Clinical Case Reports

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Intraventricular Baclofen via neuronavegated Ommaya reservoir as Bridge Therapy in Refractory Status Dystonicus: A Case Report

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Introduction: Status dystonicus (SD) is a life-threatening neurologic emergency that requires prompt recognition and intervention. Intraventricular baclofen (IVB) has been shown to be an alternative therapy with promising results in refractory cases.

Case Presentation: We present a 12-year-old male with Wilson's disease and acquired generalized dystonia who developed status dystonicus after a gastrostomy procedure. Due to refractory, his dystonic symptoms despite of optimized intensive medical management, including intravenous sedation and invasive ventilatory assistance, we implemented intraventricular baclofen therapy via a temporary Ommaya reservoir. This allowed for rapid stabilization and resolution of the SD, prior to attempting to a more definitive surgical solution.

Conclusion: Temporary Ommaya-based IVB is a feasible and cost-effective bridge therapy for SD management. It provides rapid symptomatic control and may enable strategic planning of definitive interventions, when applicable

Keywords: intraventricular, baclofen, ommaya catheter, status dystonicus

INTRODUCTION

Status dystonicus (SD), is a neurological emergency marked by severe, often continuous exacerbations of generalized or focal hyperkinetic movements requiring urgent hospitalization due to life-threatening complications, irrespective of the patient's baseline neurological status (14). The transition from stable dystonia to SD is not clearly delineated and should be considered in the presence of bulbar dysfunction, respiratory compromise, metabolic derangements, intractable pain or exhaustion (10).

SD may arise from various etiologies, including genetic and acquired dystonic syndromes, such as cerebral palsy and Wilson's disease. It may be prompted by metabolic stress or infectious/inflammatory conditions, drug toxicity, medication withdrawal and structural brain insult (11). SD may occur as a single episode or be recurrent and difficult to control. The management strategies of SD remain complex and continually evolving, nevertheless still strongly relying on a multidisciplinary intensive supportive care. Optimal medical management, psychological patient-centered and parental support, physical and respiratory rehabilitation, and in selected cases timely invasive interventions are crucial for improving outcome (7,15).

Among pharmacologic strategies, baclofen—a selective GABA_B receptor agonist— has been available for several decades as one of the main agents in the medical management of spasticity, but it also plays a supportive role as an off-label treatment in dystonia, particularly in complex cases, when other treatments are insufficient. Its mechanism involves both presynaptic inhibition of excitatory neurotransmitter release via calcium influx suppression and postsynaptic hyperpolarization via potassium efflux enhancement (1). Intrathecal baclofen (ITB) delivery provides direct access to the cerebrospinal fluid (CSF), allowing for lower therapeutic concentrations with substantially fewer systemic side-effects in comparison with oral therapy. Initially employed for spasticity management in cerebral palsy, ITB demonstrated notable benefits in dystonic patients with refractory symptoms (16).

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Subsequently, Albright and colleagues reported improved speech, functional status and quality of life in this group treated with ITB (19). Although its use in SD remains underrepresented in the literature, several case reports have documented favorable outcomes (5,8), but with much higher doses compared with predominantly spastic patients.

Intraventricular baclofen (IVB) administration has recently gained attention as an alternative route, potentially offering equal or superior efficacy with even significantly lower daily doses. Comparative studies suggest that IVB is at least as safe as ITB, with some evidence indicating higher cortical drug concentrations following ventricular administration (2,12). In the context of SD, early IVB administration has been shown promising results in case reports, including in patients refractory to prior invasive therapies (13,9). However, these reports typically describe delivery through permanent implanted pumps, which may delay urgent treatment due to the need for expensive dedicated devices and time needed for progressive dose titration.

We propose the temporary use of an Ommaya reservoir—cheaper and readily available solution most neurosurgical centers—as an alternative intraventricular access for baclofen administration in the acute management of dystonic status. This approach may serve as a bridge therapy to achieve rapid symptomatic control, reduce dependence on continuous anesthetic and sedative agents and reduce critical time until definitive treatments, such as permanent intraventricular pump, deep brain stimulation (DBS) or ablative procedures like pallidotomy, can be safely pursued.

CASE REPORT

A 12-year-old male with previous diagnosis of Wilson's disease and irregular follow-up consultations was admitted to our institution for optimization of medical and nutritional management.

Upon admission, the patient presented with a Dystonia Severity Scale (DSS) score of 3. Due to significant nutritional deficiencies identified during the initial evaluation, a gastrostomy procedure was indicated. Shortly thereafter, however, the patient developed status dystonicus, necessitating endotracheal intubation and the initiation of multiple sedative agents to control the severe and refractory neurological symptoms.

