ORIGINAL ARTICLE

Multimodality Intraoperative Neurophysiological Monitoring (IONM) with Selective Dorsal Root Rhizotomy For patients with Cerebral Palsy

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ABSTRACT

Selective Dorsal Root Rhizotomy (SDR) is a neurosurgical procedure aimed at alleviating spasticity in cerebral palsy patients by selectively severing sensory nerve rootlets in the spinal cord. This process helps reduce spastic muscle activity while preserving motor function. The use of multimodal intraoperative neurophysiological monitoring (IONM) techniques—such as Somatosensory Evoked Potentials (SSEP), Transcranial Motor Evoked Potentials (TCeMEP), and Electromyography (EMG)—has significantly enhanced surgical precision, leading to improved outcomes. Long-term studies report enhanced motor function and quality of life, with reduced reliance on further treatments, positioning SDR as a valuable option for managing spastic cerebral palsy. This literature review explores the evolution of SDR, its clinical effectiveness, patient selection criteria, and long-term outcomes for spasticity management in CP. The review consolidates findings from various studies to assess the impact of SDR on motor function, complications, and quality of life while comparing it with alternative spasticity management options. By synthesizing the current literature, this review aims to offer a comprehensive perspective on the role of SDR in enhancing functional outcomes for children and adults with CP.

INTRODUCTION

Cerebral Palsy (CP) is a broad term that covers a range of neurological disorders affecting movement, posture, and motor function. CP stems from non-progressive interference, lesions, or abnormality in the developing brain, impacting motor and developmental skills, affecting 2–3 infants per 1,000 live births in the United States. The highest incidence occurs in babies with birth weights ranging from 1000 grams to 1499 grams (59.18/1000 live births) and in preterm babies born before the 28th week of pregnancy (111.8/1000 live births). Research suggests that preterm males are more susceptible to white matter injury and intraventricular hemorrhage than females, potentially leading to a higher prevalence of CP among males. Cerebral palsy can result from brain injury before the completion of cerebral development during prenatal, perinatal, or postnatal periods, as cerebral development continues during the first two years of life. 70 to 80 percent of CP cases are acquired prenatally and largely have unknown causes. Brain hypoxia during birth or the perinatal period is a common mechanism of brain injury contributing to approximately 6% of cases of spastic cerebral palsy [1].

Spastic cerebral palsy is a form of CP that impacts the cerebral cortex, particularly the corticospinal (pyramidal) tracts of the developing brain. This results in hypertonic muscles in children. Depending on which part of the developing brain is affected, patients may display symptoms of spastic hemiplegia, spastic diplegia, spastic quadriplegia, or monoplegia. In addition to motor symptoms, children with CP often experience cognitive and sensory impairments, epilepsy, and nutritional deficiencies [2]. Early diagnosis of CP enables early implementation of medical interventions to optimize motor and cognitive functions, taking advantage of brain plasticity, preventing secondary lesions, and improving the child's quality of life. Treatment of spasticity involves systematic rehabilitation and pharmacotherapy using Gamma Amino Butyric Acid (GABA) analogs for generalized spasticity. Botulinum toxin (Botox) injections are used to alleviate focal spasticity, and physiotherapy or surgical interventions may also be utilized. Neurosurgical procedures include continuous intrathecal infusion of Baclofen through a pump, selective peripheral neurotomy, and selective dorsal root rhizotomy.

SDR originated as an approach to reduce hyperreflexia in patients with spastic conditions. The procedure involves the selective severing of dorsal sensory nerve roots, reducing afferent input to the spinal cord and decreasing motor neuron hyperactivity [3]. Initially introduced in the early 20th century, SDR underwent various modifications to enhance its safety and efficacy. In the 1970s and 1980s, the modern technique of selective sectioning was refined, mainly through the work of Fasano and Peacock, who emphasized the importance of selective cutting to minimize sensory deficits while maximizing reduction in spasticity [4].

Improvements in intraoperative monitoring and patient selection criteria drove the widespread adoption of SDR in the 1980s and 1990s. Electromyographic (EMG) monitoring during surgery allowed for more precise identification of the nerve rootlets contributing to spasticity, reducing the risk of adverse effects such as muscle weakness or sensory loss [5,6]. These advancements established SDR as a viable option for reducing spasticity in selected patients with CP.

Selective Dorsal Rhizotomy (SDR) is a promising treatment involving dissection of dorsal nerve roots while preserving muscle strength and function. When combined with a multimodality Intraoperative Neurophysiological Monitoring (IONM) protocol utilizing Somatosensory Evoked Potential (SSEP), Transcranial Motor Evoked Potential (TCeMEP), Electromyography (EMG), and Train of Four (TOF), SDR has shown encouraging results. Generally, the use of IONM has been linked to improved postoperative outcomes and significantly fewer neurological complications in spinal surgeries [6]. This study explores the beneficial application of IONM in SDR for the treatment of spastic cerebral palsy.

