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Improving Quality of Life Through Selective Dorsal Rhizotomy in a Pediatric Patient with Cerebral Palsy and Spastic Quadriparesis: A Rare Case with Twisted Conus Medullaris

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Abstract

Background: Cerebral palsy (CP) is a permanent movement disorder characterized by abnormal motor control and posture, often leading to significant physical disability. A common feature of CP, spasticity, results from damage to upper motor neurons, causing muscle stiffness and involuntary contractions. Selective dorsal rhizotomy (SDR) is a neurosurgical procedure aimed at reducing spasticity in CP patients.

Case Description: A 6-year-old male with spastic quadriparesis due to infancy spasticity in the right hand and lower limbs underwent SDR after physiotherapy failed to alleviate his severe spasticity. Neurological examination revealed spastic paraparesis in the lower extremities and a history of seizures. MRI confirmed the necessity for surgical intervention due to abnormalities from vertebrae Th12 to L1. The patient underwent SDR under general anesthesia, targeting spinal roots from L3 to S1. The immediate postoperative period was uneventful, with no major complications reported. This suggests that SDR can be safely performed in pediatric patients with CP and severe spasticity.

Conclusion: SDR presents a valuable surgical option for managing severe spasticity in children with CP. The positive outcomes observed in this case highlight its potential to enhance motor function and overall quality of life. However, careful patient selection and multidisciplinary management are crucial for achieving optimal results. Long-term follow-up is essential to assess the durability of spasticity reduction and improvements in motor function, emphasizing the need for further studies on the long-term benefits and risks of SDR.

Keywords: Cerebral Palsy, Spastic Quadriparesis, Selective Dorsal Rhizotomy, Pediatric Neurosurgery, Spasticity Management

INTRODUCTION

The management of pediatric patients with cerebral palsy (CP) and spastic quadriparesis presents unique challenges, particularly when accompanied by anatomical anomalies such as a twisted conus medullaris. Selective dorsal rhizotomy (SDR) has emerged as a surgical intervention aimed at alleviating spasticity and improving functional outcomes in this population. This procedure involves the selective cutting of sensory nerve roots in the spinal cord, which can lead to a significant reduction in muscle tone and spasticity, thereby enhancing the quality of life for affected children. The implications of SDR extend beyond mere physical improvements; they encompass psychological and social dimensions that are crucial for holistic patient care.

Cerebral palsy is characterized by a range of motor impairments due to non-progressive disturbances occurring in the developing fetal or infant brain. Spastic quadriparesis, the most severe form of CP, results in significant motor dysfunction and often

coexists with other complications, including cognitive impairments and musculoskeletal issues. The presence of a twisted conus medullaris adds another layer of complexity, as it may influence the efficacy of conventional treatments and necessitate tailored surgical approaches. The integration of SDR into the treatment regimen for these patients has been shown to yield positive outcomes, including improved mobility, independence in daily activities, and enhanced overall quality of life. [1]–[3]

Cerebral palsy (CP) is a disorder distinguished by irregular tone, posture, and movement. It is clinically categorized according to the prevailing motor syndrome, which includes extrapyramidal or dyskinetic spastic hemiplegia, spastic diplegia, and spastic quadriplegia. CP occurs at a rate of 2–3 per 1,000 live births. [4] A recognized consequence of an upper motor neuron (UMN) lesion, such as cerebral palsy (CP), is spasticity. Spasticity is frequently considered the most prevalent motor impairment in CP. [5] Regarding the contribution of spasticity to hypertonia and, more specifically, to the gait abnormalities observed in CP, there are numerous uncertainties. [6], [7] Various methods have been employed to treat individuals with spastic diplegia, including a procedure known as selective dorsal rhizotomy (SDR). Despite the effectiveness of SDR in treating spastic patients, there is a lack of awareness among neurologists and neurosurgeons regarding the procedure, its indications, and expected outcomes. This is mainly due to the limited number of centers that offer this treatment. [8]

