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Case Report

Prepontine placement of an intrathecal baclofen pump catheter for treatment of dystonia

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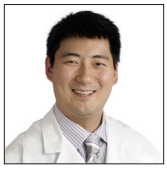

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ABSTRACT

Background: Cerebral palsy with medically refractory spasticity and dystonia is a condition that often benefits from intrathecal baclofen pump therapy to treat these symptoms. In this case report, an intracranial baclofen catheter was placed in the prepontine space to improve withdrawal symptoms in a patient unable to undergo new lumbar catheter placement due to infection.

Case Description: A 22-year-old female with past medical history of cerebral palsy presented with baclofen pump failure and was unable to undergo placement of a new lumbar baclofen catheter due to an infection in her lower back precluding safe and efficacious catheter placement. It was decided the patient would benefit from intrathecal baclofen administered in the prepontine space as a means to avoid a lumbar catheter and thus bypass this prior infection site. An endoscopic third ventriculostomy (ETV) was performed with the endoscope and the distal end of the baclofen pump catheter was fed through this ETV into the prepontine space. Placement in the prepontine space was confirmed by a follow-up head computed tomography. There was a significant improvement in autonomic symptoms and spasticity. By postoperative day 5, the patient was surgically and medically cleared for discharge.

Conclusion: In cases of severe baclofen withdrawal due to dysfunctional pumps, immediate reversal is preferred but may not be feasible due to factors such as infection. This case report has demonstrated that prepontine catheter placement can be effective for the administration of baclofen to reverse withdrawal symptoms in these types of patients.

Keywords: Baclofen pump, Catheter, Prepontine

BACKGROUND

Cerebral palsy is a non-progressive lesion in the developing brain that is often associated with increased muscle tone.[7] Baclofen can be used to treat this dystonia by reducing pain and muscle spasms through its agonistic effect on gamma-aminobutyric acid (GABA)-B receptors.[1,5] Baclofen can be administered orally or intrathecally through a catheter, often placed in the lumbar spine.

With prolonged use or in particularly severe cases, oral baclofen may be ineffective or require doses large enough to cause unwanted sedation.[7,8] To administer sufficient doses, intrathecal baclofen may be employed. Intrathecal baclofen is connected to an abdominal pump that directly

infuses the pharmaceutical into the intrathecal space.[10] Intraventricular catheters may be used as a last resort when traditional intrathecal catheters are not an option for narcotic pain medications. Intraventricular catheter placement allows a drug, such as baclofen, to disperse throughout the cerebral spinal fluid space, with potentially greater, more uniform distribution.[2] Our prior experience has shown that although intraventricular therapy is a rescue option, patients do poorly with this therapy especially in the area of spasticity control.

Early baclofen withdrawal typically results in itching, fever, confusion, and worsening spasticity but can progress to more severe symptoms, including full autonomic dysfunction syndrome and death.[1] These symptoms can emerge within hours of therapy cessation and progress rapidly.[8] Compared to oral baclofen, the withdrawal effects from intrathecal baclofen have a more rapid onset and carry higher mortality and morbidity.[6]

This case report describes a 22-year-old female with cerebral palsy who presented with severe withdrawal symptoms from a nonfunctioning intrathecal baclofen pump requiring intubation due to autonomic dysfunction. Due to her history of lumbar infections and prior scoliotic correction with Harrington rods and multiple prior intraventricular catheters providing inadequate spasticity relief, the decision was made to place a catheter in the prepontine space in an effort to improve the distribution of the baclofen the cerebrospinal fluid space.

CASE REPORT

A 22-year-old female with a past medical history of cerebral palsy and prior spinal fusion for scoliotic deformity with Harrington rod constructs from T1 to pelvis presented to the office with baclofen withdrawal symptoms secondary to a nonfunctional intrathecal baclofen catheter as diagnosed on pump interrogation. She also had signs concerning for an infection of her lumbar catheter site at the incision. She had an intrathecal baclofen pump initially placed for medically refractory spasticity and had undergone three prior revisions. Although tentatively scheduled to undergo a baclofen pump revision with an associated 5–7-h surgery that involved drilling through the Harrington rods to replace the intrathecal catheter, the patient went into moderate to severe baclofen withdrawal, prompting her to present to the emergency department with symptoms of worsening spasticity, tachycardia, and tachypnea with altered mental status. She was intubated and placed on a propofol drip for autonomic dysfunction syndrome and baclofen withdrawal.