METHODS

Patient Selection:

Patient selection is crucial for the success of SDR. Not all children with CP are suitable candidates; careful screening is necessary to ensure optimal outcomes. The ideal candidates are typically those with spastic diplegia, good cognitive function, and adequate strength in their lower extremities (Park et al., 2013). Additionally, candidates should not have significant dystonia or athetosis, as these conditions are less responsive to SDR and may result in suboptimal outcomes [7].

A multidisciplinary evaluation involving neurologists, orthopedic surgeons, and physiatrists is recommended to determine a child's suitability for SDR. Preoperative assessment often includes gait analysis, MRI imaging to assess the integrity of the brain and spinal cord, and physical therapy evaluation to determine muscle strength and functional abilities [8]. The selection process aims to identify children most likely to benefit from reduced spasticity without experiencing significant adverse effects such as muscle weakness or impaired balance.

Anesthesia requirement:

Total Intravenous Anesthesia (TIVA) with Propofol and Remifentanil should be employed, and no muscle relaxant should be given. A train of four (TOF) is used to monitor the level of muscle relaxation by stimulating the posterior tibial nerve at the medial malleolus and recording from the abductor hallucis muscle in the foot.

Benefits of Intraoperative Neurophysiological Monitoring

Intraoperative neurophysiological monitoring (IONM) plays a crucial role in the success of SDR. The primary methods of IONM used during SDR include Somatosensory Evoked Potential (SSEP), Transcranial Motor Evoked Potential (TCeMEP), Electromyography (EMG), and Train of Four (TOF)EMG monitors the response of individual nerve rootlets to electrical stimulation, helping the surgical team determine which rootlets are contributing most to spasticity. SSEPs assess the integrity of sensory pathways during the procedure, ensuring that critical sensory functions are preserved [8].

The benefits of IONM in SDR are numerous. First, it allows for a more selective approach by identifying which dorsal rootlets are most responsible for hyperactive reflexes, thereby improving the precision of the surgery. This selectivity minimizes the risk of postoperative complications, such as sensory loss or muscle weakness, if too many or the wrong nerve rootlets are severed [5]. Additionally, IONM provides real-time feedback to the surgical team, reducing the likelihood of intraoperative errors and improving overall safety (Park et al., 2013). IONM has been shown to enhance surgical outcomes, leading to a better reduction in spasticity and improved motor function in patients with CP.

Transcranial electrical Motor Evoked Potential (TCeMEP):

Transcranial electrical Motor Evoked Potential (TCeMEP) assesses functional integrity and promptly addresses any damage in the descending corticospinal tracts. Corkscrew electrodes are placed on the scalp at C1, C2, C3, and C4 levels per the international 10-20 system. Subdermal needle electrodes are placed in the upper extremity abductor pollicis brevis and abductor digitiminimi muscles as a control. Electrodes are also placed in the in the lower extremity muscles adductors, quadriceps, tibialis anterior, medial gastrocnemius, abductor hallucis, extensor hallucis brevis, and the anal sphincter. Urinary catheter electrode is used for bladder recordings [9]. Low-frequency filters were set at 10 Hz, while the high-frequency filters were set at 5000 Hz. The inter-pulse stimulation interval was established within a range of 2-4ms. A stimulation rate of 250-500 Hz and a gain of 200 microvolts per division is used. The recording sweep is set at 10ms per division (100ms). The alert criteria for TCeMEP are established as a 70-80% decrease in amplitude from the baseline, a change in the wave morphology, or an increase in the stimulation threshold of 100 volts or more [10].

Somatosensory Evoked Potential (SSEP):

Somatosensory Evoked Potential (SSEP) is an IONM modality utilized to evaluate the sensory or ascending pathways of the nervous system. SSEP is elicited by placing the stimulation electrodes distally on the peripheral nerves of the upper and lower extremities and the recording electrodes along sensory pathways to record peripheral, subcortical, and cortical responses [11]. Stimulation surface adhesive electrodes or subdermal needle electrodes are used to simulate the ulnar nerve at the wrist in the upper extremity and the posterior tibial nerve at the medial ankle in the lower extremity. The anode electrodes were placed 2-3 centimeters distal to the cathode electrode. The low-frequency filters were set at 30 Hz, while the high-frequency filters were set at 500 Hz (cortical) and 1500 Hz (subcortical and peripheral). Recording was performed at multiple locations along the sensory pathway (brachial plexus, popliteal fossa, brainstem, and somatosensory cortex). The recording electrodes are placed at FPz, CPz, CP3, CP4 according to the international 10-20 system for cortical recordings. For cervical recording, electrodes were placed at Cv5, and for peripheral recording at the Erb's point of the popliteal fossa. The recording sweep is set at 5ms per division for the ulnar nerve and 10ms per division for the posterior tibial nerve. The alarm criteria for SSEP are a 10% increase in latency or a 50% decrease in amplitude.

