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Spastic paralysis after sustaining cervical spinal cord injury: 13 years on a baclofen pump — case report

Abstract

Spasticity is a very troublesome symptom, which causes further deterioration in physical disability and in many cases precludes effective treatment. The aim of this paper is to recall different methods for treating spasticity, with particular consideration of the modern method of employing a baclofen pump with a patient who had suffered trauma to the cervical spinal cord.

Key words: spasticity, baclofen, baclofen pump, cervical spinal cord

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Introduction

Spasticity is a very troublesome symptom, which causes further deterioration in physical disability and in many cases precludes effective treatment. The aim of this paper is to recall different methods for treating spasticity, with particular consideration of the modern method of employing a baclofen pump with a patient who had suffered trauma to the cervical spinal cord.

According to Lance's classical definition, spasticity is a motor disorder characterized by a velocity-dependent increase in tonic stretch reflexes (muscle tone), resulting from the stretch reflex as one component of upper motor neurone syndrome [1, 2].


Its modification by Young and Sheen points to cortical control of muscle tension involving pyramidal, reticulospinal, vestibulospinal and rubrospinal tracts, as much as reflex control by alpha motor neurones, excitatory and inhibitory interneurons and polysynaptic reflexes. The spinal control mecha-

nisms, such as the Renshaw cell system and Golgi tendon cells, take part in controlling and also in supraspinal actions such as cerebellum and reticular formation, the cerebral cortex and the autonomic system [2, 3].

Spasticity results from an increased activity of the gamma motor system. Thus, it is the result of distortion in balance between mechanisms activating and inhibiting alpha and gamma motor neurones in the spinal cord. It can also be explained by the unlocking of control of spinal reflexes by supraspinal sites, which means the withdrawal of the influence of gamma motor neurones by inhibitory centres. This leads to increased activity of muscle spindles. A typical symptom of spasticity is called the clasp-knife response [2].

Soon after spinal cord injury, there is an initial phase of spinal shock, a term first used by M. Hall in 1841, which is characterized by the loss of all reflexes below the injured segment. At the time of losing cortical control over the spinal cord there is

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a great increase in the activity of motor neurones. This presents itself in spasm: prolonged, uncontrolled and excessive contraction of skeletal muscles and pathologically exaggerated responses to every kind of stimulation. Subsequently, an increased and greatly spreading muscle contraction in response to stretching appears. Clinically, it is manifested by hyperactive reflexes: increased muscle tension due to passive movement. The spinocerebellar tract, which normally plays an important role in passing information from the peripheral to the central nervous system, becomes totally useless as the whole spinal organization loses meaning. Urinary bladder distension, skin irritation, the blowing of cold air, or any other stimulus, provokes the same response, independent of its location or significance to the organism. All stimuli may elicit an increase in spasticity. Loss of control by descending tracts leads to hyperactivity of interneurons with simultaneous, spontaneous, persistent neuronal firing. Freed from inhibitory and coordinating influence, the mechanisms within the gamma loop become excessively activated. Fast passive movement is met by increasing resistance which suddenly melts away — hence the term clasp-knife reflex. The reticulospinal tract, being the stretch reflex activator, also plays a role in sustaining spasticity. Increased muscle tension is accompanied by an exaggerated stretch reflex, especially within flexor muscles. Spastic muscle tone in paralyzed patients, with a tendency to flexion and adduction in the hip joint and overflexion at the knee, makes caring for them even more difficult. Other symptoms of spasticity include the following: exaggerated deep tendon reflexes, pathological reflexes, pathological synkinesis and clonus, autonomic system hyperactivity, paresis, muscle palsy, increased muscle weakness, lack of movement precision and loss of superficial reflexes [2, 4].

Spastic jerks may sometimes cause the patient to fall from a bed or wheelchair. Gradual limitation of movement within joints and contractures follows. Increased spasticity predisposes the patient to bedsores resulting from skin abrasions during involuntary limb movements. A considerable level of spasticity is a very annoying symptom, further exacerbating physical disability and quality of life. Thus, prevention or treatment of spasticity becomes one of the most important elements in the rehabilitation of patients with central paralysis. The treatment is aimed not only at increasing physical condition and a maximal reduction in disability, but also at prevention of complications such as bedsores, periarticular ossification, contractures, joint insta-

bility and luxation, foot deformities, and dysfunction of urinary and digestive systems. Proper treatment of spasticity also decreases pain sensations related to spasms and makes it easier to care for a patient [1, 2, 5].

One of the most modern methods of treatment for spasticity is therapy based on the subarachnoid infusion of baclofen. A surgically implanted dosimeter automatically dispensing regularly-spaced doses guarantees constant drug concentration in the spinal canal, which is necessary for the treatment to be effective. Side effects with baclofen treatment, such as hypotension, drowsiness, dyspnoea, coma, photophobia, respiratory disorders, convulsions, pain at the site of pump implantation and infection, are rare. Any serious symptoms most commonly result from a badly-set dose or simply in the overdosing of baclofen [5].