Research indicates that the benefits of SDR are not limited to physical improvements; they also encompass emotional and psychological well-being. Children who undergo SDR often experience a reduction in the stigma associated with physical disabilities, leading to enhanced self-esteem and social integration. Furthermore, the procedure can alleviate the burden on caregivers, who often face significant emotional and physical strain when caring for children with severe disabilities. By improving the child's functional abilities, SDR can foster greater family cohesion and reduce caregiver stress, thereby enhancing the quality of life for both the patient and their family. [9]–[11]

The decision to proceed with SDR must be made on a case-by-case basis, considering the unique anatomical and functional characteristics of each patient. In cases where a twisted conus medullaris is present, careful preoperative assessment using advanced imaging techniques is essential to determine the optimal surgical approach. The potential risks associated with SDR, including postoperative complications and the need for rehabilitation, must also be weighed against the anticipated benefits. Multidisciplinary collaboration among neurosurgeons, physiotherapists, occupational therapists, and psychologists is crucial in developing a comprehensive care plan that addresses the diverse needs of these patients. [12]–[14]

Selective dorsal rhizotomy represents a promising intervention for improving the quality of life in pediatric patients with cerebral palsy and spastic quadriparesis, particularly in the context of complex anatomical challenges such as a twisted conus medullaris. The potential for enhanced mobility, reduced spasticity, and improved psychosocial outcomes underscores the importance of this surgical option in the comprehensive management of these patients. Ongoing research and clinical evaluation will be essential in refining surgical techniques and optimizing patient selection criteria to maximize the benefits of SDR in this vulnerable population.

CASE REPORT

A 6-year-old boy presents with the chief complaint of stiffness in the right hand and both lower limbs. Previously referred with a diagnosis of Cerebral Palsy post VP Shunt and Spastic paraplegia, the patient is scheduled for Selective Dorsal Rhizotomy (SDR) and

a thoracolumbar MRI (Figure 1). Currently, the patient remains conscious and active, yet lacks eye contact and verbal communication, only able to vocalize screams without clear words. The stiffness in the limbs has been persistent since infancy, showing improvement with physiotherapy. The patient maintains good oral intake, without vomiting, and has a history of seizures, with the last episode occurring in December 2023. The seizures, resembling daydreaming, lasted approximately 10 minutes without biting or bed wetting, followed by crying. Past medical history includes VP Shunt placement in 2017 and a shunt revision in 2021. The patient was born via cesarean section, with prenatal ultrasound indicating a buildup of fetal brain fluid. Developmentally, the child can sit independently but is non-verbal. Immunizations are up-to-date, and current medications include Depakene (Valproic acid), Lioresal, Brainact, and Neurotam. No allergies are reported. Neurological examination reveals spastic paraparesis with varying degrees of spasticity and range of motion in different segments, with sensory evaluation posing challenges. Postoperative wound assessment shows no complications.

The motor examination reveals that the patient presents with inferior paraparesis, indicating weakness in the lower extremities, coupled with spasticity, which refers to increased muscle tone and stiffness. The assessment of specific spinal segments shows varying degrees of spasticity and range of motion. In the cervical spine, segments C5, C6, and C7 exhibit spasticity but maintain free range of motion. However, at the C8 and T1 levels, spasticity is present bilaterally, suggesting more pronounced stiffness in these segments. Moving down the spine to the lumbar region, segments L2 demonstrate normal range of motion bilaterally, while segments L3, L4, and L5 display spasticity bilaterally, indicating increased muscle tone and stiffness in these areas. Similarly, at the sacral level (S1), spasticity is observed bilaterally (Figure 2). This detailed motor examination provides valuable insight into the patient's neurological condition, highlighting the extent and distribution of spasticity along the spinal cord.

Diagnostic procedure indicated for selective Dorsal Rhizotomy Spinal Roots of Lower Extremity L3-S1, and later carried out. At present, the lower extremities seem to be unaffected by spasticity, and the legs can be moved through tactile stimulation. Reports of stiffness and seizures are refuted.