Electromyography (EMG):

An electromyogram (EMG) records the electrical activity of muscles. There are two types of EMG recording: spontaneous (s-EMG) and triggered (t-EMG). The s-EMG passively monitors nerve roots and muscles without stimulation and is sensitive to injury. At the same time, t-EMG is beneficial in identifying, mapping, and evaluating the functional continuity of nerve fibers. EMG is recorded from the same muscles used for TCeMEP. In EMG, subdermal needle electrodes are placed bilaterally in corresponding muscle groups to record compound action potential (CMAP). The lower extremity muscles recorded are adductor longus (L2-L4), vastus medialis (L2-L4), gastrocnemius (S1-S2), tibialis anterior (L4-S1), abductor hallucis (S2-S3), external anal sphincter (S2-S4) and external urinary sphincter (S2-S4) [12]. Low-frequency filters are set to 10 Hz, and high-frequency filters are at 5,000 Hz for EMG. In s-EMG, the time display is set to 300 milliseconds/division; in t-EMG, the time display is set to 10 milliseconds/division. A hand-held monopolar probe is used to perform t-EMG. Furthermore, t-EMG has two stimulation settings: single pulse t-EMG— 2.79 Hz frequency and 200 microseconds duration, and reflex t-EMG— with a frequency of 50Hz, was used with a 200-microsecond duration and train length of one second. The benefit of using single t-EMG is motor root identification, as the intensity of the motor threshold is lower than sensory roots. Reflex 50 Hz T-EMG helps identify normal and abnormal responses, further assisting in grading the abnormal responses from 0-4.

Train of four (TOF)

During TOF monitoring, peripheral nerves are stimulated and assessed for neuromuscular blocking agents using TCeMEP and EMG monitoring. The posterior tibial nerve is stimulated distal to the surgical site at the medial malleolus, and the compound muscle action potential (CMAP) is recorded from the abductor hallucis muscle located at the foot. The stimulation frequency is set to 2 Hz, pulse width to 200 microseconds, and duration to 2 seconds. A TOF of 4/4 indicates a blockage of less than 5% of the body's muscles. Three twitches (TOF=3/4) correspond to 75% blockage, TOF=2/4 to 85% blockage, TOF=1/4 to 95% blockage, and TOF=0/4 to 100% blockage. For intraoperative EMG, t-EMG, or TCeMEP, at least 4 out of 4 twitches must be present (TOF=4/4).

Selective Dorsal Rhizotomy (SDR):

Surgical Procedure:

Selective Dorsal Rhizotomy (SDR) is a neurosurgical procedure used to reduce spasticity in patients with conditions such as cerebral palsy (Figure 1). The patient is positioned face down during the procedure, and a small incision is made in the lower back. The L1 vertebrae are removed to expose the spinal cord, and ultrasound and X-ray are used to identify the specific nerves. Selectively cutting the dorsal rootlets involves several vital steps to ensure maximum reduction of spasticity while minimizing adverse effects. During SDR, the surgeon exposes the lower spinal cord and identifies the dorsal roots of the lumbar and sacral spinal nerves. Each dorsal root is then divided into smaller rootlets, individually stimulated using lowintensity electrical currents. The responses are monitored using EMG to determine the degree of hyperactivity. Responses upon stimulation are identified as contributing to spasticity and are selectively severed. This approach allows for precise targeting, ensuring that only the problematic rootlets are cut while preserving those necessary for normal sensory function. The goal is to balance, reduce spasticity, and maintain adequate sensory input to avoid complications such as numbness or weakness.