Case report

A 13-year-long observation of a patient, following injury to the cervical spinal cord, implanted with a baclofen pump in order to decrease spasticity of the lower limbs.

On November 13, 1994 the patient, now 52-years-old, fell from a height of 1.5 m and suffered trauma to the spinal cord at the C6–C7 level. Immediately after the injury he was treated surgically by stabilizing with the use of a titanium plate. The injury left him with paralyzed lower limbs, upper-limb paresis (P > L) with bladder and anal sphincters also paralyzed.

Due to a great degree of spasticity in the lower limbs, the patient stayed unresponsive to large doses (150 mg per day) of baclofen taken orally and suffered from a great deal of pain (NRS: 8). The decision to implant the baclofen pump was taken in order to free the patient from this. Constant infusion of baclofen to the subarachnoid space is indicated in cases of lack of response or side effects from oral treatment with baclofen and positive results from a baclofen test. The baclofen test is based on injecting single doses (25–100 mg) of baclofen into the subarachnoid space. The interthecal method of injecting baclofen allows for reducing the drug dose more than 100 times with good therapeutic results [6]. The described patient met all the criteria. After receiving positive baclofen test results the procedure of pump implantation was performed on March 19, 1996 at the City Hospital in Koblenz, which was one of the first centres in Europe to implant baclofen pumps. The model ARROW-THEREX 3000 was implanted by Professor Muller. A sub-

arachnoid catheter was inserted at the L3–L4 level ending at T6. The capacity of the implanted pump is 30 ml and the optimal time required to empty it is 50 ± 3 days. The initial dose of baclofen at the time of implantation was 14 mg and it is presently set at 18 mg. The very small increase in the required dose of baclofen shows a lack of tachyphylaxis to the drug applied through the spinal canal.

Baclofen is a spasmolytic drug, derivative of GABA, and is commonly applied in the treatment of spasticity. It decreases skeletal muscle tone by inhibiting mono- and polysynaptic reflexes in the spinal cord, which is its site of uptake. The detailed mechanism of the action of baclofen is unknown but its relaxing effect is based on interaction with the alpha motor system and dependent on decreasing activity in the gamma motor system. Baclofen does not decrease conductivity within the neuromuscular junction or sensitivity of the sensory ending of muscle spindles. The oral dose varies from 10 to 150 mg per day. Baclofen is well tolerated but its side effects include dizziness, vertigo, muscle weakness, tiredness, headaches, sleeplessness, speech disorders, blurred vision, hypotension, nausea, constipation, skin rash, excessive sweating, increased activity of aminotransferases and alkaline phosphatase, increased glucose level and, rarely, depression. Contraindications to treatment with baclofen include hypersensitivity to the preparation, or peptic or duodenal ulcers. Caution is advised while treating psychiatric patients with schizophrenia or psychosis, epilepsy, stroke, and those with renal or liver failure. The direct infusion of the medication to the subarachnoid space through the intrathecal pump allows binding of the drug particles with the receptors in the dorsal roots, permitting treatment with 100 times lower doses compared with those administered orally, and it results in a weak penetration of baclofen through the blood-brain barrier. At present, baclofen is available as an injection preparation under the names of Loresal and Kemstro [7–10].

From the beginning of our patient's treatment, pump refilling was performed in our centre. Twice during our 13-year-long care of the patient, he was, due to urinary or respiratory infections, staying in other hospitals at the time of the required pump refilling. Since both hospitals had Pain Centres, on site doctors attempted to refill the pump. Unfortunately, the pump was damaged each time and baclofen intoxication followed, requiring mechanical ventilation and consequent pump replacement. The symptoms of baclofen intoxication included sudden respiratory failure, depressed states of con-

sciousness, muscle flaccidity and fever. Damage to the pump resulted from employing incorrect needles and poor procedural technique. The pump implantation took place in October of 1996 and August of 1998.

Due to the poor penetration of baclofen through the blood-brain barrier, the only effective way of applying the drug is directly to the cerebrospinal fluid. The most convenient method is constant infusion through the pump. In order to ensure its proper function it is required that the procedure of refilling is performed in a sterile environment and that proper needles are used. The best needles used for refilling are either the Hubner type needles or Port 22G, which have been specially ground to avoid membrane rupture. For the best results, the refilling should be performed in one centre.

