Clinical Case Reports

ISSN: 2675-3626

Selective Ventral and Dorsal Rhizotomy for Refractory Status Dystonicus Cerebral Palsy: A Case Report

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Introduction/Background: Status dystonicus (SD) is a life-threatening neurological emergency, particularly in children with mixed-type cerebral palsy (CP), where dystonia and spasticity coexist. Although deep brain stimulation (DBS) is a recognized therapeutic option, its use in acute settings and resource-limited environments can be challenging.

Case Presentation: We report the case of a 4-year-old boy with mixed CP secondary to traumatic brain injury, who developed refractory SD. The patient presented with severe dystonia, spasticity (Ashworth grade 3) and required intensive care due to rhabdomyolysis and autonomic instability. After failure of clinical management, a mixed selective rhizotomy (MVR) was performed as an emergency intervention.

Conclusions: MVR resulted in immediate motor relief, full resolution of SD, and functional improvement of the upper limbs. This case highlights MVR as an effective, low-cost and viable alternative for controlling refractory SD in patients with mixed motor phenotypes, especially in resource-limited settings.

Keywords: Status dystonicus, cerebral palsy, selective rhizotomy

INTRODUCTION

Dystonia is a movement disorder characterized by sustained or intermittent muscle contractions that result in repetitive movements, abnormal postures or both. When exacerbated acutely and persistently, it may progress to a medical emergency known as status dystonicus (SD)—a severe, potentially fatal condition that requires prompt and specialized intervention.(1,4) SD represents the worst form on the dystonia spectrum, marked by acute neurological deterioration, autonomic instability, multi-organ failure and intense pain. It is more prevalent in pediatric population, especially among patients with acquired dystonia, as in the case of developmental encephalopathies and dyskinetic cerebral palsy, but can also occur in genetic forms of dystonia syndromes as DYT-TOR1A, GNAO1 and inborn errors of metabolism(1,4).

The causes of SD are diverse and may be classified as genetic or acquired. Among genetic ones, mutations in DYT-TOR1A, GNAO1, KMT2B and ARX are frequently associated with early-onset, generalized dystonia with an aggressive course.(1,2) Acquired causes include dyskinetic or mixed (spastic-dystonic) cerebral palsy, perinatal brain injury, inherited metabolic disorders (e.g., glutaric aciduria type I, Lesch–Nyhan syndrome), neurodegenerative diseases and central nervous system infections or inflammatory conditions(1,4).

Triggers include infection, fever, pain, dehydration, abrupt drug withdrawal and constipation. In some cases, SD may be the first manifestation of a previously unrecognized neurological condition. Clinical presentation includes progressive dystonia, sympathetic hyperactivity, metabolic disturbances and potentially multiorgan failure. Diagnosis is clinical and severity can be graded with the Dystonia Severity Scale (DSS)(1,6).

In children with mixed-type CP, spasticity and dystonia frequently coexist. Spasticity exacerbates pain, spasms and dystonic movements. Selective dorsal rhizotomy (SDR) is used to treat spasticity by targeting afferent sensory inputs. Whereas selective ventral rhizotomy (SVR) addresses efferent motor outflow.(2,5,7,9) The combined ventral and dorsal rhizotomy (CVDR) can interrupt both mechanisms, offering dual therapeutic action. SDR typically targets lumbar roots (L2-S1), focusing on lower limbs, often with beneficial upper-limb effects. Intraoperative neurophysiology guides root selection(8,9).

Submitted: 30 June 2025
Accepted: 31 August 2025
Published: 13 September 2025

SVR is less commonly performed, but in severe dystonia, it may be essential to complement SDR. SDR alone could worsen dystonia by removing sensory inhibition.(2,7) Recent literature has explored ablative strategies, such as pallidotomy, for refractory dystonia or intraventricular baclofen pumps (3,4,10). However, spinal interventions offer faster relief in critical settings.

CVDR may be particularly indicated in patients with dyskinetic spastic phenotype, lack of response to medication, no indication or availability for neuromodulation techniques, life-threatening SD and predominant lower limb and axial involvement(4,10).