Given her history of multiple pump failures and the added complexity of pump revision through a fusion mass, her small body mass, likely lumbar incision infection and autonomic instability, it was decided, she was risky candidate to go through a long surgery for a new lumbar catheter replacement, as detailed above. Oral baclofen was provided, but due to the high dose required by the patient, it was insufficient. Given the limitations of her anatomy and the infection concern, as well as the failure of oral baclofen to control her potentially lethal baclofen withdrawal, it was decided to proceed with placement of the catheter into the prepontine space in an effort to reduce the risk of it failing.

The patient was placed supine with her head turned to the left on a horseshoe head holder. An endoscopic third ventriculostomy (ETV) entry point was planned using the Stealth neuronavigation station. We used a C-shaped skin incision followed by an oval burr hole craniotomy with an acorn drill bit. The old baclofen pump was removed from the abdominal subfascial pocket and a new Medtronic Ascenda 8781 proximal catheter was tunneled from inferiorly to superiorly from the abdomen to this cranial incision.

The dura was opened with bipolar cautery and an 11-blade scalpel, using further electrocautery to cauterize the dura and the pia. On a single pass, we placed the 16-gauge sheath with the aid of neuronavigation to enter the right lateral ventricle. We then passed the MINOP Neuroendoscopy System into the right lateral ventricle through the sheath.

The endoscope was navigated into the third ventricle and an ETV was performed with forceps through the port [Figures 1 and 2]. A second pial opening was created slightly anterior to our initial pial opening and the distal baclofen pump catheter was then inserted into this opening and was advanced parallel to the MINOP. We visualized the entry into the right lateral ventricle. Using the endoscopic forceps and movement of the MINOP, we were able to guide the baclofen catheter into the third ventricle and then advanced it into the prepontine cistern under direct visualization [Figures 3 and 4].

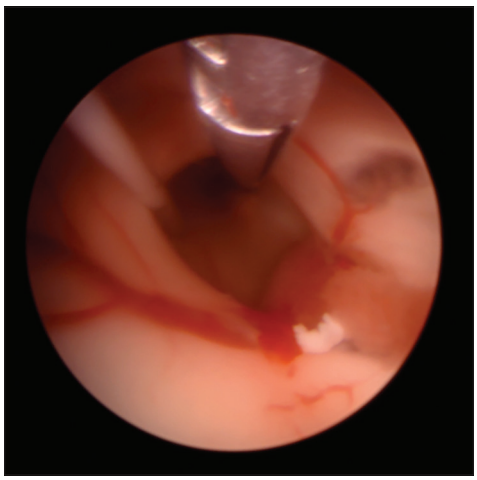

Figure 1: Endoscopic view from the lateral ventricle into the third ventricle. Alligator forceps are visible to the images right and the catheter itself on the left.

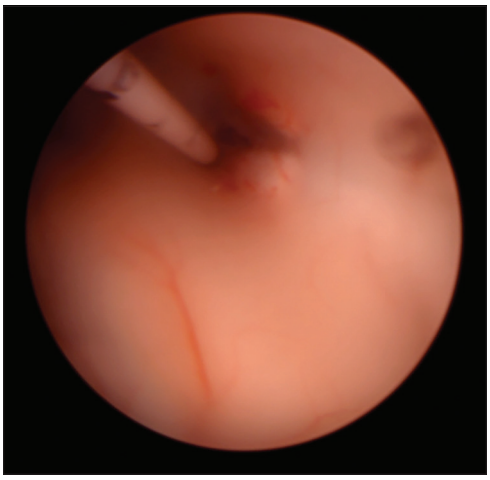

Figure 2: Intended site of the endoscopic third ventriculostomy.