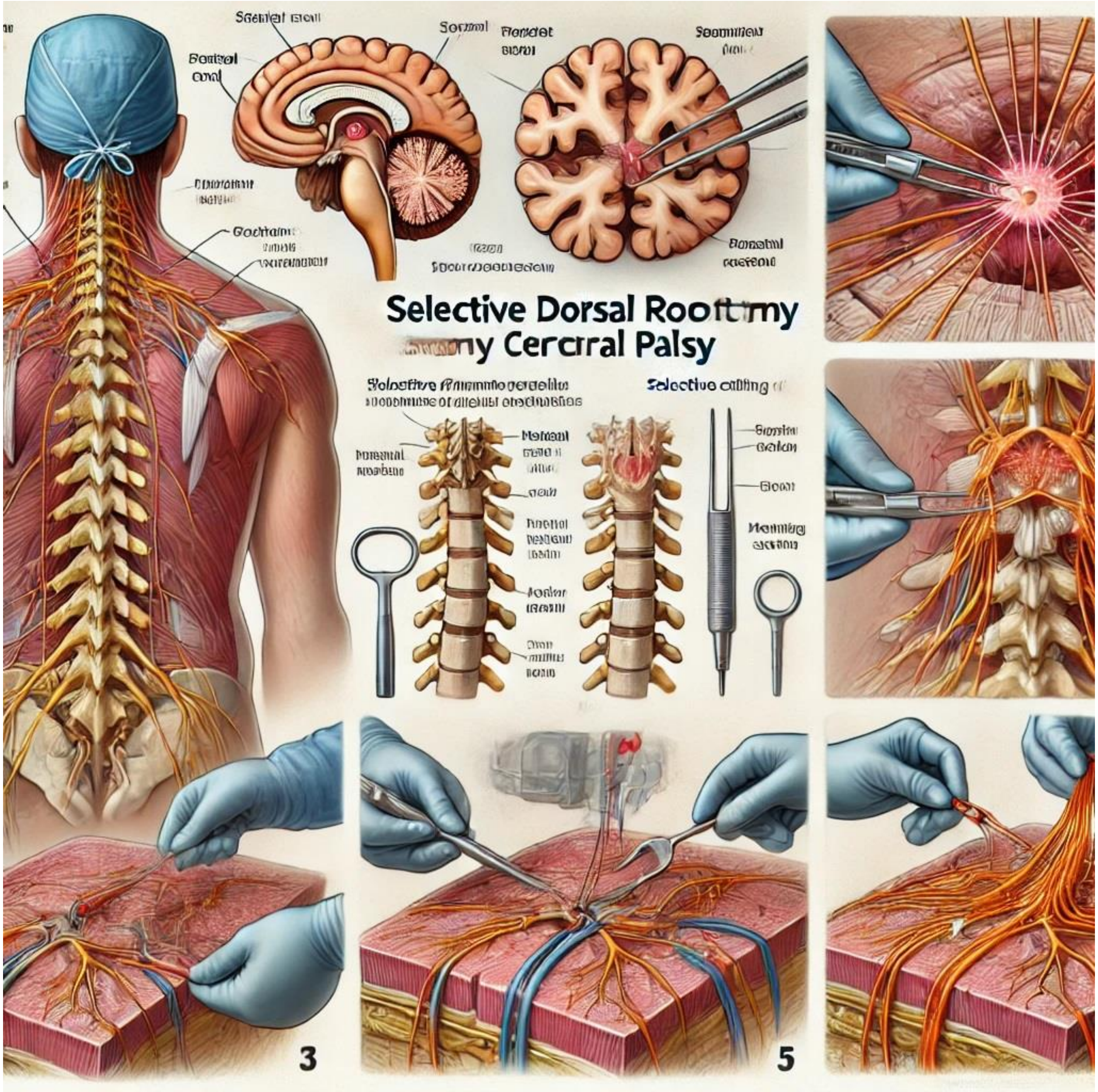

Figure 1. An illustration depicting the stages of selective dorsal root rhizotomy (SDR) for cerebral palsy, including an overview of the spinal cord, exposure during surgery, rootlet identification, selective cutting, and postoperative changes.

The use of IONM, including EMG, SSEPs, TCeMEP, and TOF, helps guide the surgeon in making informed decisions about which rootlets to cut, thereby improving the overall safety and efficacy of the procedure. The nerves are then separated by placing a rubber pad in between the sensory nerve above and the motor nerves beneath, preventing surgical damage. After exposure, each sensory nerve was divided into 3-5 rootlets and tested for spasticity with t-EMG using two fine-tip monopolar hook probes. The responses are graded from 0 (nonspastic) and 1 (mildly spastic) to 4 (severely spastic). Abnormal rootlets are resected, and the procedure was repeated for the L2 and S1 segments.

Multimodality IONM is used to determine the selection of severed nerve roots and avoid accidental involvement of other motor nerve roots. Constant current stimulation and recording EMG responses from the limb muscles help categorize the nerve rootlets to be transected. The rootlets graded 3 or 4 are cut, and the ones graded two are preserved unless indicated otherwise. This ensured that only the roots closely associated with spasticity were transected. Furthermore, it provided a detailed insight into the patient's initial spinal-neurofunctional state, thus providing information on the reorganization process.

Grading Criteria for Rootlets with 50 Hz Stimulation

During SDR, the rootlets are evaluated using electrical stimulation at 50 Hz, and their responses are graded on a scale from 0 to 4 to determine which rootlets should be severed (Figure 2). The grading criteria are as follows:

- Grade 0: No response to stimulation. This indicates that the rootlet is not contributing to spasticity and should be preserved.
- Grade 1: Minimal response with slight muscle contraction. These rootlets are typically preserved unless other factors indicate a need for sectioning (Figure 3).
- Grade 2: Moderate response with visible muscle contraction. These rootlets may be considered for cutting depending on the overall assessment of spasticity.
- Grade 3: Strong response with significant muscle contraction. Rootlets graded at this level often contribute substantially to spasticity and are generally recommended for sectioning.
- Grade 4: Maximal response with exaggerated and sustained muscle contraction (Figure 4). These rootlets contribute to severe spasticity and are prioritized for cutting [3].

