Clinical Case Reports

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Intraventricular baclofen and bilateral pallidotomy in a pediatric patient with acquired generalized dystonia and refractory status dystonicus: illustrative case

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Background: Status dystonicus (SD) is a life-threatening emergency in pediatric patients with severe dystonia, often requiring intensive care. Surgical interventions, such as deep brain stimulation, intrathecal baclofen (ITB), or pallidotomy, are considered for refractory cases, but evidence for intraventricular baclofen (IVB) and bilateral pallidotomy (BP) in children is limited. We report a novel combined approach using IVB followed by BP in a 3-year-old boy with refractory SD secondary to hypoxic-ischemic encephalopathy.

Case Description: A 3-year-old male developed SD and spastic quadriparesis following cardiorespiratory arrest due to airway obstruction. Despite aggressive medical management (levodopa, clonazepam, tetrabenazine, oral baclofen, sedatives), SD persisted, requiring prolonged mechanical ventilation. An ITB test (100 mcg) reduced the modified Ashworth scale from 5 to 3 and dystonic episodes from 8 to 2/day. IVB was placed via a third ventricle catheter with a pump delivering 100-1050 mcg/day, achieving spasticity control but fluctuating dystonia response. Due to recurrent SD, poor prognosis, and malnutrition, BP was performed using stereotactic microelectrode guidance and radiofrequency lesions (80°C, 60s). Postoperative MRI confirmed precise pallidal lesions. The patient achieved full SD resolution, was weaned from ventilation, and stabilized at home with tracheostomy and gastrostomy.

Conclusions: bilateral pallidotomy and IVB should be considered in the treatment of refractory SD, particularly for patients with contraindications for DBS and concurrent severe spasticity. The literature review support this findings, providing an overview that can guide palliative management in similar cases. Further studies are needed to establish comprehensive guidelines for surgical management of SD in children.

Keywords: Intraventricular baclofen, bilateral pallidotomy, dystonia, status dystonicus

INTRODUCTION

Status dystonicus (SD) represents a significant source of disability with a high burden of disease.1 Dystonia severity can range from mild to severe, with SD being the most extreme form, representing a life-threatening emergency associated with considerable morbidity.2 Children with SD are at risk of respiratory dysfunction, metabolic imbalances, pain, and fractures, among other complications, requiring critical care in the pediatric intensive care unit for airway management and sedative administration, with mortality rates reaching up to 12.5%.3,4 Surgical treatments are a valuable therapeutic option when a refractory SD is established and among surgical alternatives deep brain stimulation (DBS) has demonstrated efficacy.5 Another valid alternative is bilateral pallidotomy (BP) in patients with cognitive impairment, young age, cachectic state, inability to comply with follow-up visits, failed DBS or patients with contraindications for DBS.6 Intrathecal baclofen (ITB) is an accepted surgical option for pediatric population with dystonia showing spasticity and dystonia concurrently.7 ITB baclofen has been tried in a small number of patients with refractory status dystonicus with various reports of benefit and intraventricular baclofen (IVB) is described as an alternative, when ITB is not feasible or are contraindications to this route of administration.8 Despite previous publications, there remains a need for comprehensive guidelines for treating SD in children. We present the case of a 3-year-old boy with refractory SD secondary to hypoxic-ischemic encephalopathy and the outcomes of a novel surgical approach involving IVB followed by bilateral pallidotomy.

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CASE REPORT

A previously healthy 3-year-old male patient presented with severe hypoxic-ischemic encephalopathy secondary to a cardiorespiratory arrest, which occurred as a result of upper airway obstruction caused by a Melicoccus bijugatus seed. This event precipitated the development of SD accompanied by severe spastic quadriparesis. The patient initially received intensive medical management, including administration of levodopa, clonazepam, tetrabenazine, oral baclofen, and high doses of sedative medications. Despite these efforts, the patient's condition remained refractory to treatment, necessitating prolonged mechanical ventilation in a pediatric intensive care unit setting (Figure 1).

