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Baclofen Pump Implantation for Cerebral Palsy

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<http://dx.doi.org/10.5772/intechopen.79619>

Abstract

Programmable baclofen pump implantation is used to provide the patient with minimal intrathecal dose of baclofen to provide relaxation when the oral permitted doses are no longer withstand able by the patient. We discussed the efficiency of programmable baclofen pump implantation in treating spasticity by reviewing several international papers. Satisfactory relaxation was noticed in most of the patients. The complications following intrathecal baclofen (ITB) surgeries are not uncommon. ITB is an advised method for treating spasticity whether due to cerebral or spinal causes. It has significant improvements with minor complications. It needs special trained multidisciplinary team to manage it.

Keywords: spasticity, baclofen, drug infusion pump, treatment, cerebral palsy

1. Introduction

Spasticity is defined as a velocity-dependent resistance to the passive movement of a joint and its musculature [1]. It usually occurs with hyper-excitability of the stretch limb reflexes. It is related to the loss of inhibition from descending supra spinal neurons.

The contraction of the skeletal muscles can cause involuntary jerky like movements with difficulty in relaxation, coordination and movement control. It is often seen with ordinary activities like changing positions, stretching or by just touch. The severity of the spasticity is quite variable from patient to another and even with the same level or type of insult.

Spasticity is not always treated, because, in many conditions, it compensates for the loss of motor power helping the patient to mobilize. However, treatment indicated when the hypertonicity causes significant functional impairment or limb and spine deformities.

Modified Ashworth rating scale	
0	Normal muscle tone
1	Slight increase in resistance, catch with movement
1+	Catch plus minimal resistance through a range of motion
2	More marked increase but limb easily flexed
3	Considerable increase in the tone throughout the range of motion
4	Affected part rigid

Table 1. Modified Ashworth rating scale of spasticity [1].

Spasticity may result from either cerebral lesion (e.g., dystonia and cerebral palsy) or spinal cord lesion (e.g., spinal cord tumors, injury and syrinx).

The degree of spasticity is usually assessed using the Modified Ashworth rating scale (**Table 1**). Those with grade 3 and above are regarded as disabled persons.

2. Treatment modalities

2.1. Non-pharmacologic methods

A variety of physiotherapeutic modes are applied in order to stretch the muscles and increase their range of motion:

1. Splints (orthotics) or braces and taping.
2. Electrical or vibratory stimulation to the opposing muscle to attempt to relax the spastic muscle.
3. Physical and occupational therapy.
4. Massage, ice or heat
5. Serial or inhibitory casting – by this process we cast the limb repeatedly and progressively closer to the desired position so as to “reset” the muscle stretch reflexes and to lengthen muscles/tendons.

The advantage to these non-pharmacologic measures is that they have minimal side effects and can be used in addition to medications. However, in many patients, these measures are not sufficient to control the muscle tightness or spasms to the desired extent.

2.2. Pharmacologic methods (medications)

2.2.1. Regional (*in situ*) treatments

Injections of anesthetics, nerve blocks or neurotoxins such as botulinum toxin (Botox) are used if spasticity that requires treatment is limited to one or a small area of the body.

2.2.2. Oral medications

Patients with more profound spasticity need oral medications. They include baclofen, tizanidine, clonidine, clonazepam, gabapentin and dantrolene.

Although these oral medications are very effective in treating spasticity, they have several disadvantages. Among them sedation (sleepiness) and fatigue. They may need periodic blood monitoring for some of these drugs that can have potential side effects on liver function or blood counts and some may interact with other medications.

2.3. Surgical methods

The surgical treatment can be either ablative procedures, such as dorsal entry zone rhizotomy or augmented therapy such as baclofen pump implantation. The augmented therapy is a neuromodulation technique which modulates the function of the nerves without creating irreversible damage to any neuronal element. All these surgical options are provided only when other modes of treatment fail to relax the patient.

Baclofen pump implantation is a surgical procedure performed to permanently implant a pump that delivers baclofen to the spinal fluid to treat severe to moderate-severe spasticity that is refractory to oral medications.