This grading system allows for a systematic and objective evaluation of each rootlet, ensuring that only those rootlets contributing significantly to hyperactive reflexes are severed. 50 Hz stimulation helps differentiate between normal and hyperactive reflex pathways, thereby aiding in the selective nature of SDR.

The procedure aimed to divide approximately 60% to 70% of the sensory nerve roots between L2 and S1. Rootlets with grades 3 to 4 are associated with incremental, clonic, multiphasic, and sustained wave responses, and thus, they are preferentially divided and transected.

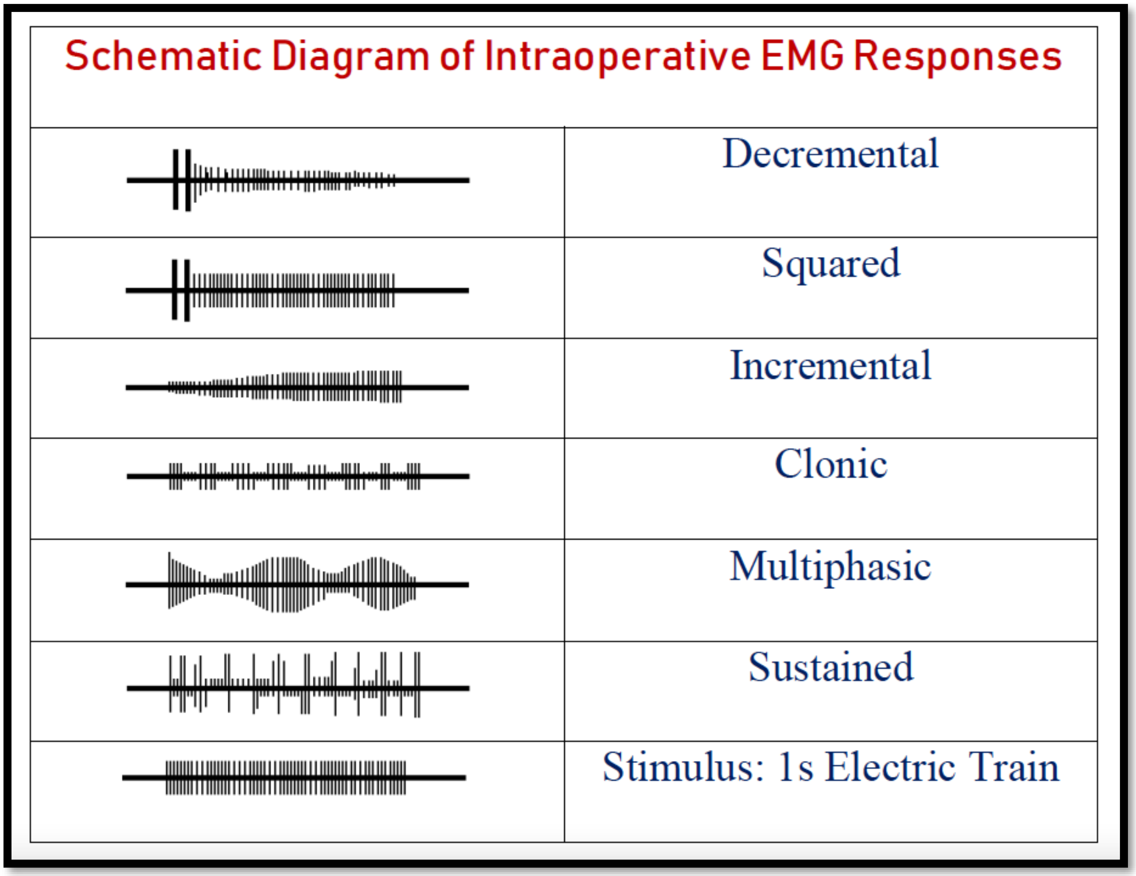

Figure 2. Illustration of the normal and abnormal 50 Hz reflex triggered electromyographic response during dorsal root rhizotomy procedure.

