Indwelling intrathecal catheter with subcutaneous abdominal reservoir: a viable baclofen delivery system in severely cachectic patients

Technical note


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Intrathecal baclofen (ITB) is a reversible treatment that reduces muscle tone to ameliorate spasticity and dystonia in patients with cerebral palsy (CP). The resulting decrease in energy expenditure allows patients to gain much-needed weight, albeit temporarily. Modern techniques require sufficient abdominal musculature and subcutaneous fat to permit the implantation of an indwelling pump. In patients with extremely low muscle bulk, visceral pumps may be impractical or impossible, with increased risks of dehiscence and infection. The authors describe a variation of the classical procedure in a young patient with severe cachexia.

A 10-year-old boy with spastic-dystonic quadriplegic CP was admitted to the neuromedical unit. Numerous drug trials had failed, and surgical intervention was deemed necessary but was complicated by his cachectic body habitus. The authors inserted a lumbar intrathecal catheter and subcutaneously tunneled it to the anterolateral abdomen, where it was connected to a subcutaneous injection port. Baclofen was continuously infused into the subcutaneous port using a noncoring needle connected to an external pump. The needle and line were changed every 5 days to minimize the risk of sepsis.

Although other techniques, such as intraventricular baclofen delivery, have been described, these are largely dependent upon sufficient musculature to support a visceral pump. A subcutaneous injection port system represents an alternative approach that reduces the risk of sepsis and may be better tolerated in cachectic patients.

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**Key Words** • intrathecal baclofen • spasticity • dystonia • technique • status dystonicus • cachectic • cerebral palsy • functional neurosurgery

**Abbreviations used in this paper:** CP = cerebral palsy; DBS = deep brain stimulation; ITB = intrathecal baclofen; SD = status dystonicus; SDR = selective dorsal rhizotomy.

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**Intrathecal baclofen (ITB)** was first described as an effective and reversible treatment for neuromuscular spasticity secondary to spinal cord injury. Baclofen is an agonist of the inhibitory neurotransmitter, γ-aminobutyric acid. Intrathecal delivery is useful because of the drug’s limited ability to penetrate the blood-brain barrier. Usually, an indwelling intrathecal catheter is subcutaneously tunneled to the anterolateral abdominal wall, where it is connected to a subfascial intrathecal pump that delivers the drug to its target site. The site itself is dependent on indication, and the catheter tip is usually placed at C5–T2 for spastic tetraparesis and C1–4 for generalized secondary dystonia. Intrathecal pumps contain built-in reservoirs that can be refilled as appropriate, under ultrasound guidance if necessary. Reductions in spasticity and dyskinetic movements release muscles from chronically contracted states, thus decreasing energy expenditure and allowing weight gain and symptom relief. In fact, weight gain may be extreme in some cases. Intrathecal baclofen has been shown to be cost effective in controlling spasticity when noninvasive interventions have failed, in conditions such as traumatic brain injury, spinal cord injury, and cerebral palsy (CP). It provides an important intervention in the last group of patients (those with CP) given that almost 75% of them exhibit spasticity. Furthermore, dyskinetic CP responds poorly to medication, so neurosurgical intervention is often necessary. Due to their high energy expenditure caused by
chronically contracted muscles, many patients with CP are underweight, making them unsuitable candidates for indwelling device implantation. Nutritional interventions beyond gastrostomy have a limited evidence base and do not solve the fundamental problem of calorie usage. Here we describe the case of a severely cachectic patient with spastic-dystonic quadriplegic CP, in whom we employed a novel strategy to deliver ITB.

Case Report

A 10-year-old boy with a known history of spastic-dystonic quadriplegic CP was admitted with emerging severe status dystonicus (SD). The patient was born prematurely at 25 weeks’ gestation, with periventricular leukomalacia visible on MR images. He had a tendency to exhibit athetoid and dystonic posturing in his upper limbs, which increased with activity and frustration. His truncal control was limited, and his minimal rolling ability caused him considerable discomfort during dystonic episodes. He also had marked hip adductor spasticity that had responded poorly to intramuscular botulinum injections. The patient had been previously treated by the orthopedic team due to bilateral dislocated hips that had been reconstructed prior to admission and had exacerbat-ed his dystonia. His gross motor functional classification system score was Level V, but he retained good verbal communication and cognitive function. His symptoms had been largely unresponsive to numerous medications, including high-dose oral baclofen, diazepam, tetrabenzine, morphine, chloral hydrate, trihexyphenidyl, and levodopa. Clonidine provided marginal benefit, but his weight remained extremely low at 16 kg (below the 0.4th centile), with a cachectic body habitus. A gastrostomy tube was trialed, but neurosurgical intervention was ultimately deemed necessary.