During the hospitalization, the patient required up to seven sedative agents (midazolam, fentanyl, clonidine, baclofen, biperiden, clonazepam, diazepam) with progressive dose escalation. His DSS worsened to 5. The clinical course was further complicated by ventilator-associated pneumonia, urinary tract infections, fungal sepsis, and elevated creatine phosphokinase (CPK) levels exceeding 1200 mg/dL.

In view of the persistent and refractory generalized dystonia, surgical intervention became necessary. Although a bilateral pallidotomy was initially planned, delays in material availability and emerging supportive evidence for intraventricular baclofen led to a change in strategy. Given the presence of an available Ommaya reservoir and literature supporting its use for baclofen delivery, an intraventricular baclofen infusion was proposed as a bridge therapy to reduce sedation burden and stabilize the patient for posterior definitive surgical treatment.

The procedure was performed 15 days after the onset of refractory dystonia. Under general anesthesia, the patient was positioned supine with the head elevated at 45 degrees and kept in a neutral position (Figure 1). A curvilinear incision was made at the right Kocher's point (1 cm anterior to the coronal suture along the midpupillary line). After standard antiseptic preparation, the subgaleal layers were dissected, a burr hole was created, and the dura was opened (Figure 2). Cortical coagulation was performed to facilitate catheter placement.

Figure 1 – Patient's positioning and trichotomized area

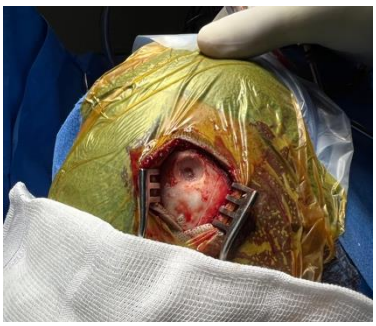

Figure 2 – Burr hole and dura-mater after subperiosteal layer dissection

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Neuronavigation was employed to guide precise insertion of the catheter into the third ventricle, ensuring optimal cerebrospinal fluid (CSF) distribution of the drug (Figure 3). Once accurate positioning was confirmed, the catheter was connected to an Ommaya reservoir, which was secured in a subperiosteal pocket to reduce the risk of manipulation-related complications (Figure 4 A). The procedure was completed uneventfully. Postoperative computed tomography (CT) showed satisfactory catheter placement and no evidence of surgical complications (Figure 4B)

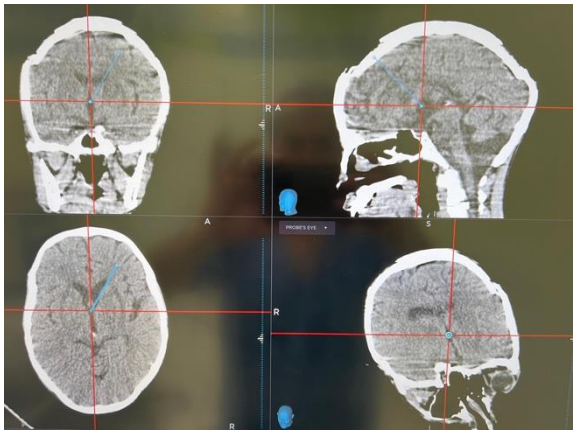

Figure 3 – Neuronavigation planning trajectory and target in the third ventricle

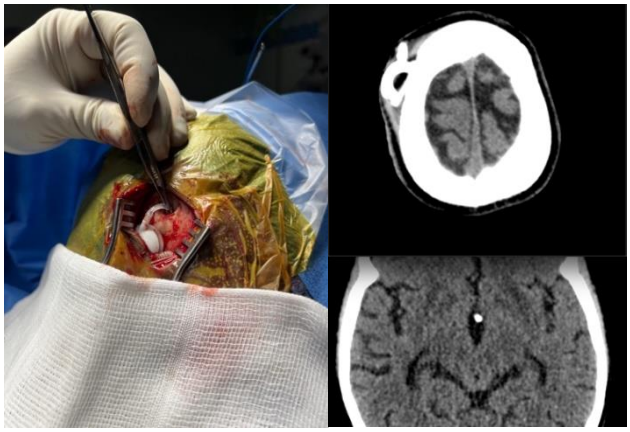

Figure 4 – A - Ommaya's reservoir connected to proximal catheter and subperiosteal insertion . B - Post op Computerized Tomography with Ommaya's Reservoir position.