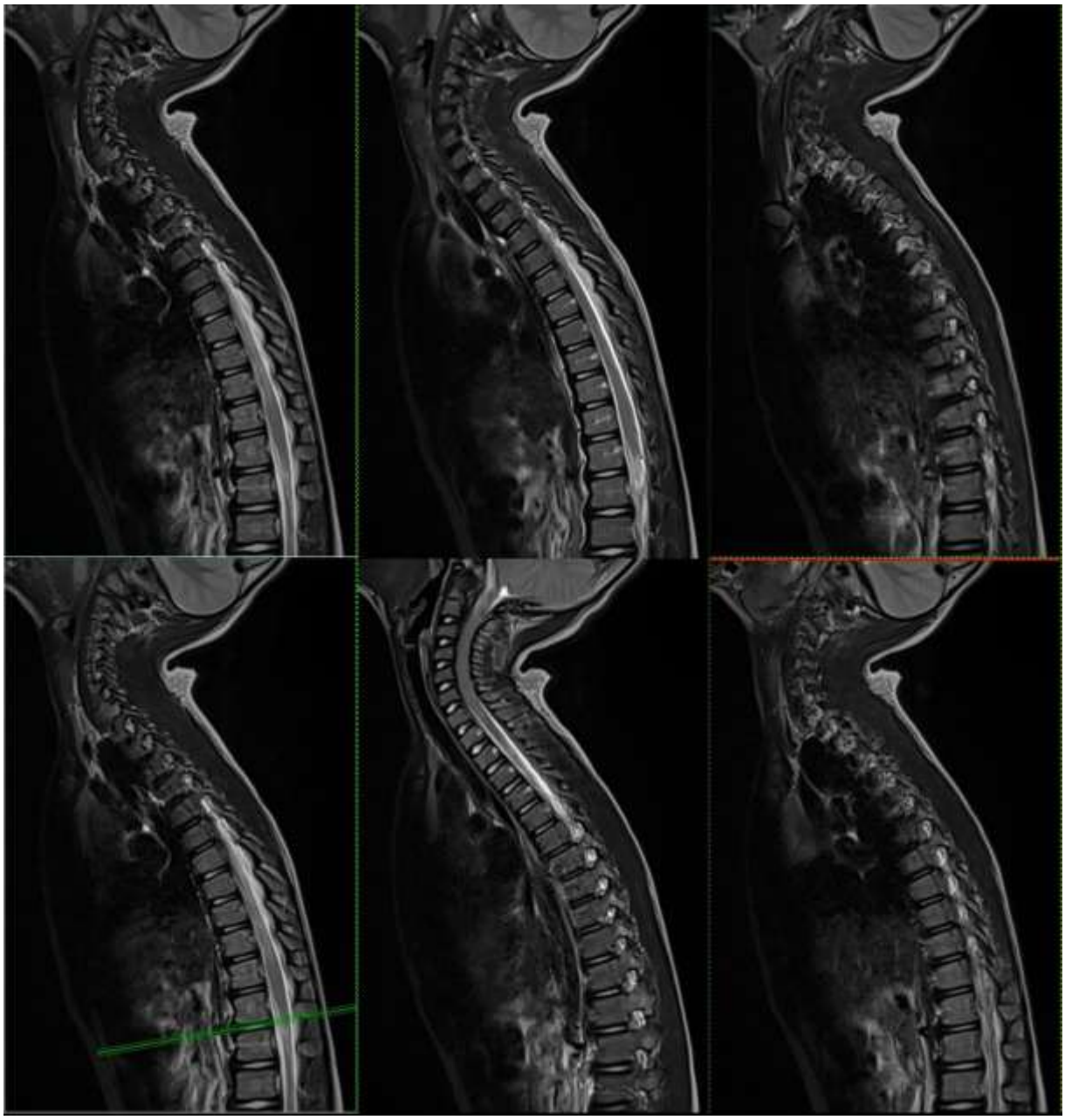

Figure 1. Spine MRI of Thoracal and Lumbal

Figure 2. Durante SDR Operation

DISCUSSION

Selective dorsal rhizotomy (SDR) is a specialized surgical procedure designed to alleviate spasticity, particularly in children diagnosed with cerebral palsy (CP) exhibiting spastic quadriparesis. [15], [16] This intervention involves the careful division of specific sensory nerve roots, followed by a rigorous and structured rehabilitation program. The significance of SDR lies in its ability to enhance motor function and improve the overall quality of life for individuals facing severe spasticity due to CP. This case report aims to elucidate the importance of SDR through a detailed examination of a patient with unique anatomical challenges and end-stage spasticity. [17]