Final diagnosis

Injury to the cervical spine at the C6–C7 level in November, 1994. Paraplegia. Neurogenic bladder. Cervical spine stabilization complicated by broncho-oesophageal fistula. Reconstructive operation of broncho-oesophageal fistula and pleural empyema in May, 1995. Resection of left inferior pulmonary lobe in January, 1996. Chronic bronchitis. Long-term treatment with baclofen. Baclofen pump implantation in March, 1996. Damage to the pump complicated by intoxication with baclofen and pump replacement in October, 1996 and August, 1998. HepB infection in 2000. Chronic urinary infection. Degenerative joint disease of spine and main joints, especially the hip. Obesity. Chronic constipation.

Discussion

Methods of treatment leading to the improvement or elimination of spasticity can be divided into three groups: physiotherapy, pharmacology and surgical. Physiotherapy is aimed at decreasing muscle tone, blocking pain receptors, decreasing impulse flow to the CNS and temporary weakening of nerve conduction [2, 3].

Pharmacological treatment includes preparations with general activity, such as baclofen, tolperisone, dantrium, mephenesin, myolastan, methocarbamol, pridinol, phenoprobamat and benzodiazepines and preparations acting directly on the spinal cord, nerve roots, motor points and peripheral nerves. Pharmaceuticals administered orally decrease the tonus of all muscles, including the non-spastic ones. Their side effects include a deterioration in cogni-

tive functions, drowsiness, weakness, confusion, vertigo, phlebitis or liver damage [2].

In patients suffering from a degree of spasticity great enough to prevent normal functioning, following the unsuccessful employment of all non-invasive methods of treatment, a surgical interruption of the reflex arch can be conducted. These surgical methods usually involve mielotomy, rhizotomy, DREZ-tomy or procedures on tendons and muscles [1, 2, 11].

Among the various methods of treating spasticity of different origin (spinal, cerebral or combined), there is a therapy based on the subarachnoid infusion of baclofen with the use of a pump. The method was first described in 1984 and is now widely employed in the treatment of spasticity due to sclerosis multiplex, spinal cord and cerebral injuries, infantile cerebral palsy and in dystonias.

The baclofen pump is of little more than 7 cm diameter (Figure 1AB) and is implanted in the sterile environment of an operating room. The pump is placed in a subcutaneous pocket, at a depth of 4 cm, on the front of the abdominal wall. It is connected with a catheter placed in the subarachnoid space, through which a permanent flow of baclofen ensures a constant concentration of the drug interthecally (Figure 2). The next step is to establish the minimum effective dose of baclofen. Immediately

after the implantation procedure the patient is examined in terms of respiratory rate, muscle tone, presence of spasticity and involuntary movements. The spasticity assessment is most commonly based on the Ashworth Scale, characterized by a high degree of data compliance and repeatability. Since 1987, a modified Ashworth Scale [12] has become more common for assessing spasticity (Table 1). This method of treatment is combined with the intensive education of patients and their families relating to the handling of the implanted pump. Physical rehabilitation reinforced by educating the patient in this subject takes place concurrently. Since the pump is a pneumatic device lacking electronic parts, its expected lifespan is about 25 years, provided it is not damaged at the time of repeated refilling through poor procedural technique or the use of incorrect needles (Figure 3).

The baclofen pump has been in use in Poland for the last few years in cities such as Poznań, Szczecin, Gdańsk, Kraków and Zakopane. Until recently, the NFZ had not been refunding the costs related to the pump implantation, since the equipment itself is quite expensive (accounting for 30,000 zł) with the cost of medication in addition to this. Gathering such funds was impossible for many patients. Increasing acceptance of this procedure by the NFZ has made a real breakthrough in accessibility of this method for patients with spinal injury, MS and infantile cerebral paralysis, and all others with proper.