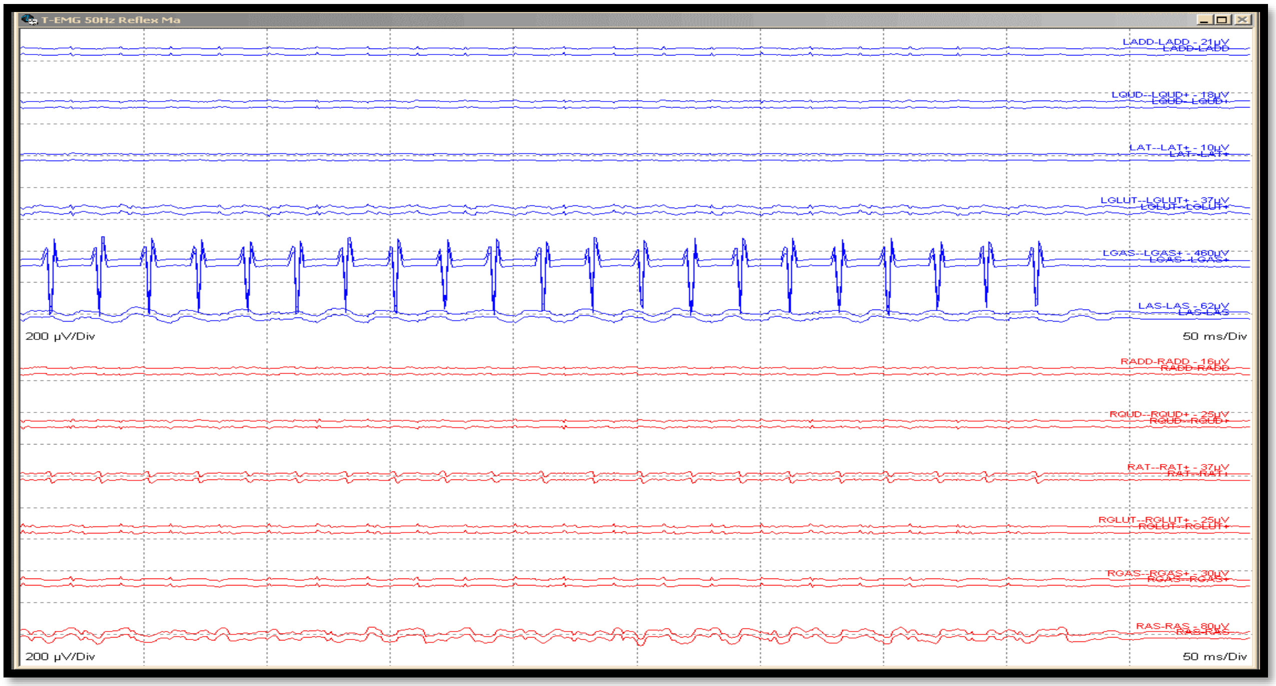

Figure 3. Grade 1 squared 50 Hz reflex triggered electromyographic response from one muscle during dorsal root rhizotomy procedure.