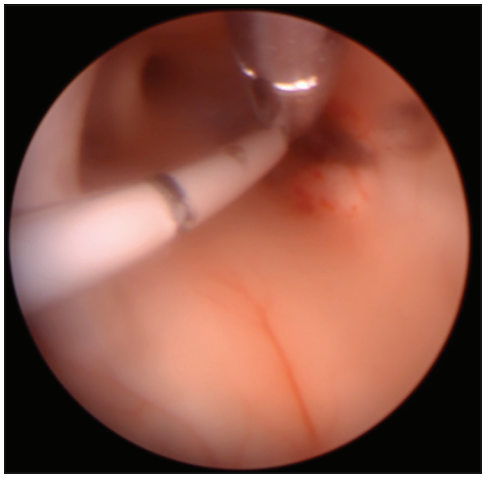

Figure 3: Using the alligator forceps, the tip of the catheter is inserted into the third ventriculostomy defect.

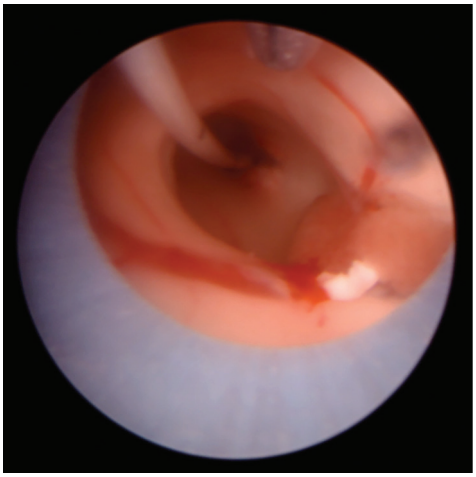

Figure 4: With the camera pulled back into the lateral ventricle, the baclofen catheter is directly visualized inside the third ventriculostomy defect, in the prepontine space.

We removed the MINOP making sure not to back out the catheter and removed the sheath. The intraventricular catheter was cut and measured to the appropriate length and the straight connector was used to connect the proximal and intracranial portions of the catheter. A dog-bone plate and screws were placed on the skull and a 2–0 silk tie secured the straight connector and the catheters. We filled a new 40 cc pump sterilely with 500 mcg/mL of intrathecal baclofen and it was programed to start at 100.1 mcg/day (compared to her preoperative dose of 998.9 mcg/day. The incisions were irrigated thoroughly with antibiotic irrigation and closed. The patient was then taken back to the intensive care unit intubated.

Placement in the prepontine cistern was confirmed by a postoperative computed tomography head. By the time, the dose was increased to 200 mcg/day, the patient was extubated, as she no longer showed signs of autonomic dysfunction and the spasticity in her upper extremities was significantly reduced.

By postoperative day 5, the patient was at a dose of 400.28 mcg/day and was discharged home with minimal upper and lower extremity spasticity (modified Ashworth scores of 1 in uppers and 3 in lowers compared to 4 throughout preoperatively).

DISCUSSION

Cerebral palsy is a disorder arising from a non-progressive lesion in the developing brain commonly associated with increased muscle tone leading to dystonia in up to 25% of patients, often refractory to medical therapy.[2,4] In these cases, baclofen is effectively used to treat patients with spastic cerebral palsy and dystonic cerebral palsy.[5] In addition, baclofen has become widely used for its antispastic effects in patients with spinal cord injury, multiple sclerosis, and traumatic brain injuries.[8,12] It exerts its effects through an agonistic effect on the GABA-B receptors, producing an inhibitory effect on presynaptic transmitter release through the restriction of calcium influx into presynaptic terminals.[1,7,9,14] GABA-B also affects postsynaptic terminals by decreasing neuronal activity through increased potassium conductance.[7] The agonistic effect of GABA-B inhibits spinal reflexes which reduces muscle spasm and pain.[8]