Postoperatively, a protocol of daily intraventricular baclofen administration was initiated via percutaneous puncture of the Ommaya reservoir using a sterile 13 × 0.45 mm needle. The dose was gradually increased to a maximum of 560 mcg/day with reevaluation every 48 hours. This regimen facilitated a progressive reduction in both the daily doses and number of sedative agents needed to control his dystonic symptoms, slowly enabling transition to oral medications where feasible. Nevertheless, likely due to the long ICU stay and severity of his condition, the patient remained depended of mechanical ventilation.

On postoperative day 26, a cerebrospinal fluid fistula at the surgical site was detected, which was successfully managed without further complications. On day 33, following acquisition of the necessary materials, the patient underwent bilateral posteroventral pallidotomy. The Ommaya reservoir was removed during the same surgical session, and sedative medications were gradually withdrawn. The patient achieved resolution of status dystonicus and was subsequently transferred to a tertiary care facility in his home state for continued multidisciplinary rehabilitation.

This case report was approved by the Institutional Research Ethics Committee and written informed consent was obtained from the patient's legal guardians for publication.

DISCUSSION

SD represents a neurological emergency with significant morbidity and mortality, particularly in the pediatric population. Despite increasing awareness and advances in supportive care, the pharmacologic and surgical management of refractory SD remains complex. Well-documented complications—including respiratory failure, rhabdomyolysis, dysphagia, anarthria, thromboembolic events, gastrointestinal bleeding, traumatic injuries, fractures, and sepsis - contribute to poor functional outcomes (20,21).

Intrathecal baclofen (ITB) remains a cornerstone of neuromodulatory treatment (1,6,19). However, its implementation may be hindered by anatomical constraints, infection risks or limited access to costly implants and medical expertise—issues especially relevant in pediatric and resource-limited settings (5,8).

In this context, temporary intraventricular baclofen (IVB) via an Ommaya reservoir might be a surgical alternative in severe refractory SD cases. Evidence suggests that IVB may achieve higher cortical cerebrospinal fluid (CSF) concentrations than ITB, offering potentially greater efficacy—particularly in hyperkinetic movement disorders with significant cortical and subcortical involvement (12, 3).

The present case illustrates this approach, showing rapid titration of baclofen directly into the ventricular system, without the logistical and financial burden associated with implantable intraventricular pumps, which can exceed USD 30,000 in Brazil. In contrast, Ommaya reservoirs are widely available in neurosurgical centers at a cost of approximately USD 500, representing a viable option in urgent and resource-constrained scenarios (13).

From a technical standpoint, neuronavigation-guided catheter placement into the third ventricle ensured precise drug delivery and minimized perioperative complications. Subperiosteal placement of the reservoir further reduced risks associated with skin tension and external manipulation. Nonetheless, a delayed CSF fistula was observed, which was likely related to the prolonged hospitalization, systemic infection, low body weight, young age and repeated reservoir taps for drug administration. Early recognition and prompt management were essential to prevent serious sequelae such as meningitis or ventriculitis, which can be catastrophic in this population (9,18). In future cases, the use of permanent catheter attached to the Ommaya's reservoir to infuse the medication might be an alternative to prevent this complication.

Clinically, Ommaya-based IVB therapy led to a marked reduction in sedative requirements and stabilization of dystonic symptoms, facilitating the eventual transition to definitive treatment via bilateral pallidotomy. Avoiding prolonged deep sedation is particularly advantageous, as it reduces the risk of complications such as respiratory depression, thromboembolic events, bedsores, withdrawal syndromes and ventilator-associated morbidity as laryngotracheomalacia (22).

In this context, IVB via a temporary Ommaya reservoir served effectively as a bridge therapy, enabling stabilization in a critically ill child and obviating the need for emergency neurosurgical intervention under suboptimal conditions (8,14). To our knowledge, this is the first reported pediatric case employing bedside IVB administration via Ommaya reservoir in the management of SD. Prior literature has focused predominantly on permanent pump systems, which although effective, may be impractical in resource-limited settings (8,2,3,15).

Future directions should include systematic evaluation of pharmacokinetics, optimal dose titration and safe withdrawal for IVB via temporary access systems in SD. Collaborative, multicentric efforts are essential to define the role of this approach within the broader treatment algorithm for SD and to assess its impact in the context of acute and subacute intensive pediatric care (10,15).

CONCLUSION

This case report underscores the efficacy and safety of intraventricular baclofen administration via an Ommaya reservoir as a bridging strategy in the acute management of refractory dystonic storm. The intervention enabled a marked reduction in the requirement for continuous sedation, contributing to clinical stabilization and allowing for the strategic planning of definitive therapies, such as deep brain stimulation or pallidotomy.