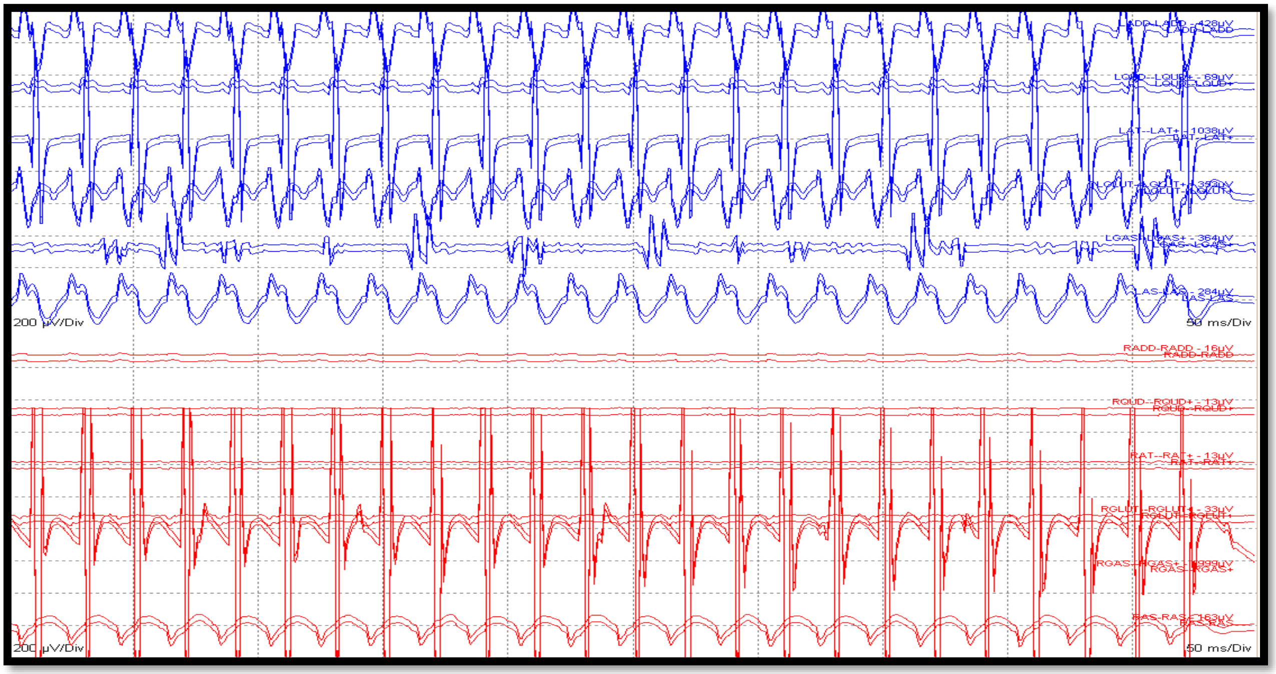

Figure 4. Grade 4 multiphasic 50 Hz reflex triggered electromyographic response from multiple muscles bilaterally during dorsal root rhizotomy procedure.

DISCUSSION

Clinical Efficacy of SDR

Several studies have demonstrated the efficacy of SDR in reducing spasticity and improving motor function in children with CP. A landmark study by Park et al. (2013) reported significant reductions in spasticity, measured by the Ashworth Scale, and improvements in Gross Motor Function Classification System (GMFCS) levels following SDR. Patients exhibited enhanced mobility, reduced need for orthopedic interventions, and improved quality of life. The study concluded that SDR is most beneficial for children between 3 and 7, as their neuroplasticity allows for better motor adaptation post-surgery.

Similarly, a meta-analysis by Grunt et al. (2011) found that SDR led to significant long-term reductions in spasticity and improvements in gross motor function compared to non-surgical management [8]. The meta-analysis emphasized that children with spastic diplegia—those with spasticity predominantly affecting the lower limbs—benefited the most from SDR. These findings are corroborated by observational studies, which indicate that children who undergo SDR experience sustained improvements in gait and functional independence [13].

It was postulated by Turner et al. [14] that the use of IONM aided in selective sectioning of the dorsal nerve roots that cause spasticity by sending excitatory stimuli. Applying electric current stimulation and reading the EMG responses helps to intraoperatively categorize and grade the rootlets of the nerve to be transected. This is a crucial part of the surgical decision-making process. The patient is saved from the loss of crucial sensory functions and does not experience the adverse effects otherwise manifested as post-op complications in SDR. SDR using IONM techniques has shown benefits excelling beyond lumbosacral function. Improvements in upper extremities, speech, as well as refined cognitive functioning are noted, mostly in severely disabled CP children.

Cerebral Palsy is a disease that, if left untreated, can lead to decreased muscle power, joint pain, and spasticity of muscles. This spasticity results in “early aging” of the patient and has not been shown to improve spontaneously. Thus, surgical correction is crucial to improve the mobility of these patients. Selective dorsal rhizotomy (SDR) has been proven to reduce muscle spasticity and enhance ambulation greatly. In a study conducted by Park et al. [15], the functional outcomes of 95 patients were evaluated after 20-28 years of childhood SDR for spastic CP. SDR was shown to have a positive impact on the quality of life of 91% of patients, while 2% felt it negatively impacted their lives. The study further concluded that SDR surgery had no late complications [15]. Another study conducted by Munger et al. evaluated long-term outcomes of SDR 10-17 years after surgery. Results showed that patients of non-SDR groups underwent more subsequent treatments (surgeries and anti-spasticity injections) as compared to those who underwent SDR [16]. Similar results were deduced in a meta-analysis of randomized SDR trials assessing CP patients with spastic diplegia who underwent surgery in addition to physical therapy or physical therapy alone, concluding that the surgical division manifested better results [17].

A study by Nordmark et al. [18] reviewing the 5-year follow-up of 35 children who underwent SDR, and postoperative rehabilitation showed a marked reduction in muscle tone and improved mobility. However, four patients developed increased lordosis, three had spondylolisthesis, and five showed scoliosis. In addition, seven children had an occasional urinary tract infection, problems with micturition, constipation, and sleep problems. Park et al. [15] mentioned that pain was noted in 38% of patients, specifically in the back and lower limbs, with mean pain ranging from 4.2 ± 2.3 according to the Numeric Pain Rating Scale (NPRS). Transient dysesthesias were also seen in 2.5% to 40% of patients. However, this was seldom permanent and usually resolved within a few weeks [18]. On the other hand, Paul Steinbok concluded that participants who underwent SDR had decreased lower limb spasticity and increased upper limb range of motion. A positive impact was noted on functional limitation dimensions and motor functions [19]. The employment of IONM in SDR has drastically reduced the occurrence of these complications. Finally, the success of the SDR procedure depends on the expertise of the surgeon and the neuromonitoring team. A well-versed surgeon and an experienced team are crucial for a successful procedure outcome.

