally, in neuromuscular patients, ensure that both cortical and subcortical recordings are utilized for SSEPs with multiple recording sites, as single channel cortical signals are potentially unreliable.[10,19,20]

Decision-Making with No Signals

The rate of postoperative neurologic deficit in neuromuscular patients, based on Scoliosis Research Society Morbidity and Mortality data, is approximately 1%.[21,22] Without neuromonitoring, Thuet et al. demonstrated that risk of postoperative neurologic deficit increases more than six-fold, from 0.77% to 5.08%.[15]

Spinal cord injury can result in neurogenic bowel and bladder, with loss of ability to void and defecate spontaneously. Previously incontinent patients can no longer just be diapered; clean intermittent catheterization

(CIC) is generally the preferable method of bladder drainage, typically required several times daily to prevent urinary distention.[23,24] CIC can be poorly tolerated in neuromuscular patients, as spasticity and adductor contractions cause difficulty with positioning and visualization of the meatus.[25] However, indwelling catheters (urethral catheters or suprapubic tubes) can cause additional complications such as urinary tract infections, urinary tract fistulae, strictures, stones, and even cancer.[24,26,27] A neurogenic bowel can likewise deteriorate quality of life, typically requiring rectal therapy for emptying, enemas, and surgical interventions.

Parents of patients with neurogenic bowel and bladder have reported these issues to be more stressful than motor disability in conditions such as spina bifida.[28] Without adequate treatment, serious complications can occur, such as renal failure, urosepsis, and even death. Upper urinary tract dysfunction from neurogenic bladder was previously the top cause of mortality following spinal cord injury, due to recurrent urinary tract infection and renal failure.[27] Mortality rates have dramatically improved in recent years, but neurogenic bladder still confers high morbidity and greatly impacts quality of life and psychosocial well-being of patients and caregivers.

Spinal cord injury can also compromise protective sensory function in neuromuscular patients, increasing risk of decubital ulcers. Recurrent ulcers with need for wound care and other interventions adversely affects quality of life. Although the flaccidity resulting from iatrogenic spinal cord injury is often not overtly detrimental to GMFCS V spastic patients, the loss of ability to void and defecate spontaneously, and also the loss of protective sensory function, can be devastating.

At induction, if SSEPs are absent but TcMEPs are present, it is likely reasonable to proceed. If SSEPs are normal but TcMEPs are absent or irregular despite troubleshooting, then it also is likely reasonable to proceed, especially if the patient has a large magnitude curve with no meaningful lower extremity motor function. However, if there are no SSEPs or TcMEPs at induction, then the decision should be based on careful preoperative discussion with the family regarding a 5% risk of neurologic injury. The ability to perform a Stagnara wake-up test, depending on baseline cognitive and motor function, may increase the safety profile of proceeding with surgery. These decisions have lifelong implications and should be patient-focused and individualized.

Many patients with NMS can have labile blood pressures during surgery for a variety of reasons. Many GMFCS IV and V patients have smaller amounts of blood as a result of lower relative amounts of muscle mass and they are often relatively hemo-concentrated from baseline dehydration. Even though a patient may start with a hemoglobin of 15 g/dl, the combination of rehydration with IV fluids and large volumes of blood loss can quickly lead to anemia and hypotension which effects IONM. As such, the team has to be vigilant on resuscitation with blood products as the case evolves. It is not uncommon that at the end of a long case that IONM signals drop significantly. As such, the team should have discussed this possibility with the parents and come to a decision on whether to continue the case. It is often wise to leave the operating room and directly discuss with the family that despite a successful instrumentation and correction, the IONM have decreased, and it is difficult to predict any morbidity. Shared decision-making is needed to decide whether to continue the case with the understanding the IONM signals will likely improve with resuscitation and without any sequalae or to remove the instrumentation and hope that this will improve the signals with a later operation to finish the correction. The latter choice brings certain morbidity for risk of infection and other complications that a frail child may experience as a result of two large spine operations.

Summary

The use of IONM is recommended during spinal fusion for neuromuscular patients, including severely affected GMFCS IV and V spastic quadriplegic CP patients with epilepsy (Table 1). Weight-bearing ability, baseline motor function, supratentorial brain lesions, and antiepileptics are associated with ability to achieve baseline signals.

Table 1. Summary of Preoperative and Intraoperative Decision Making for Absent Neuromonitoring Signals, with Associated Risks of Absent Signals and Decision-Making

Preoperative Considerations	Intraoperative Troubleshooting	Risks of Absent Signals	Decision-Making
Careful history and exam: • Weight-bearing ability • Usable motor/sensory function • Bowel/bladder function Risks for absent signals: • Nonambulatory • Severe motor impairment • Antiepileptic medications • Supratentorial brain lesion: ◦ Hydrocephalus ◦ Focal periventricular leukomalacia ◦ Encephalomalacia Discussion with family: • Potential consequences of iatrogenic neurologic deficit • Develop clear plan for how to proceed if signals are absent	Surgical: • Remove traction Anesthetic: • Blood pressure/volume management • Medications – refer to Part 1: AIS Technical: • Neuromonitoring systematic troubleshooting • Utilize both cortical and subcortical recordings for SSEP • Hydrocephalus: wider electrode spacing for TcMEP stimulation	Absent IONM signals: • Neurologic deficit risk 5% • Potentially lower if patient can comply with Stagnara wake-up test Sequelae of neurologic deficit: • Flaccidity • Loss of protective sensory function ◦ Decubitus ulcers • Neurogenic bowel & bladder impairs quality of life ◦ Bladder: requires catheterization (intermittent vs. indwelling) ◦ Bowel: meds, rectal therapy for emptying, enemas ◦ Potential for serious complications &/or surgical intervention	Balance between anticipated curvature progression & sequelae without surgery VERSUS risk of neurologic deficit from surgery Absent SSEP, present TcMEP: • Likely reasonable to proceed Present SSEP, absent/irregular TcMEP: • May be reasonable to proceed, especially if: ◦ Large curve ◦ No meaningful lower extremity function ◦ Bowel/bladder incontinence ◦ Underlying supratentorial brain lesion Absent/irregular SSEP and TcMEP: • **Decisions based on plan discussed with family preoperatively** • Consider proceeding carefully if minimal functional loss would be expected in the setting of neurologic injury

Without reliable IONM signals, the decision to proceed should be based on careful preoperative discussion with the family regarding anticipated consequences of curvature progression versus 5% risk of neurologic deficit from surgery, potentially causing neurogenic bowel and bladder or loss of protective sensory function.

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