Figure 1 – Status dystonicus in Pediatric ICU

An intrathecal baclofen test was conducted using 100 mcg of baclofen. The test yielded a positive result, demonstrating a significant improvement in clinical status. Specifically, the modified Ashworth scale score was reduced from 5 to 3 across all four limbs, and the frequency of dystonic episodes decreased from 8 to 2 per day. Additionally, there was a 50% reduction in the dose of sedative drugs required to manage the patient's condition. Despite this positive response, the patient's dystonic status continued to present challenges, and further intervention was deemed necessary. Due to the the positive result in the intrathecal baclofen test, the decision was made to proceed with intraventricular baclofen (IVB) therapy. A catheter was inserted into the third ventricle using precise neuronavigation techniques, and the catheter was secured at the cranial exit (Figure 2). The catheter was tunneled subcutaneously to the right lower quadrant of the abdominal subfascial tissue to house the pump (Figure 3). Postoperatively, the baclofen infusion pump was programmed to deliver a constant rate of 100 mcg/day. Throughout the patient's hospitalization, the baclofen dosage was adjusted daily based on the clinical response, gradually increasing to 850 mcg/day over the course of the first two weeks. This adjustment led to a substantial improvement in spasticity, along with resolution of SD. However, the response in dystonia remained fluctuating, indicating the need for further evaluation. Despite the improvement in spasticity, the patient's SD recurred, prompting a thorough review of the IVB system's function. Contrast infusion testing was conducted to assess the adequacy of the pump's patency, which confirmed appropriate function. Given the poor neurological prognosis, along with the patient's malnutrition state, a bilateral pallidotomy (BP) was considered as the next step in treatment. Preoperatively, the patient's Burke Fahn Marsden rating scale score was recorded at 96 points, indicating the severity of the dystonic condition.

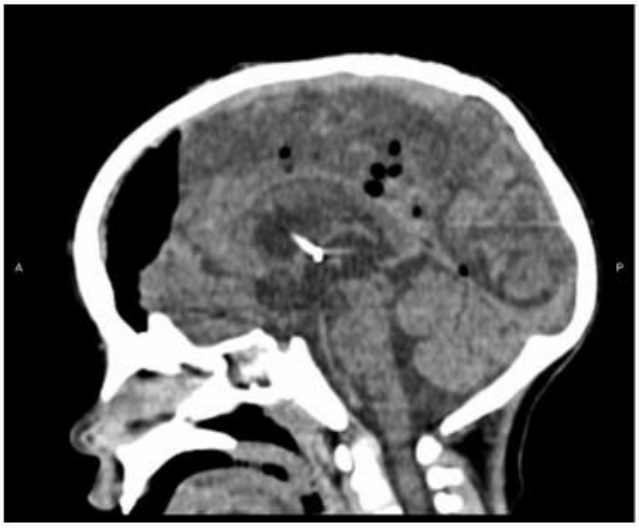

Figure 2 – Immediate postoperative CT scan showing thrid ventricle intraventricular catheter of Baclofen pump

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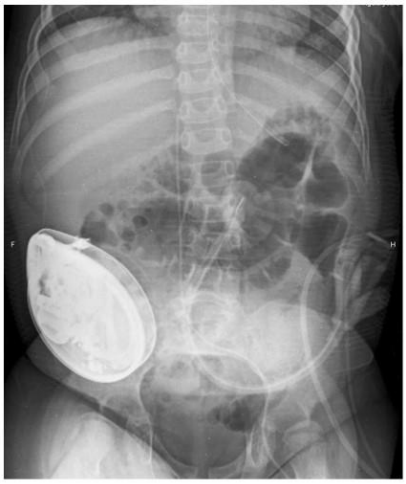

Figure 3 – Baclofen pump in abdominal region

The bilateral pallidotomy procedure was performed using a stereotactic guide with microelectrode guidance. Both sides of the pallidum were targeted in a single surgical intervention. Radiofrequency lesions were applied with a temperature of 80°C for 60 seconds. Magnetic resonance imaging (MRI) was conducted postoperatively to confirm the precise location of the lesions (Figure 4). The procedure was completed without any intraoperative complications. Following the pallidotomy, the patient's initial response was partial, with gradual but significant improvement. Over time, the patient achieved full resolution of SD and was successfully weaned from mechanical ventilation. The baclofen infusion rate was further adjusted to 1050 mcg/day, optimizing spasticity control. The patient's clinical condition continued to improve, and he was gradually stabilized (Figure 5). He has been at home for just over a year, utilizing a tracheostomy and gastrostomy as part of his ongoing rehabilitation management.