As compared to baclofen taken by mouth, the direct infusion into the intrathecal space will reduce the amount needed to provide the same degree of relaxation. We can give even higher doses that cannot be tolerated by the patient when taken orally. It will limit or eliminate the undesirable side effects associated with taking oral baclofen. The pumps can provide a consistent and precise dosage throughout the day. It will avoid the peaks and valleys of taking oral baclofen. The dosing is very flexible and can be programmed in many ways, for example, we can give continuously the same dose for the whole day or administer on an hourly basis for 24 h a day, to different doses at different times of the day or the week.

There are two types of drug infusion pumps:

2.4. Fixed-rate pumps

This is now historical, and no one is manufacturing it any more. They operate mechanically, no battery is required. It operates with a butane gas pressure chamber surrounding a flexible inner reservoir. There are either 20 or 40 mm drug reservoir to minimize refilling while keeping overall dimensions as compact as possible.

Gas in the pressure chamber is warmed by body heat and expands, squeezing the inner chamber to drive the drug through a filter then to the catheter ending to the site of action in the body. The device designed to provide highly accurate flow rate.

Depending on treatment needs, the dose of spasticity medication is adjusted to individual needs, making the device operation simple and safe.

The pump is designed for life long. The purely mechanical pump operates without a battery.

Changes in pressure or temperature have influence on the flow rate, especially in hot countries. A baclofen overdose reported in some cases during summer time in several hot countries.

2.5. Variable-rate pumps

These are now widely used nowadays. These pumps have a drug reservoir from which they automatically dispense a programmed amount of medication through a catheter. These pumps have a small, on-board battery and integrated microelectronic circuits to control the drug delivery.

In this chapter we will discuss the rationale for the use of this mode of treatment in spasticity.

3. Indications for ITB

Baclofen pump implantation is indicated if patients have:

1. Sever spasticity grade 3 and above (equal or more than 12 months duration). This spasticity significantly interfere with voluntary movement, causing difficulty in maintaining range of motion or position. It even affects safety, contributing to pain or skin breakdown and making personal care difficult for you or your caregiver.
2. Spasticity refractory to oral medications (baclofen) or having unacceptable side effects.
3. Positive response to ITB at test dose 100 µg with no response to placebo.
4. No hypersensitivity to baclofen.
5. No medical comorbidity to surgical intervention.

Patients with profound pain are preferred to have morphine pumps rather than baclofen.

4. Screening test before implantation

All candidates for intrathecal baclofen (ITB) therapy were planned to perform screening trials before pumps implanted. The patient receives 25 µg of baclofen through lumbar puncture. The resulting hypotonia is measured using the Modified Ashworth score. Pulse rate, blood pressure, respiratory rate and any adverse effects (including seizures) should be recognized. A test is successful if the Ashworth score is reduced by at least two points from 4 to 8 h following administration. If this reduction is not achieved, a bolus dose is given the next day, with an increment of 25 µg up to a maximum bolus of 75 µg. The drug amount that reached a satisfactory test result is also used to guide us to give the initial dose after pump implantation. If the response obtained with 25 µg for 24 h or more, then the initial dose of the implanted pump will be 25 µg/day; if the duration of relaxation were less than 24 h, the initial dose of the implanted pump would be 50 µg/day.

5. Operation of pump implantation

The pump is inserted under the covering of the abdominal muscles, while the patient is under a general anesthetic. A small catheter is then inserted through a Touhy needle into the thecal sac. When spinal fluid appeared, a spinal catheter threaded upward to a site depends on the distribution of the patient's spasticity (i.e., to the T10-11 spinal segment level for patients with spastic paraplegia and the C7-T4 spinal segment level for those with spastic quadriplegia). The catheter is then tunneled under the skin to the abdomen and is connected to the pump. The incision is closed with suture material or surgical staples. The procedure usually lasts 1–2 h.

