Hypertonia caused by dystonia and spasticity can be debilitating in pediatric patients with cerebral palsy, spinal cord injury, or traumatic brain injury. First-line treatment includes physical and occupational therapies, muscle relaxants, oral baclofen, phenol injections, and/or botulinum toxin injections. Patients with persistent hypertonia despite these therapies have refractory hypertonia, and many of these patients are candidates for intrathecal baclofen (ITB). ITB is an effective neuromodulatory therapy for reducing muscle tone and improving function in pediatric patients with hypertonia. Candidates who either have undergone a successful ITB bolus trial or have been evaluated and referred by pediatric movement disorder (PMD) specialists are considered for the implantation of a subcutaneous pump and intrathecal catheter system.

Traditionally, the ITB catheter tip is located dorsally in the lumbar or thoracic spine, leading to documented

**ABBRIVIATIONS**

BADS = Barry-Albright Dystonia Scale; ITB = intrathecal baclofen; ITBP = ITB pump; NG = nasogastric; PMD = pediatric movement disorder; POD = postoperative day.

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improvements in lower-extremity hypertonia and a more modest effect in the upper extremities.\(^5\)\(^-\)\(^8\) This is likely due in part to the correlation of segmental effects of ITB on the spinal cord, where the highest concentration of medicine is located at the catheter tip; this effect on tone is less at spinal cord segments farther away, as the concentration of ITB decreases.\(^9\) The cervical spine may have roughly 25\% of the total introduced baclofen dose, with the concentration affected by both diffusivity and specific gravity.\(^9\) The cervical tip location has traditionally been avoided because of concerns of ITB-mediated hypoven- tilation and pneumonia; however, significant increases in these complications in cervical compared with thoracic or lumbar placement have not been reliably proven.\(^5\)\(^,\)\(^10\)\(^,\)\(^11\) Fortunately, one study in adults demonstrated success in treating upper-extremity hypertonia with a rostrally placed catheter without complications relating to tip positioning.\(^6\) One small case series demonstrated that pediatric patients with cervical tip location for ITB delivery due to prior thoracolumbar fusion mass secondary to scoliosis did not experience complications.\(^12\) Concerns persist regarding the effect of high concentrations of cervical ITB on bulbar symptoms leading to breathing and swallowing dysfunc- tion;\(^20\) outcomes and complications in a larger pediatric cohort have yet to be reported.

In this retrospective study, we analyzed the outcomes and complications of cervical ITB tip location in the largest modern cohort of pediatric patients with hypertonia.

Methods

Patient Selection

Institutional review board approval was obtained prior to initiation of this study. A retrospective database of all patients who underwent ITB pump (ITBP) insertion or revision between April 2022 and October 2023 was reviewed; all surgeries with a cervical catheter tip were filtered. Twenty-five patients met the inclusion criteria. Inpatient admissions, outpatient follow-up, and neuroimaging were reviewed.

Surgical Procedure

After appropriate antibiosis, patients were placed in the left or right lateral decubitus position under general anesthe- sia. Every step of the Cerebral Palsy Research Network (CPRN) surgical implant protocol for pump implantation was followed for each case. The attending surgeon decontaminated the skin using a 4-step preparation including 70\% alcohol, Betadine scrub, Betadine prep, and 2\% chlorhexidine, and draped the patient. Gloves were changed and an 8-cm incision was made where the pump would be implanted, as well as a 3-cm incision overlying the intended site for access to the CSF space. Most commonly, CSF space access begins with a percutaneous placement of a 14-gauge Tuohy needle under fluoroscopic guidance. The catheter is navigated using fluoroscopy to the intended spinal location. All patients with mixed hypertonia and/or dystonia received a cervical catheter, and patients with quadriplegic spasticity generally received a cervical catheter unless the family objected. All diplegic spastic patients received a thoracic catheter around T10 targeting the lumbar expansion. Once the catheter was at the target, an incision was made to the fascia. The catheter stylet and needle were removed and the silastic anchor was secured with 2-0 Ticron sutures (Medtronic). The 8-cm incision was opened and a subcutaneous or subfascial pocket was created depending on the patient’s body habitus and intended pump size. The remaining spinal segment was tunneled to the abdominal pocket and cut to an appropriate length, which was connected to the pump segment. The pump segment was then connected to a SynchroMed II pump (Medtronic) filled with 500 \(\mu g/mL\) baclofen. The side port was injected with 10 mg vancomycin, and each wound was irrigated with 1 L dilute Betadine and 1 L vancomycin irrigation (1 g/L). Vancomycin powder (1 g) was placed within the wounds, and they were then closed in a typical layered fashion. These patients were admitted and managed by our PMD subspecialty physiatrists to ensure correct baclofen dosing prior to discharge.