Often high oral doses are needed before clinical benefits occur because baclofen does not readily cross the blood-brain barrier.[6,11] Consequently, orally administered baclofen may be ineffective in as many as 35% of cases because the required dose to achieve muscle relaxation may be prohibitively high.[8] As a result of this high requirement, oral baclofen doses are often limited by the unwanted side effects of gastrointestinal symptoms, hemodynamic problems, or oversedation, which can be lethal in some cases and are often

associated with a host of morbidities including pneumonia and urinary tract infections.[6,7]

In the instances where oral baclofen is a poor option, the medication may also be delivered through a surgically implanted catheter.[3,10,12] The catheter, typically intrathecal, is connected to a subcutaneous or subfascial pump that controls the release of baclofen from a reservoir that is easily accessible for periodic refills.[5,10]

If intrathecal catheters are not viable for various reasons, including infection, intrathecal scar tissue, spinal cord injury causing myelographic blockage, or difficulty with operative access to the intrathecal space, intraventricular catheter placements have been described.[2,3] Previous literature has commented that intraventricular placement increases the concentration of baclofen in the intracranial subarachnoid space, potentially circumventing these overdose effects, though we have not found this to be the case in our institution's experience.[4]

Prior reports have described intraventricular baclofen catheters implanted into the third ventricle instead of the lateral ventricles so that the baclofen can more directly drain to the subarachnoid space and distribute over the cerebral convexities where it produces its effects through the inhibition of the excessively stimulated premotor cortex and supplementary motor cortex.[2,3] Theoretically, catheter placement into the third ventricle decreases the likelihood that baclofen in the cerebral spinal fluid will stagnate in the lateral ventricles,[2] and also optimizes symmetrical subarachnoid drug concentrations.[4] Our prior experience with intraventricular baclofen catheters has not provided the desired clinical effect, as we have had to increase the baclofen dose to >1000 mcg/day without any significant improvement in spasticity. In our experience, it is not uncommon to have patients request that the catheter be moved back intrathecally and we have had success with that approach to this patient population. We are aware, Dr. Albright's group has the largest intraventricular intrathecal catheter experience in the world; our understanding is that we have had the second largest population with similar results.

In theory, prepontine baclofen catheter insertion provides superior benefits to lateral and third ventricular baclofen catheter placement. There could be a lower occlusion rate due to the reduced interference of choroid plexus that may occlude the catheter. We have found this concept to be true with regards to prepontine shunt catheters. In addition, the prepontine technique provides more direct access to the subarachnoid space communicating with the cervical, thoracic, and lumbar spinal cord. Prepontine proximal shunt catheters have been placed previously through a similar approach and have demonstrated safety and efficacy.[13] In the case of our patient, the prepontine catheter provided excellent distribution of baclofen, as her autonomic dysfunction and upper extremity spasticity improved significantly postoperatively. Her lower extremity spasticity did improve, but was not quite back to her baseline when she was reliant on an intrathecal catheter.

All in all, this patient did exceptionally well from the procedure and nearly improved to her baseline status except her lower extremity spasticity when she had a working intrathecal catheter.

Baclofen pumps and catheters are already prone to failure and any efforts to add surgical techniques to improve patients' outcomes and prevent future morbidity and mortality associated with baclofen withdrawal is always ideal.

CONCLUSIONS

Baclofen is often used to treat dystonia secondary to cerebral palsy through its agonistic effect on GABA-B receptors. It can be administered orally or through a catheter placed intrathecally or intraventricularly. Withdrawal can result in fever, worsening spasticity, or dysautonomia and can occur within 2 days of therapy cessation. Intrathecal or intraventricular baclofen causes particularly serious withdrawal effects.[6]

In cases of severe baclofen withdrawal due to dysfunctional intrathecal catheters, surgical repair is recommended. However, the replacement of intrathecal catheters may not be feasible. This case report demonstrates that prepontine catheter placement can be safe alternative. It can allow for a greater distribution of baclofen in the subarachnoid space and provides a higher clinical benefit compared to other intraventricular techniques.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

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