6. Dosing of intrathecal baclofen

After implantation, we will fill the pump with baclofen, the pump is then programmed with certain external programmer, for example, N-vision. In all the cases, we prefer to use the "simple continuous infusion". ITB doses are titrated over the next few months. Initially, the infusion often begins at 25–50 µg/day and the dose is titrated gradually by 10–20%, a week increment until the patient's spasticity significantly reduced. The dose then adjusted with less frequent periods on an outpatient basis. In most of the patients, the satisfactory dose is usually achieved within 2 months.

Refills are generally done every 3–9 months, a refill kit is provided with a sterile needle and syringe. The filling is done in the outpatient just by passing the needle to the pump through the skin of the abdomen. Pumps hold 20 or 40 ml and will signal with a beeping noise if the amount gets below 2 ml. The devices also sound alarms if there is a malfunction, or if the battery runs low (generally between 3 and 7 years of use). When the battery runs low, the implant will be replaced under local anesthesia.

7. Prognosis

We conducted a study of 55 patients with spasticity, who had baclofen pump implantation in Neuroscience Hospital, Baghdad, from October 2011 to December 2016 [2]. The spasticity decreased, significantly in the lower extremities and remained so during the 2.9 years of infusion. In 55 patients (41 patients with a spinal cause of spasticity and 14 patients with spasticity caused by cerebral palsy), the results were remarkable.

Comparing our data with other international studies (**Table 2**) [3–10], the 2.9 years follow-up quite objectively concluded the efficiency of use of the pump to treat spasticity.

Many review papers were published to assess the efficiency of the baclofen pump. Butler and Campbell [11] in 2000 reviewed 12 published studies on ITB. They concluded that spasticity improved and that limb functions probably improved with ITB.

Reference	Country	Patient no.	Type of study	Result
Kvascevicus et al. [5]	Lithuania	5	Prospective	Very good
Baker et al. [6]	United States	117	Prospective	Excellent
Penn [7]	United States	18	Double blind	Excellent
Coffey et al. [8]	United States	75	Double blind	Excellent
Muller [9]	Germany	211	Prospective, multicenter	Excellent
Lazorthes et al. [10]	Belgium/Holland	18	Prospective	Excellent
Jierski et al. [11]	Germany/Sweden	28	Prospective, multicenter	Excellent
Penn et al. [12]	United States	62	Prospective	Excellent

Table 2. Comparison of the outcomes from international data.

In our study, we concluded that spasticity decreased significantly in the lower limbs and to a lesser extent in the upper extremities. The Ashworth scale remains low during the years of infusion. Westbom et al. in 2003 reported in a multicenter study a sustained decline in the Ashworth scores in both lower and upper extremities during a 6-year follow-up [12].

The reduction of upper limbs spasticity is usually less than that noticed in the lower extremities; if the catheter tip was at the mid-thoracic level or lower. Motta et al. [13] evaluated ITB's effect on the upper extremities of 20 patients with quadriplegic cerebral palsy with a mean age of 11.4 years. The spasticity of the upper extremity decreased ($P < 0.05$), and the range of movement improved remarkably.

The range of motion in the lower limbs increased following ITB therapy due to the reduction of spasticity. Further, this accompanied by an improvement in the gait. In our study, improvement in gait noticed in 10 patients (4 with multiple sclerosis, 3 with cerebral palsy and 3 with partial spinal cord injury). Gerstzen et al. [14] studied the effect of ITB on gait. They classified gait functionality into four types (community, household, non-functional and non-ambulatory). In their study, 24 patients had some ambulation before ITB therapy. The level of ambulation was improved by one level in nine patients, worsened in three and unchanged for the remaining patients.

8. Complications

1. Pain, numbness, weakness or paralysis due to nerve damage (rare)
2. Cerebrospinal fluid leak
3. Bleeding/injury to blood vessels
4. Infections
5. General anesthetic complications
6. Hardware-related complications during surgery are rare, but can include catheter migration. After surgery though, the potential hardware complications include catheter fracture or migration and infection or pump malfunction.

The most frequent complication was catheter malfunction (e.g., disconnection of the catheter tip from the pump, drug leakage from the catheter and migration of the catheter outside the thecal sac).