Outcome Measures and Data Analysis

Postoperative surgical site complications including wound dehiscence and infections, as well as effects from baclofen toxicity including respiratory depression, pneumonia, and swallowing dysfunction, were recorded. Wound dehiscence was defined as wound separation necessitating revision surgery. Surgical site infection was defined by positive cultures requiring antibiotics. Pneumonia was defined by the clinical and radiological diagnosis of pneumonia, and readmission for pneumonia was also recorded. Respiratory depression was defined as a decrease in respirations necessitating intervention within 48 hours postoperatively. Swallowing dysfunction was defined as a change of swallowing function leading to diet modification.

Results

Twenty-five patients (8 female) were included. Patient demographics are described in Table 1. The mean age at surgery was 12.44 ± 4.25 years. Sixteen patients (64\%) underwent initial placement of ITBP following a bolus test dose, and 7 patients (28\%) underwent replacement ITBP with catheter revision to the cervical level. Two patients (8\%) underwent initial ITBP without a test dose. Overall, 56\% of patients had a history of prematurity. Fifteen patients (60\%) had a gastrostomy tube, and 3 patients (12\%) had a tracheostomy tube.

Operative characteristics are presented in Table 2. Most catheter tips were located at C6 and C7 (44\% and 24\%, respectively). There was 1 C1–2 cervical catheter puncture (4\%) due to a spinal fusion; the remaining 24 patients (96\%) underwent traditional midlumbar access. Twenty pumps (80\%) were placed subcutaneously, with the remaining 5 (20\%) placed in the subfascial plane. Fifteen pumps (60\%) had a volume of 20 ml, while 10 pumps (40\%) held 40 ml. The mean operative duration was 90.33 ± 49.92 minutes, and the mean length of stay was 5.29 ± 4.74 days. The mean follow-up was 70.46 days (range 12–280 days). The mean aggregated Barry-Albright Dystonia Scale (BADS) score decreased by 9.5 points (\(p = 0.01\)) postoperatively.\(^21\) The mean aggregated modified Ashworth scale score in the upper extremities decreased by 2.14 points (\(p = 0.04\)),
and that in the lower extremities decreased by 4.98 points (p < 0.01) postoperatively.22

Complications are shown in Fig. 1. The overall complication rate was 12%. Two patients (8%) had respiratory depression, 1 case of which (4%) was attributed to baclofen toxicity. This patient also had swallowing dysfunction and temporarily required a nasogastric (NG) tube when her spinal catheter was changed from midthoracic to cervical; the NG tube was removed when the 2400-µg/day dose was decreased to 1900 µg/day. One patient (4%) acquired a methicillin-susceptible Staphylococcus aureus surgical site infection following primary implant, necessitating pump explantation. There was no incidence of pneumonia or wound dehiscence. Increased seizure activity was observed in 1 patient immediately postoperatively; it remains unclear whether this was attributable to the change in catheter location.

Illustrative Case

A 16-year-old female with severe generalized idiopathic dystonia who previously underwent bilateral globus pallidus internus deep brain stimulation and midthoracic ITBP placement presented with continued episodes of status dystonicus. She had a preoperative BADS score of 30 with examination findings including axial hypotonia, neck extension, windswept spine, and rigid extremities. Given these findings, she underwent a lumbar-sacral ventral-dorsal rhizotomy with a catheter revision to C2 for optimized baclofen dosing with increased proximity to the brain. Her original ITBP dosing was 2400 µg/day. On postoperative day (POD) 2 she began experiencing vertical nystagmus, dysphagia, lethargy, and respiratory depression requiring increasing oxygen by nasal canula. Head and cervical spine CT scans were noncontributory. Given the concern for baclofen toxicity, her baclofen dose was decreased by 7.5% to 2220 µg/day. Her nystagmus improved in a few hours; however, she required NG tube placement and a high-flow (20 L/min) nasal canula. Her baclofen was decreased every 24 hours by 4.9% to 7.5% for 2 days to arrive at 1955.6 µg/day with improvement of oxygen requirements. She was able to be weaned to room air on POD 11 and was discharged to inpatient rehabilitation on POD 14. Her swallowing improved enough at inpatient rehabilitation for NG tube removal on POD 20, with a BADS score reduction to 17.