The patient in this case report was diagnosed with cerebral palsy following the placement of a ventriculoperitoneal (VP) shunt, presenting with spastic paraplegia. The clinical presentation included pronounced stiffness in the right hand and both lower limbs, significantly impairing mobility and daily functioning. In managing such complex cases, a thorough selection process is crucial. This includes comprehensive assessment protocols that evaluate the patient's neurological status, spasticity levels, and overall functional capabilities. The surgical team must ensure that the decision to proceed with SDR is grounded in robust clinical evidence, as well as a clear understanding of the patient's unique condition. [18]–[20]

One of the key components of SDR is the identification and division of the appropriate dorsal rootlets responsible for spasticity. Research has consistently shown that SDR can reduce spasticity in a predictable and significant manner, leading to improved motor function. The delineation of spasticity

across specific spinal segments provides invaluable clinical insights, allowing clinicians to tailor treatment strategies effectively. By pinpointing areas of heightened spasticity, targeted pharmacological interventions or neurorehabilitation techniques can be employed to address specific neurological deficits, ultimately enhancing motor function and quality of life. [21]

During the surgical procedure, an unexpected anatomical configuration was revealed: the conus medullaris exhibited twisted radicles, with the right radicles positioned posteriorly and the left radicles anteriorly. This anatomical anomaly was not detected during preoperative magnetic resonance imaging (MRI) examinations, adding a layer of complexity to the surgical intervention. Such variations in anatomy can significantly impact surgical planning and execution, underscoring the necessity for detailed anatomical assessments prior to SDR.

Despite the common practice of utilizing intraoperative monitoring (IOM) to enhance surgical precision and safety, this particular procedure was conducted without IOM. The decision was made based on the patient's established end-stage spasticity scores and the understanding that the primary goal of the surgery was to improve the patient’s quality of life rather than restore ambulation. This approach reflects a growing recognition that, in certain cases, the benefits of SDR can extend beyond mere mobility improvements, addressing the broader spectrum of functional capabilities and well-being.

The patient's spasticity was classified as end-stage, indicating that traditional interventions had been exhausted. In such cases, the severity of spasticity necessitates a surgical approach to provide any significant relief. The decision to proceed with SDR, despite the challenges posed by the unique anatomical configuration and the absence of IOM, underscores the potential of this procedure to deliver meaningful outcomes even in complex scenarios.

Comparing this case to other reports in the literature, it becomes evident that SDR holds promise for improving the quality of life in patients with severe and multifaceted spasticity profiles. This case highlights the critical importance of thorough anatomical and functional assessments in tailoring surgical interventions. It also demonstrates the feasibility of performing SDR without IOM in select cases, particularly when the primary focus is on enhancing the patient's overall quality of life rather than solely on mobility restoration.

The Gross Motor Function Measurement scale (GMFM) serves as a pivotal tool in assessing the extent of spasticity reduction, improvements in strength, range of motion, fine motor skills, and daily activities following SDR. [22], [23] Additionally, it allows for the evaluation of hip spasticity, the need for subsequent orthopedic procedures, bladder and sphincter function, and the potential complications that may arise postoperatively. While there is a possibility of complications associated with SDR, the procedure is generally considered safe and effective, providing long-lasting and substantial functional improvements for well-selected children with spasticity related to cerebral palsy. [24], [25]

CONCLUSION

In conclusion, this case report underscores the transformative potential of selective dorsal rhizotomy in managing spasticity in children with cerebral palsy. By carefully selecting patients and employing a comprehensive approach to assessment and management, clinicians can navigate the complexities of surgical intervention, even in the presence of unique anatomical challenges. The evidence supporting SDR efficacy in reducing spasticity and improving functional outcomes is compelling, reinforcing its role as a critical option in the management of severe

spasticity. As the field continues to evolve, ongoing research and clinical experience will further elucidate the best practices for optimizing outcomes in this vulnerable population, ultimately enhancing the quality of life for children with cerebral palsy.

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Institutional Review Board approval is not required.

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The authors certify that they have obtained all appropriate patient consent.

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