An intrathecal catheter was inserted into the lumbar spine with a standard technique and was subcutaneously tunneled to the anterolateral abdominal wall, where it was connected to a subcutaneous titanium injection port with a self-sealing silicone septum (INFU-KT, FB Medical). A noncoring 19-gauge needle (GRIPPER Needle, Smiths Medical) and line were used to continuously infuse baclofen into the reservoir via an external bedside infusion pump (Perfusor Space, B. Braun Medical Inc.) (Fig. 1). The GRIPPER needle was changed every 5 days, a regimen routinely employed for such ports, and the infusion set was changed every 2 days to ensure drug preparation stability. To prevent fluctuations in dose due to infusion set or needle changes, we administered a very low concentration (50 µg/ml). The device remained in situ for approximately 6 months, during which time baclofen was titrated up to a dose of 700 µg/day (0.58 ml/hour). The patient’s dystonia and spasticity improved considerably during this time, which allowed us to reduce the diazepam and morphine doses that had a sedative effect, causing respiratory depression and bowel dysmotility, as well as allowing a considerable weight gain of 8.5 kg (50% of his initial body weight). Unfortunately, the patient’s postoperative course was eventually complicated by an infection of his central venous access device (Portacath) for intravenous drug administration, which was removed. He then developed an encephalopathy, and a lumbar puncture revealed evidence of Pseudomonas meningitis necessitating device removal and treatment with antibiotics. His recovery was uneventful, and although consideration was given to the placement of a permanent indwelling pump, the patient’s weight was still too low to accommodate it. Nevertheless, he had gained a significant amount of weight (recorded at 22.5 kg, 0.4th centile), which was credited to the beneficial effects of our treatment plan. Unfortunately, the patient has required high doses of numerous sedating drugs that have prevented him from conversing and interacting, which he was able to do while receiving ITB. He is currently awaiting implantation of a deep brain stimulation (DBS) device.

Discussion

A recent classification system for CP distinguished between spastic and dyskinetic forms. However, both commonly coexist in the extremities of patients with CP. Dyskinetic CP does not respond well to medication, and there are limited treatment options that cause side effects that may outweigh their benefits. Spasticity is more common than dyskinesia, and CP is the most common cause of this upper motor neuron syndrome in children. However, not all patients with spasticity are candidates for treatment, and a careful consideration of functional status and other impairments is imperative for each individual. Indications for treatment include severe pain, desired posture improvement, and to limit the possibility of deformity. Although a number of drugs are licensed for treating spasticity, there is a limited evidence base for their use. Diazepam appears to be particularly effective, but its sedative effects and risk of dependence are well documented. A few studies have supported the use of clonidine, which appeared to be particularly effective in our patient. Certainly, the high drug doses required to control spasticity may be beyond the current evidence base. Pharmacotherapy is further complicated by the coexistence of spasticity and dystonia, which may reduce treatment effectiveness. For these reasons, neurosurgical management may be warranted in a subset of patients.
Intrathecal catheter with subcutaneous abdominal reservoir

Two neurosurgical interventions are commonly employed in patients with CP: selective dorsal rhizotomy (SDR) and ITB pump implantation. The former involves permanent severing of selective dorsal spinal roots to inhibit the myotatic stretch reflex. However, it has been suggested that patients best suited to this procedure are those with spasticity predominating over dystonia, which fails to improve with SDR. Caution is advised even in patients with spastic quadriplegic CP because a significant component of the hypertonia is likely due to concomitant dystonia. Although spasticity reduction is maintained in the long term, this procedure does not seem to preclude the need for additional orthopedic interventions, making its usefulness questionable.

Intrathecal baclofen is a reversible alternative that can be used in patients with dystonia and is usually preferred to SDR. The selection for this procedure is usually based on a trial ITB period with drug delivery via lumbar puncture or an external pump. Evidence for ITB therapy is currently limited, but initial experiences are promising even in nonambulant patients. However, indwelling devices are difficult to implant and poorly tolerated by patients with low muscle bulk, who require special consideration of implantation site and sepsis risk. Older techniques involved the creation of a craniocaudal subfascial pocket for the pump, but placement of the suture line directly over the device was associated with risks of wound dehiscence and infection. Kopell et al. improved this technique by placing the suture line either above or below the device, but their decision to cut the lateral fascial thickening of the rectus sheath, the linea semilunaris, may weaken the anterior wall of the pump.