![FIG. 1. Complication rates. The overall complication rate was 12%. Two patients (8%) had respiratory depression, 1 case of which (4%) was attributed to baclofen toxicity. One patient (4%) acquired a surgical site infection following primary implant, necessitating pump explantation.](image.png)
Discussion

Our study is the largest modern series to describe the outcomes and complications of continuous dosing cervical ITB therapy in pediatric patients with hypertonia. The overall complication rate is low; respiratory depression was the most frequent complication, occurring in 8% (2/25) of patients, requiring ITB dose weaning in 1 case of baclofen toxicity.

Enteral baclofen is commonly used as an antispasmodic drug for hypertonia in children because of its centrally acting gamma-aminobutyric acid (GABA) B receptor agonism. First-pass metabolism limits the effect of enterally dosed centrally acting medications like baclofen, requiring higher doses that cause side effects. The first documented use of ITB in a child was reported by Dralle and colleagues in 1985. Intrathecal administration delivers baclofen directly to its site of action at very small doses, which results in fewer systemic side effects. ITB has become a standard therapy for severe hypertonia in children, and in some cases it allows for reduction of polypharmacy.

The highest concentration of medication occurs at the catheter tip, with a drop in concentration as the drug diffuses across the spinal cord rostrally. Only approximately 25% of the total infused dose reaches the cervical spine from below, leading to appreciable declines in drug efficacy. As a result, it was postulated that placing the catheter tip in a more rostral position could offer better distribution of medicine throughout the spinal cord.

Lumbar cisternal access is the most common way to access the CSF space, largely because it is safe. Classic teaching is to place the ITB spinal catheter tip into the thoracic or lumbar spine to provide adequate coverage for patients with lower-limb hypertonia. Higher placement in the cervical spine is technically challenging from a lumbar entry point.

McCall and MacDonald compared 23 patients (22 adult, 1 adolescent) who received ITB with a cervical catheter tip to a group receiving a thoracic catheter tip and found similar functional outcomes and complication rates between both groups. A systematic review evaluated 13 studies of cervical ITB catheter tip location and found that 88% of patients experienced functional improvement in the upper extremity and 50% had improvements in both upper and lower extremities. Those authors concluded that drug-related (1%) and technical (21%) complications did not occur more frequently in cervical tips than in thoracic or lumbar tips. The effect of cervical baclofen on respiratory function and sleep apnea was not studied, and the only etiology studied was spasticity.

We highlighted 1 patient within our cohort who experienced respiratory depression due to baclofen toxicity at 2400 µg/day. Primary implants were started at doses of 100–450 µg/day. Patients who required catheter revisions remained at their therapeutic doses of 150–1200 µg/day without experiencing respiratory depression, suggesting that an upper limit of baclofen dosing may exist within the range of 1200–2400 µg/day. Similarly, McCall and MacDonald described 1 patient who experienced hypoventilation and developed aspiration pneumonia.

These two similarly sized studies suggest a nonzero baclofen dose–induced bulbar dysfunction complication rate. It can be inferred that baclofen toxicity may be dose dependent within the cervical spine, although it may also be time dependent. A 2400-µg/day dose titrated over many months may not cause the same baclofen toxicity as a relocated catheter tip at this dose. Although high doses can be tolerated at the lumbar level, patients with comparatively naive cervical spinal cords may need time to build up tolerance. There is limited knowledge regarding the steady-state dose achieved in the CSF or the concentration gradient throughout the CSF. Currently, there is no reliable method to predict the therapeutic dose of individual patients. This will need to be prospectively studied.

Limitations

This is a single-institution, single-surgeon retrospective case series that is at risk of selection bias. Our sample size was relatively small, and we included a heterogeneous dataset of pediatric patients with multiple forms of PMD. Our results may not be generalizable. Future studies with longer follow-up are necessary to determine safety and efficacy parameters.

Conclusions

Cervical ITB is effective in significantly reducing the symptoms of dystonia and spasticity in the upper and lower extremities. Pediatric patients with quadriplegic hypertonia, including from cerebral palsy, can safely receive a cervical catheter tip for ITB dosing, although a percentage may require dose adjustments to avoid bulbar symptoms. Additional studies evaluating cervical ITB may help us determine a time- or dose-dependent effect of ITB on the cervical spinal cord.

References


Disclosures
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Conception and design: Raskin, Abdelmageed, Votoupal.
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