8.1. Baclofen withdrawal

Sudden cessation of ITB infusion can occur when the pump running empty; programming error; incorrect baclofen concentration; a problem with the catheter system and a problem with the battery. Some of these problems will activate the alarm. It is followed within a few hours by symptoms of generalized pruritus, increased spasticity and agitation. The symptoms may range in severity from mild to severe [15, 16]. The Classic symptoms of baclofen withdrawal are a sudden increase or return of your spasticity or tone, profuse sweating and itching without an associated rash. Fever, tachycardia, tachypnea, hypotension or hypertension or even confusion. Severe withdrawal symptoms include hallucinations or delirium, seizures, rhabdomyolysis, organ failure and even death. We noticed some patients might not recognize the mild withdrawal symptoms; consequently, the interruption in therapy may not detect. Severe withdrawal symptoms are severe complications and must manage urgently. Moderate withdrawal symptoms are managed by high doses of oral baclofen (e.g., 10–20 mg every 4 h), intravenous benzodiazepines (diazepam, 2–5 mg every 6 h) or bolus baclofen injections can be given too [17]. Despite the complications reported in many papers, almost 90% of patients with baclofen pump implantation at the end of the battery life prefer to have a new pump exchanged and to continue ITB therapy [18].

8.2. Baclofen overdose

This is far less common than withdrawal. The overdose could be a human error in dosing, programming or filling the pump; system malfunction or unsafe combination of intrathecal and oral medications. Most of the time, mild overdose symptoms can be easily managed by turning the pump rate of infusion down. The mild symptoms are characterized with low muscle tone, low attention, lightheadedness and sleepiness. Moderate symptoms are bradycardia, respiratory depression and difficulty awakening. When sever symptoms developed the hypotonia spreads to the trunk, arms, face and neck. Stupor, coma, seizures, severely slowed breathing that needs mechanical ventilation and eventually death. Some papers recommend an intravenous injection of 1–2 mg of physostigmine to treat overdoses, but their therapeutic effects are minimal and brief [21, 22]. Severe overdoses may necessitate assisted ventilation. CSF aspiration may also help with severe overdoses: 20 ml of CSF is withdrawn and replaced with 20 ml of a 5% dextrose to 0.25% normal saline solution 2 or 3 times [22].

8.3. Infections

Infection occurs in 5–10% of patients mostly caused by *Staphylococcus aureus* [19].

Most infections that start in the first 3 weeks after surgery are due to bacterial contamination at the time of pump insertion. Those related to future pump refills are extremely rare. Infections can occur either at the site of pump implantation, that is, anterior abdominal wall. This will result in erythema, swelling, tenderness and fever. It is treated with intravenous antibiotics and surgical cleaning of the pump wound. In severe cases, the infection may affect the whole

system, including the catheter and cerebrospinal fluid (CSF). It requires removal of the entire pump with postoperative parenteral antibiotics for 3 weeks [19].

8.4. CSF leaks

Leak may appear due to insertion of Tuohy needle with a catheter. The incidence is from 5 to 15% in patients with CP (most of whom are children), in contrast to the 3% leak rate reported in adults. The difference explained by malnutrition of chronically disabled children, thinner tissue masses, the smaller body size and the presence of higher CSF pressures associated with occult hydrocephalus [20].

9. Regular checkup and replacement

This process is quick. It involves “interrogating” or analyzing the pump using the handheld programmer, for example, N-Vision Programmer. We have to check whether the pump is working correctly and looking for any trauma at the pump site. If the patient has an MRI appointment, we must stop the pump before examination, then the pump is checked within 2 h after MRI to make sure the pump rotor has restarted itself after an expected stall because of the magnetic effect on the rotor.

Replacement is needed usually after 5–7 years. It is replaced with surgery. Unless there is a problem with the catheter system, the catheter will not require replacement, and the section near the pump will be simply reconnected to the new pump.

10. Conclusions

ITB is a good method for managing spastic cerebral palsy, especially in patients with some potentially useful extremities. Performing ITB requires a well-trained multidisciplinary team that can deal with patient education, pump refills and dose adjustments.

However, it is expensive and associated with some complications; yet, it is one of the best therapeutic options available for patients with spasticity.

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