Long-Term Outcomes and Complications

Long-term outcomes of SDR have been generally favorable, with sustained reductions in spasticity and improvements in motor function. A 10-year follow-up study by Langerak et al. (2014) demonstrated that most patients maintained lower spasticity levels, improved gait, and increased participation in daily activities [9]. However, the study also noted that long-term physical therapy is essential to support functional gains, emphasizing the importance of postoperative rehabilitation.

Complications associated with SDR are relatively uncommon but can include sensory deficits, muscle weakness, and bladder dysfunction [5]. The risk of complications is minimized through careful patient selection and modern surgical techniques, including intraoperative neurophysiological monitoring. Postoperative weakness, often transient, can occur due to reduced afferent input to the spinal cord, necessitating intensive rehabilitation to regain strength and coordination [3].

A retrospective cohort study by Steinbok (2001) highlighted the importance of postoperative care, noting that patients who received consistent physical therapy were more likely to achieve positive long-term outcomes [19]. The study also pointed out that complications such as spinal deformities might develop over time, particularly in patients with pre-existing orthopedic issues. Therefore, regular follow-up with orthopedic assessments is recommended for patients who undergo SDR.

Comparison with Other Spasticity Management Strategies

SDR is one of several options available for managing spasticity in CP, with other interventions including botulinum toxin injections, intrathecal baclofen therapy, and orthopedic surgeries. Each approach has distinct advantages and limitations, and treatment choice often depends on the individual patient's needs and the severity of spasticity.

Botulinum toxin injections are commonly used for focal spasticity management and provide temporary relief by inhibiting acetylcholine release at the neuromuscular junction [20]. While effective for targeted muscle groups, the effects are short-lived, typically lasting 3-6 months, necessitating repeated injections. In contrast, SDR permanently reduces spasticity, making it more suitable for patients with generalized lower limb involvement [6].

Intrathecal baclofen therapy (ITB) is another option for managing spasticity, particularly in patients with widespread involvement or dystonia. ITB involves the delivery of baclofen directly into the spinal fluid via an implanted pump, allowing for more effective spasticity control with fewer systemic side effects compared to oral baclofen [21]. However, ITB requires ongoing pump management, including refills and potential complications such as infection or catheter malfunction. SDR is a one-time surgical intervention compared to ITB, requiring a more extended initial recovery period and extensive rehabilitation [13].

Orthopedic surgeries, such as tendon lengthening or osteotomies, are often performed to address musculoskeletal deformities resulting from prolonged spasticity. These procedures are frequently used with SDR to optimize functional outcomes, particularly in older children or adolescents with contractures or skeletal abnormalities [22]. The combination of SDR and orthopedic interventions can provide a comprehensive approach to improving mobility and reducing pain in patients with CP.

Quality of Life and Functional Independence

Several studies have focused on the impact of SDR on quality of life, with most reporting positive outcomes in terms of mobility, independence, and overall well-being. A study by McLaughlin et al. (2002) found that children who underwent SDR experienced significant improvements in their ability to perform activities of daily living, such as dressing and walking independently [23]. Parents also reported reduced caregiver burden, as their children required less assistance with mobility and self-care tasks.

Furthermore, the psychological benefits of improved mobility should not be overlooked. Increased independence often leads to better social integration, higher self-esteem, and enhanced participation in school and recreational activities [9]. These improvements contribute to a better quality of life for the patients and their families, who benefit from the reduced physical and emotional demands of caregiving.

CONCLUSION

Selective dorsal root rhizotomy is a well-established surgical intervention for managing spasticity in children with cerebral palsy, particularly those with spastic diplegia. The procedure has evolved significantly over the past century, with advances in surgical techniques and patient selection contributing to its success. The literature supports the efficacy of SDR in reducing spasticity, improving motor function, and enhancing quality of life, particularly when combined with comprehensive postoperative rehabilitation.

While SDR offers a permanent solution for spasticity, it is not suitable for all patients, and careful selection is essential to maximize benefits and minimize risks. SDR provides a unique advantage in improving motor function compared to other spasticity management options, such as botulinum toxin injections or intrathecal baclofen therapy. However, the need for intensive postoperative care and the potential for long-term complications must be considered when deciding on the most appropriate intervention for each patient.

Multimodality intraoperative neurophysiological monitoring (IONM) during SDR is instrumental in achieving optimal outcomes. By combining EMG, SSEPs, TCeMEPs and other monitoring techniques, multimodality IONM provides comprehensive, real-time feedback that enhances the precision of rootlet sectioning while minimizing the risk of complications. This approach improves the safety and efficacy of SDR and contributes to more consistent and favorable long-term outcomes for patients with CP. The integration of multimodality IONM represents a significant advancement in the surgical management of spasticity, underscoring its importance in modern neurosurgical practice.

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