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Short Communication Open Access

Neuromonitoring in Neuromuscular Scoliosis

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Short Commentary

Neuromuscular disease was first described in 1836 by Conte and is accompanied by spine deformity [1] in 60% to 75% of quadriplegic cerebral palsy children, 90% of spina bifida children (above the sacral level), and nearly 100% of Duchenne muscular dystrophy (DMD) children, who have not been treated with long term deflazacort glucocorticoid [2-4]. Neuromuscular scoliosis (NMS) presents earlier than idiopathic scoliosis (IS), and is progressive because of the abnormal biomechanical loading of the spine due to muscular imbalance and asymmetrical, Heuter-Volkmann induced growth of immature spinal vertebrae [5]. Anticipation is regarded by DiMeglio as a very successful method of managing NMS scoliosis [6], and while orthoses may be used indefinitely to treat children with mild cerebral palsy or alternatively to maximize the nonoperative management of sitting ability and postural care in children with severe scoliosis, bracing does not alter progressive neuromuscular deformities that are ≥ 20° [7]. Iatrogenic spinal cord injury remains one of the most devastating complications of neuromuscular spine deformity surgery. The incidence of neurological complications in NMS scoliosis, varies from 0.5% to 4.6%, and is higher than that in IS (0.5% to 0.72%) [8,9]. Higher intraoperative blood loss that compromises spinal cord vascularity, in combination with distraction techniques, that are occasionally adopted to address the severest and stiffest neuromuscular deformities, may account for this discrepancy [10,11].

Neuromonitoring was introduced by Nash et al. in 1977, and monitors the function of the spinal cord [12]. Prior to its introduction, the Stagnara wake up test was the only method of detecting spinal cord injury[13], and while still regarded as the standard to assess global motor function, this test is not always practical in NMS patients who have either intellectual disabilities, muscle weakness or both. In addition, an ischemic spinal cord injury may not present immediately following a correctional maneuver, and the patient may be able to move the lower extremities voluntarily at the time of the wake up test, only to demonstrate paralysis on emergence from anesthesia. In contrast, neuromonitoring provides a continuous means of assessing spinal cord integrity and offers early detection of reversible neurophysiological dysfunction that enables prompt intervention to prevent permanent neurological deficit. MacEwen et al. found that the recovery of a neurological deficit is directly proportional to the speed of removal of malpositioned instrumentation [14].

Spinal cord monitoring consists of somatosensory evoked potentials (SSEPs), transcranial electric motor evoked (MEPs), and H reflexes. Intraoperative monitoring using somatosensory evoked potentials (SSEPs) alone is inadequate for monitoring the descending spinal cord motor tracts or the spinal gray matter, as SSEPs are mediated by the posterior sensory column of the spinal cord [15]. Transcranial electric motor evoked (MEPs) potentials are an effective and clinically practical way to monitor spinal cord motor function in real time during corrective spine surgery [16]. Schwartz et al. reported that transcranial MEPs were 100% sensitive in detecting evolving neurological injury, whereas SSEPs were only 43% sensitive [17]. In addition to better sensitivities, transcranial MEPs detect emerging spinal cord motor injury at an average 5 minutes earlier than SSEPs [17]. The differential sensitivities of transcranial MEPs and SSEPs to evolving spinal cord injury are thought to be related to the vascular supply of the motor pathways. The anterior horn motor neurons within the spinal cord and the spinal motor interneurons have a high metabolic rate, and are vulnerable to vascular insult. Since most neurological injuries during deformity surgery are thought to be ischemic in nature, transcranial MEPs are more likely to change first during these corrective maneuvers than SSEPs [17]. Transcranial MEPs have been previously demonstrated to be reliable in identifying cord ischemia during abdominal aortic aneurysm repair and spinal operations [5].

Spinal cord monitoring in neuromuscular patients is variable and reflects the altered neural pathways. Single channel somatosensory evoked potentials (SSEPs) are unreliable in 16% to 28% of NMS patients, and etiology, anesthesia, blood pressure, and temperature are known to influence the quality of the SSEP tracings [18,19]. While it is challenging to consistently obtain reliable tracing in NMS patients, a decline in the amplitude of 50% of the initial baseline reading is significant, and associated with a definitive risk of spinal cord injury [15,17]. Hammett et al. evaluated 66 patients with cerebral palsy, and reliable baseline SSEPs were obtained in 88% of patients [20]. Dicindio et al. reviewed 68 patients with neuromuscular disorders and found that the reliability of the SSEP recordings in cerebral palsy was related to the severity of the condition, with reproducible SSEP potentials in 100% of patients with mild and moderate cerebral palsy, and only 70% of those with severe involvement [21]. Charcot Marie Tooth (CMT) is also associated with low reproducible SSEP tracings (50%) [19].

In contrast, Duchenne Muscular Dystrophy (DMD) (87%) and Polio (73%) have more consistent SSEP recordings, with Sewell et al. successfully obtaining SSEP tracing in 98% of their 99 NMS patients (55 DMD, 30 Spinal Muscular Atrophy, SMA and 14 miscellaneous) [19,22]. Table 1 summarises the percentage of NMS patients with monitorable SSEP at baseline.

As a result of this unreliability, Ashkenaze, Mudiyam and Boachie-Adjei recommended the introduction of alternative monitoring techniques such as subcortical, epidural and MEPs in neuromuscular patients [19]. Owen et al. found that the use of multiple recording SSEP sites, in combination with MEPs, was associated with reliable

Type of Scoliosis	SSEP
CP mild/ moderate	100%
DMD	87%
Polio	73%
CP severe	53-70%
CMT	50%

CMT: Charcot Marie Tooth, CP: cerebral palsy, DMD: Duchenne muscular dystrophy, Polio.

Table 1: Summarises the percentage of NMS patients with monitorable SSEP at baseline.

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responses in 96% of NMS patients [23]. The remaining 4% of patients with unrecordable tracings had demonstrated severe motor deficits (paraplegia) before surgery. The importance of obtaining neuromonitoring in the most severely deformed, dysfunctional and mentally impaired patients should not be underestimated. Spinal and sitting balance should only be achieved without further neurological deficit and without the risk of decubitus ulceration, and monitoring of the brachial plexus is of critical importance to those who may be totally dependent on their arms for activities of daily living.

Finally, concern over the perceived potential to initiate epileptic seizures has precluded many authors from the routine use of transcranial MEPs in NMS. However, Salem et al. recently demonstrated that transcranial MEPs do not trigger intraoperative nor postoperative seizures in NMS patients undergoing posterior spinal fusions, nor did they demonstrate deterioration in seizure control of epileptic patients [24].

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