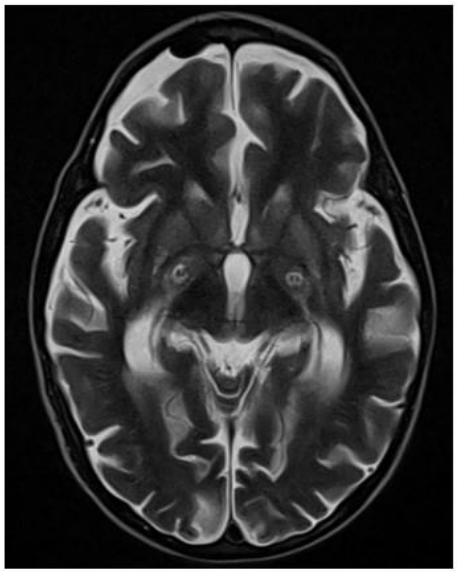

Figure 4 – Postoperative MRI showing BP in a T2 weighted sequence

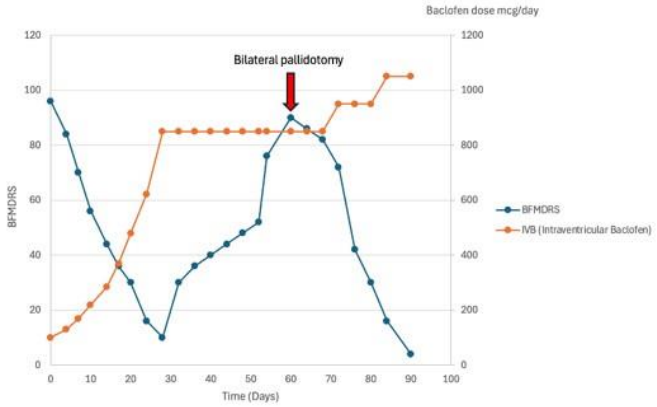

Figure 5 – Course of IVB dose and BFMDRS scores following the surgical placement of the IVB pump

DISCUSSION

The use of intraventricular baclofen (IVB) and pallidotomy in pediatric patients with refractory dystonia represents a novel therapeutic approach, as illustrated by this case. Baclofen, a GABA-B receptor agonist, is traditionally administered intrathecally to manage spasticity and dystonia in conditions such as cerebral palsy (CP) and secondary dystonia(9). However, intraventricular administration offers a targeted approach to deliver baclofen directly to the cerebrospinal fluid (CSF) surrounding the brain, potentially enhancing its efficacy in patients with severe, generalized dystonia unresponsive to intrathecal baclofen (ITB)(10). In this case, IVB was initiated to address the patient's profound dystonic symptoms, which aligns with prior reports demonstrating improved dystonia control with IVB in children with CP and traumatic brain injury (11-12).

The decision to proceed with pallidotomy following suboptimal IVB response reflects the challenges in managing refractory dystonia. Pallidotomy, a stereotactic lesioning procedure targeting the globus pallidus interna (GPi), has been largely supplanted by deep brain stimulation (DBS) in pediatric dystonia due to the latter's reversibility and adjustability(13). However, pallidotomy remains a viable option in resource-limited settings or when DBS is contraindicated, as in cases with high infection risk or hardware intolerance (14). Studies have reported significant dystonia reduction post-pallidotomy in children with primary and secondary dystonia, with improvements in Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) scores ranging from 30% to 60% at 6–12 months (15-16). In our patient, pallidotomy resulted in a notable reduction in dystonic posturing, consistent with these findings, suggesting that targeted lesioning can still play a role in carefully selected cases.

The combination of IVB and pallidotomy in this case is particularly noteworthy, as there is scant literature on their sequential or combined use in children. The synergistic effect observed here—IVB providing partial symptom control followed by pallidotomy achieving further improvement—may be attributed to their complementary mechanisms. IVB likely modulates cortical and subcortical excitability via GABAergic pathways(17), while pallidotomy disrupts aberrant basal ganglia circuitry driving dystonia(18). This case underscores the potential for multimodal strategies in refractory dystonia, echoing reports of combined pharmacological and surgical interventions yielding superior outcomes compared to monotherapies(19). Despite these encouraging results, several limitations must be acknowledged. First, as a single case report, the generalizability of our findings is limited. Second, the long-term outcomes of IVB and pallidotomy remain uncertain, as prior studies suggest potential waning efficacy or complications such as lesion-related cognitive deficits or pump infections(20-21). Third, the absence of standardized protocols for IVB dosing and pallidotomy lesion planning in children complicates reproducibility(22). Future research should focus on prospective studies to establish optimal dosing regimens, lesion targets, and patient selection criteria. Additionally, comparative studies evaluating IVB versus ITB and pallidotomy versus DBS could further clarify their roles in pediatric dystonia management.

CONCLUSION

In conclusion, bilateral pallidotomy and IVB should be considered in the treatment of refractory SD, particularly for patients with contraindications for DBS and concurrent severe spasticity. The literature review support this findings, providing an overview that can guide palliative management in similar cases. Further studies are needed to establish comprehensive guidelines for surgical management of SD in children.

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. According to institutional and national guidelines, ethics approval was not necessary for this study.

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.

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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

-**Juan Camilo Arias-Angulo**: Conceptualization, Writing – original draft
-**Sebastian Ordoñez-Curé**: Conceptualization, Writing – original draft
-**Oscar Escobar-Vidarte**: Resources, Supervision, Writing – review & editing
-Lucely Ortega-Bolaños: Conceptualization, Methodology

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