CASE REPORT

A 4-year-old male child with a diagnosis of dyskinetic spastic cerebral palsy secondary to a severe traumatic brain injury (TBI) 1,5 years before, presented with a Glasgow Outcome Scale (GOS) score of 3. His motor phenotype was dominated by dystonic features, including sustained abnormal posturing such as archer's posture, internal rotation and flexion of the right upper limb, hypotonic, functionless left upper limb and lower limbs hyperextension. He had shown partial clinical response to botulinum toxin injection. No fixed skeletal deformities observed (Figure 1) .

Figure 1 - Patient presenting archer's posture in the context of dyskinetic spastic CP

The coexisting spastic component, typical of cerebral palsy, manifested with classic signs of upper motor neuron involvement: hyperreflexia, resistance to passive movement and velocity-dependent hypertonia Ashworth 3.

The patient acutely deteriorated with SD without an identifiable trigger. He developed rhabdomyolysis, markedly elevated creatine phosphokinase (CPK) levels, 689 U/L, and autonomic instability, requiring admission to the pediatric intensive care unit (PICU) for stabilization. He was reclassified as DSS grade 4 upon admission and showed partial improvement with stabilization of CPK levels between 180–200 U/L, despite intensive medical management, remaining on refractory status pathway.

Due to the refractory nature of the dystonic crisis, a decision was made to proceed with MVR, aiming to interrupt the reflex sensorimotor loops that perpetuate the dystonic state. This approach was persued because its rapid execution, low cost, availability of functional pediatric neurosurgery team, intraoperative neurophysiology and the potential for immediate symptom relief (Figure 2). Anticipated postoperative paraparesis was not considered to increase morbidity in this particular case, as the patient was already bedridden and fully dependent on caregivers for basic activities of daily living.

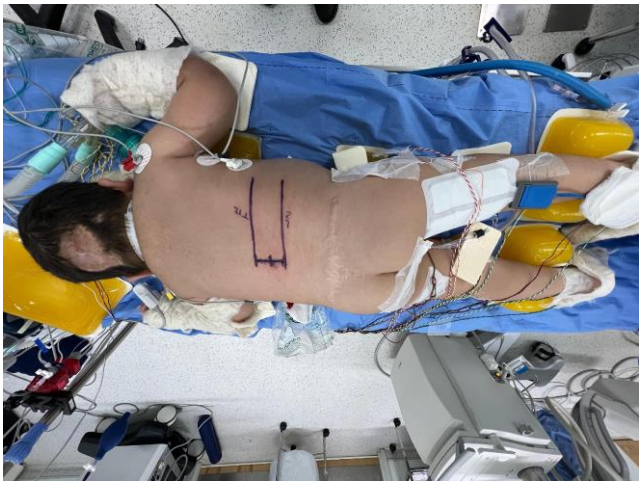

Figure 2 - Patient's positioning for the surgery

Postoperatively, the patient experienced complete resolution of the status dystonicus 5 days later and return to baseline neurological function, with marked improvement in the lower limbs, which became flaccid and CPK 92 U/L. The upper limbs remained grade 3 on the Ashworth scale, but with clinical signs of functional improvement and reduced cervical dystonia. A transient complication involved a worsening of a previously diagnosed neurogenic bladder, which had been managed with intermittent catheterization for one month due to increased urinary retention, likely exacerbated by pre-existing bladder wall thickening. The condition resolved spontaneously and the patient returned

to baseline urinary function, continuing to use diapers. The hospital discharge were around 1,5 month due to systemic infections management (not surgery related) and homecare issues.

Pharmacologically, the patient was maintained on seven medications (clonidine, baclofen, biperiden, gabapentin, carbamazepine, diazepam and clonazepam), the same number as preoperatively; however, dose reductions were achieved across multiple agents. On the DSS, the patient improved from grade 3 at hospital admission to grade 2 at discharge, with significant clinical improvement, no recurrence at 3 months follow-up and high family satisfaction degree.