Ammar et al. recently described a variation of this technique in which they extended the fascial incision laterally without severing the linea semilunaris. The lateral extension of the incision allowed them to cut the red fibers of the external oblique muscle and fashion a plane between the internal and external oblique muscles. This allowed greater coverage of the pump by the external oblique muscle and also allowed for the internal oblique muscle to act as the pump’s posterior wall. The group reported that this technique could be used in children weighing just 7 kg, with infection necessitating pump removal in only 5.5% of all cases. Rocque and Albright employed a different approach and described their experience with an extraabdominal pump implantation in 4 patients. They fashioned an infraclavicular pocket by dissecting a plane beneath the pectoralis fascia, but their suture line ultimately passed over the top tenth of the pump. Although this technique may be expected to cause more discomfort for patients, they did not find this to be the case in their small cohort. Pumps located in infraclavicular sites may be unsightly for patients, and their size is restricted by the smaller subfascial pocket, with an increased risk of sepsis due to the need for more frequent refilling.

Albright recently proposed baclofen delivery directly into the third ventricle, but this method does not bypass the need for a visceraally implanted device. Albright and Ferson used an adaptation of the technique for ventriculoperitoneal shunt placement in which they endoscopically inserted a catheter into the third ventricle and subcutaneously tunneled it to an abdominally located pump. This technique has been employed in patients with dystonia, multiple contractures, and spasticity, with good results reported in each scenario. A particularly troublesome drawback of ITB pumps is their high rate of revision, and the intraventricular route may be used as salvage therapy for patients requiring multiple revisions. Of course, the risks of cranial and spinal surgery must be carefully considered, but the spinal approach is arguably the safer method. Until the long-term outcomes of intraventricular baclofen delivery are known, widespread adaptation is likely to be limited.

Options for treatment may be limited during “dystonic storms” or SD. While there is no universal definition for this disorder, that proposed by Manji et al. is still widely used: “the development of increasingly frequent and severe episodes of generalised dystonia which had necessitated urgent hospital admission.” Status dystonicus is a life-threatening condition, with a mortality rate of approximately 10%. Precipitating causes include infection, medication change, and surgery. So careful thought should be given to minimizing these factors in patients with dystonia. As with dyskinetic CP, response to oral drugs is generally poor, with one series reporting that just 10.1% of patients responded to treatment. Tetra- benzine and benzhexol may be efficacious, but the majority of patients who receive these drugs require sedation and further intervention. Previous reports have found ITB to be efficacious in this setting, but evidence to the contrary also exists.

Narayan et al. have described postoperative SD in an adolescent with CP who required muscle paralysis and ventilatory support. Although numerous drugs failed, the patient showed a remarkable response to ITB, which allowed him to be weaned off ventilation and eventually discharged. However, ITB is not without complications, which include catheter migration, withdrawal syndromes, and overdosage, although the last problem is less likely with our setup. Alternatives to treat SD include DBS, which has replaced older lesional procedures such as thalamotomy and pallidotomy. Deep brain stimulation appears to be particularly effective for treating SD, but the risks of brain surgery may preclude surgeons from utilizing it as a first-line intervention. In routine settings, the hardware failure rate may be as high as 15%, and it can be even higher in patients with SD. Infection risk is also a concern of DBS in the setting of SD. Walscot and colleagues reported their experience with this combination of treatment and presentation in 3 patients, of whom required unilateral electrode explantation after a generator site infection developed. The patient’s recovery was uneventful after antibiotics were administered, but this case highlights the risk of deep brain infection. Intrathecal baclofen represents a safer temporary measure to address SD and allows for riskier procedures to be deferred until patients are more stable.

Most neurosurgical interventions to treat dystonia and spasticity are not adapted to patients with extremely low muscle mass. Such patients may lack the necessary musculature to support bulky, indwelling pump devices. Intrathecal baclofen represents an alternative to ITB but has the added risks of cranial surgery without remov-
ing the requirement for a visceral pump. The procedure described by Albright and Ferson may also result in complications similar to those of ventriculoperitoneal shunts, which may be poorly tolerated by malnourished patients.3

We describe an intrathecal catheter with an implantable injection port connected to an external pump as a viable alternative in patients with extreme cachexia. A more definitive self-contained system can be implanted after sufficient weight gain.

Disclosure

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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