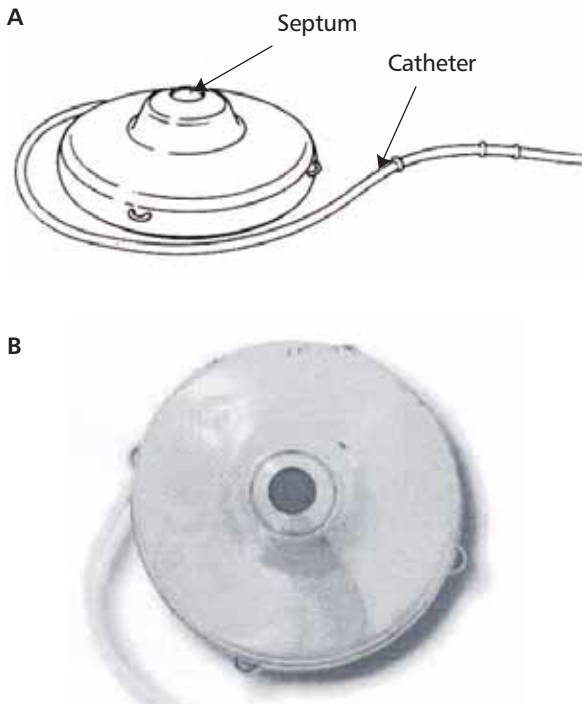


Figure 1AB. Baclofen pump

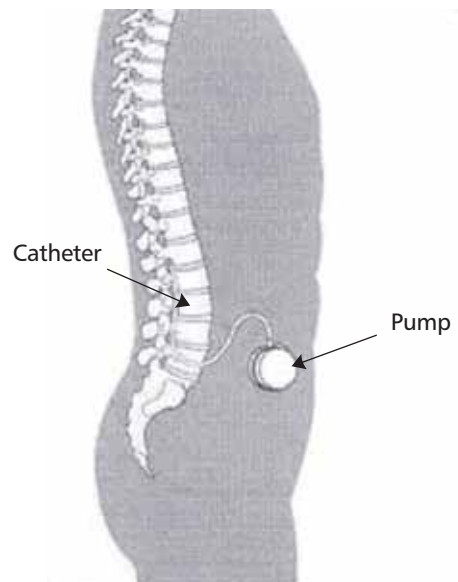
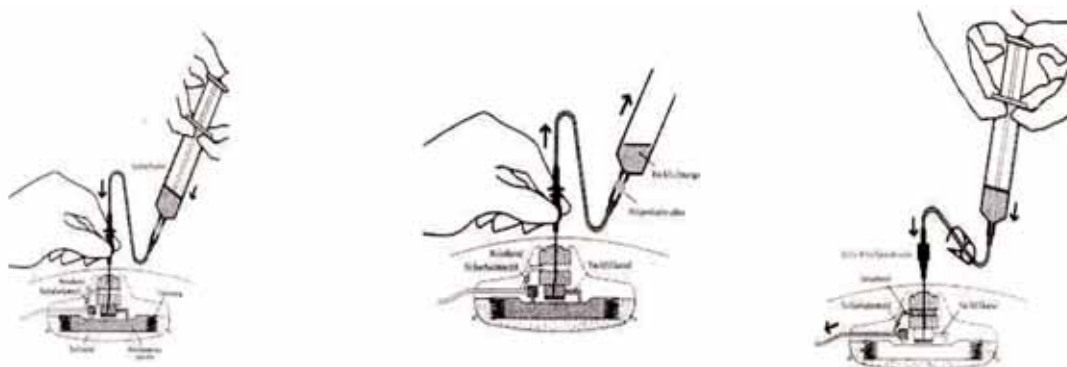


Figure 2. Catheter placed in the subarachnoid space. Permanent flow of baclofen ensures a constant concentration of the drug interthecally

Table 1. Modified Ashworth Scale for grading spasticity

0	No increase in tone
1	Slight increase in muscle tone, manifested by a catch and release or minimal resistance at the end of the ROM when the affected part(s) is moved in flexion or extension
1+	Slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the ROM
2	More marked increase in muscle tone through most of the ROM, but affected part(s) easily moved
3	Considerable increase in muscle tone, passive movement difficult
4	Affected part(s) rigid in flexion or extension

**Figure 3.** The pump filling up technique

The baseline for the lack of effective and long-lasting methods of treatment for spasticity is the complexity of its pathophysiology. Prior to making decisions about implanting the pump, it is imperative to examine the patient's physical condition carefully and thoroughly consider the indications and contraindications for this kind of treatment. A direct indication is the positive result of the baclofen test or a decrease in spinal spasticity by 2 points and cerebral spasticity by 1 point on the Ashworth Scale. Patients who meet the following criteria would qualify for pump implantation:

1. Severe spasticity due to infantile cerebral paralysis, multiple sclerosis or spinal cord injury.
2. Spasticity greatly limiting the patient's daily functioning and self-caring.
3. Improvement in functioning and self-caring can be prognosticated.
4. Spasticity is prolonged and has lasted more than 12 months.
5. The patient is over 4 years-old.
6. The implantation procedure can be performed at least 12 months after any previous surgery.
7. The patient does not tolerate, or spasticity is resistant to, orally administered drugs.
8. Any disorders within the spinal canal have been excluded.

9. The patients and their families are motivated and cooperate with medical personnel while deciding on this kind of treatment.
10. The patient's place of living is less than 200 km from the centre taking care of the patient.
11. Stabilized medical condition.

Significant contraindications to this kind of treatment are local or general infections, liver or kidney failure, allergy to baclofen, pregnancy, menopause, condition after cerebral haemorrhagic stroke, digestive system disease, and lack of cooperation. A more controversial contraindication is the patient's having a life expectancy of less than 1 year [13].

New longer-lasting and more effective methods of treatment for spasticity should still be searched for.

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