DISCUSSION

This case illustrates a safe, effective and accessible surgical option for the management of refractory status dystonicus in a pediatric patient with dyskinetic cerebral palsy. While deep brain stimulation (DBS) is the most studied neurosurgical intervention for refractory dystonia, it is associated with high cost, technological requirements and delayed clinical effects—factors that limit its use in acute dystonic crises. In contrast, the combined ventral and dorsal rhizotomy performed here demonstrated immediate motor relief without the need for electrodes, electronic permanent implanted hardware or postoperative programming. (2,3)

The literature on rhizotomy in dystonia is limited. Cury et al. documented 5 children treated with ablative procedures (including rhizotomy) among 28 with SD; 93% returned to baseline.(2) Marras et al.(3) and Garg et al(10). described pallidotomy as an emergency option for refractory SD. Ruiz-Lopez and Fasano emphasized the need for conceptual reframing and broadened surgical strategies.(4) Ahluwalia et al. also described the use of combined dorsal and ventral rhizotomy for mixed hypertonia in nonambulatory patients, though not specifically in the context of SD.(7)

Most existing surgical reports focus on ablative intracranial procedures or globus pallidus internus DBS, which are feasible preferably when the patient is clinically stable and long-term follow-up is possible. As discussed by Cury et al., spinal ablative techniques, such as rhizotomy, have been successfully employed in a limited number of patients with SD, particularly where rapid and definitive resolution is critical. (2)

This case is particularly relevant because it involves a mixed motor phenotype, in which dystonia and spasticity coexist and interact. In such scenarios, treating only one motor component—as is typical with intrathecal/ventricular baclofen for both conditions or DBS for dystonia—may be insufficient, due to hardware availability and maintenance. Combined rhizotomy directly targets both afferent (dorsal) and efferent (ventral) spinal roots, disrupting the pathophysiological circuits responsible for sustained abnormal muscle activity in both spasticity and dystonia.

To date, no pediatric case report was found in the literature describing the use of CVDR as a primary surgical strategy for acute refractory SD. This emphasizes the originality and relevance of this case. This case highlights the importance of targeting both afferent and efferent circuits in dyskinetic spastic ones. It may be life-saving when neuromodulation is not feasible.

The successful outcome in this patient suggests that combined rhizotomy is a viable, cost-effective and rapid intervention for severe secondary dystonia, particularly in settings where neuromodulation is contraindicated or unavailable. The technique can be performed in resource-limited environments, assuming the availability of basic neurosurgical infrastructure and intraoperative neurophysiological monitoring.

Finally, the limited body of published evidence regarding rhizotomy in dystonia—especially in mixed motor phenotypes with status dystonicus—emphasizes the novelty and clinical importance of this report, without previous published cases found in literature research. Further studies, including case series, are needed to better define indications, outcomes and safety profiles of this approach, taking into account that baclofen pumps are the classic reported therapy. Detailed clinical reports such as this one may serve as valuable guidance in urgent and complex clinical decision-making.

CONCLUSION

Combined ventral and dorsal rhizotomy seems to be an effective and safe alternative functional neurosurgical strategy for controlling refractory status dystonicus in patients with severe dyskinetic spastic cerebral palsy. In emergency clinical settings with limited time and resources, rhizotomy may offer a viable neuromodulatory solution providing immediate symptom relief at low cost with direct impact on clinical stabilization.

Moreover, the combined approach is particularly appropriate in patients with coexisting dystonia and spasticity, where simultaneous modulation of afferent and efferent spinal pathways may be more effective than targeting either component in isolation.

In the context of limited formal evidence on the use of rhizotomy for status dystonicus, especially in pediatric patients, this case contributes clinically relevant data and underscores the need for further investigation into mixed ablative strategies for complex, refractory movement disorders.

ACKNOWLEDGMENTS

We thank all the patients and their families who participated in this study.

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the local Ethics Committee, number: 153670588

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper

Funding

This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors

Artificial intelligence

The authors affirm that no artificial intelligence tools were used in the writing, editing, or content generation of this manuscript. All work was conducted manually, based on thorough research and academic expertise.

CONTRIBUTIONS

-**Antônio Jorge Barbosa de Oliveira**: Methodology, Writing – review & editing
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-Kesia Priscilla Omena Cardoso: Methodology, Writing – review & editing
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-Rayane Gomes de Sousa: Methodology, Writing – original draft, Writing – review & editingMethodology, Writing – review & editing

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