These findings support the potential utility of Ommaya-based intraventricular baclofen delivery as a time-sensitive, accessible alternative in emergent settings where rapid symptom control is essential. Further studies are warranted to define standardized indications, optimize dosing and safety protocols, and validate its role within the broader framework of emergency neurological management.

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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 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

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CONTRIBUTIONS

-**Antônio Jorge Barbosa de Oliveira**: Conceptualization, Data curation, Investigation, Methodology, Project administration, Resources, Software, Writing – original draft, Writing – review & editing
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-Rayane Gomes de Sousa: Methodology, Validation, Writing – review & editingMethodology, Writing – review & editing

REFERENCES

1. Albright AL. Baclofen in the treatment of cerebral palsy. J Child Neurol. 1996;11(2):77–83.
2. Albright AL, Ferson SS. Intraventricular baclofen for dystonia: techniques and outcomes. J Neurosurg Pediatr. 2009;3(1):11–4.
3. Albright AL, Barry MJ, Shafton DH, Ferson SS. Intraventricular baclofen for secondary dystonia: a phase 1 clinical trial. Pediatr Neurosurg. 2009;45(1):41–6.
4. Dalvi A, Yerramilli S, Shanske S, Khandji AG, De Vivo DC. Intrathecal baclofen in the management of primary and secondary dystonia in children. J Pediatr. 1998;133(2):252–6.
5. Dalvi A, Fahn SM, Ford B. Intrathecal baclofen in the treatment of dystonic storm. Mov Disord. 1998;13(3):611–2.
6. Ochs G, Götz M, Kappos L, et al. Intrathecal baclofen for long-term treatment of spasticity: a multi-centre study. J Neurol Neurosurg Psychiatry. 1989;52(8):933–9.
7. Grosso S, Pippa L, Gagliardi M, et al. Management of status dystonicus in children: case report and review. Eur J Paediatr Neurol. 2012;16(4):390–5.
8. Kyriagis M, Grattan-Smith P, Scheinberg A, et al. Status dystonicus and Hallervorden-Spatz disease: treatment with intrathecal baclofen and pallidotomy. J Paediatr Child Health. 2004;40(6):322–5.
9. Mingbunjerdsuk D, Mari Z. Intraventricular baclofen following deep brain stimulation in a child with refractory status dystonicus. Mov Disord Clin Pract. 2021;8(3):456.
10. Manji H, Howard RS, Miller DH, et al. Status dystonicus: the syndrome and its management. Brain. 1998;121(2):243–52.
11. Phukan J, Albanese A, Bhatia KP. Primary dystonia and dystonia-plus syndromes: clinical characteristics, diagnosis, and pathogenesis. Lancet Neurol. 2011;10(12):1074–85.
12. Rocque BG, Albright AL. Intraventricular vs intrathecal baclofen for secondary dystonia: a comparison of complications. Oper Neurosurg (Hagerstown). 2012;70(2):ons321–6.
13. Ruggiero C, De Masi S, Albanese A. Early intraventricular baclofen therapy (IVB) for children with dystonic and dysautonomic storm. Childs Nerv Syst. 2019;35(1):15–8.
14. Ruiz-López M, Fasano A. Rethinking status dystonicus. Mov Disord. 2017;32(12):1667–76.
15. Vogt LM, Bhatia KP, Albanese A. Recommendations for the management of initial and refractory pediatric status dystonicus. Mov Disord. 2024;39(9):1435–45.
16. Albright AL. Intrathecal baclofen in cerebral palsy movement disorders. J Child Neurol. 2001;16(1 Suppl):S34–9.
17. Albright AL, Barry MJ, Shafton DH, Ferson SS. Intraventricular baclofen for secondary dystonia: a phase 1 clinical trial. Pediatr Neurosurg. 2009;45(1):41–6.
18. Kim SJ, Ruge D, Carlson JD, et al. Complications of intraventricular catheter placement for baclofen delivery in dystonia. Neurosurgery. 2013;72(3):199–204.
19. 19. Albright AL. Intrathecal baclofen in cerebral palsy movement disorders. J Child Neurol. 2001;16(1 Suppl):S34–9.
20. Vaamonde J, Narbona J, Weiser R, et al. Dystonic storms: a practical management problem. Clin Neuropharmacol. 1994;17(5):344–7.
21. Marsden CD, Marion MH, Quinn N. The treatment of severe dystonia in children and adults. J Neurol Neurosurg Psychiatry. 1984;47(11):1166–73.
22. Amigoni A, Circelli A, Griggio L, et al. Sedation in pediatric intensive care: current practice and emerging evidence. J Clin Med. 2022;11(9):2467.