Pathophysiology of Spasticity

Spasticity is a major neuromuscular problem in CP. It is so deeply engrained in medical and public literature that a spastic child has come to mean a child with CP for most people around the world. Spasticity is difficult to define. The pathophysiology is obscure, findings on examination are inconsistent, and treatment is not always successful. Understanding the physiology of normal movement may help the physician in the management of spasticity.

Physiology of movement

Afferent input from the internal organs, the musculoskeletal system, and the skin converge on the medulla spinalis. This afferent input activates the stretch reflex, both directly and through the interneuron, and results in a reflex motor response [A]. The same afferent information goes to the cerebellum and the somatosensory cortex. It is processed in those centers as well as in the basal ganglia. The resulting motor response is relayed to the lower motor neuron through the pyramidal and extrapyramidal system tracts. The pyramidal tracts go directly to the lower motor neuron whereas the extrapyramidal tracts end at the interneuron. The cerebellum, basal ganglia, and extrapyramidal system nuclei modify the motor response as it goes to the medulla spinalis. In this way all motor output is influenced by the incoming sensory input and converges on the lower motor neuron. The interneurons in the medulla spinalis regulate the activity of the motor neuron.

Neural pathways regulating muscle contraction

The motor cortex is responsible for planning voluntary movement.

The corticospinal tracts carry movement order to the lower motor neuron.

The nerve impulse arising from the cerebral motor cortex is also sent to the basal ganglia and the extrapyramidal system nuclei.

The basal ganglia correct the timing of movement.

The extrapyramidal system corrects the force of contraction of the muscles involved.

The cerebellum coordinates the speed and direction of movement.

Muscle spindles in the contracting muscle, golgi tendon organs in the tendons and mechanoreceptors in the joints send information on the degree of contraction to the medulla spinalis, cerebellum and the somatosensory cortex.

The lower motor neuron sends contraction impulse to the muscle through the peripheral nerve. This is the final common pathway from the nervous system to the muscle.

These corrective impulses from the extrapyramidal system are sent to the interneurons in the medulla spinalis.

The interneurons send inhibitory or excitatory impulses to the lower motor neuron and regulate its activity.
Pathophysiology of Spasticity

1. Measurements in spasticity

   **Clinical measures**
   - Range of motion
   - Tone intensity measures
   - Modified Ashworth Scale
   - Tardieu Scale
   - Mechanical instruments
     - The pendulum test
   - Electrophysiological measures
     - The H reflex
     - Vibration inhibition index
   - Functional measures
     - Upper extremity function
   - Gait

2. Modified Ashworth Scale

   - **D**
     - 0: No increase in muscle tone
     - 1: Slight increase in muscle tone, manifested by a catch and release or by minimal resistance at the end range of motion when the part is moved in flexion or extension/abduction or adduction, etc.
     - 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 the affected part is easily moved
     - 3: Considerable increase in muscle tone, passive movement is difficult
     - 4: Affected part is rigid in flexion or extension (abduction, adduction, etc.)

3. The upper motor neuron syndrome

   CP results in an upper motor neuron syndrome [A] characterized by spasticity, exaggerated tendon reflexes, clonus, pathological reflexes, mass synergy patterns, muscle weakness, loss of selective motor control and loss of hand dexterity. Spasticity is a component of the upper motor neuron syndrome.

4. Definition of spasticity

   Muscles show a physiological resistance to passive motion. This is called muscle tone. Spasticity is the increase in this physiological muscle tone. The terms “spasticity” and “increased tone” may be used interchangeably. Spasticity is velocity dependent. The faster the passive movement, the greater the resistance of the muscle. The increase in muscle tone causes loss of trunk balance and difficulty of active movement in the extremities.

5. Pathogenesis

   The pathogenesis of spasticity is presumed to be an increase in the excitability of the lower motor neuron. This presents as hyperactive stretch reflexes [B] at clinical examination. Many hypotheses attempt to explain this hyperexcitability. One suggests a change in the balance of excitatory and inhibitory inputs to the motor neuron pool. When the inhibitory inputs are reduced, the interneurons send excitatory impulses to the lower motor neurons and they become hyperexcitable.

6. Measuring spasticity

   Spasticity can be measured by clinical examination, mechanical instruments, and electrophysiological techniques [C]. The modified Ashworth and Tardieu scales are commonly used for clinical evaluation. They measure tone intensity but do not evaluate the effect of spasticity on function. Mechanical instruments measuring the resistance of the muscle to passive stretch and electrophysiological measures showing the hyperexcitability of the stretch reflex are used only for research purposes.

   **The Ashworth scale**

   The Ashworth scale [D] is by far the most commonly used evaluation method for spasticity. Always test the patient while he or she is in a relaxed supine position. Passively move the joint rapidly and repeatedly through the available range of motion and grade the resistance using the definitions.
**The Tardieu scale** The Tardieu scale measures the intensity of muscle tone at specified velocities \[A\]. Note the joint angle at which the catch is first felt. Always grade the Tardieu Scale on the same day. Keep the body in a constant position for a given extremity. Keep the other joints, particularly the neck in a constant position throughout the test and from one test to another. Perform the test at a reproducible velocity of stretch.

Determine the effect of spasticity on the child’s function, ease of care and quality of life by using various functional scales. This guides the treatment.

**Effects of spasticity**

**Adverse effects** Spasticity causes difficulty in movement, abnormal posture in sitting and standing, contractures leading to deformities, pressure sores and pain. Increase in tone is uncomfortable. Sitting is difficult for the nonambulatory child because of increased adductor and hamstring muscle tone. The child slides out of the wheelchair and cannot be positioned properly. He cannot transfer to and from the bed, wheelchair and bathtub. Perineal hygiene and dressing the child require more effort. The ambulatory child has trouble initiating movement. He cannot wear his braces. Energy cost of movement increases. Loss of function results and parents have difficulty caring for the child.

When muscle tone increases, muscles become tight. This inhibits normal gait and posture. Normal movement patterns do not develop. Instead, the child shows abnormal or compensatory movement patterns. Spasticity affects muscle growth. Muscles need to be stretched while relaxed; failure to do this results in poor growth. Spasticity initially causes apparent muscle shortening but the passive range of motion is full. This abnormal resistance is dynamic contracture. If uncorrected, fibrosis and eventually bony deformity lock the joint into a fixed contracture. How fast a contracture will develop depends on the severity of spasticity and the muscles involved: contractures progress more quickly in some muscles.

Bone growth is distorted by the abnormal resistance of the shortened muscles. Growing bone is easily distorted by sustained pressure. Untreated spasticity puts excessive stress on bone that produces abnormal rotation or it inhibits physiological derotation of long bones. If not relieved at an early stage, bone deformities occur. Prolonged equinovarus caused by triceps surae and tibialis posterior spasticity might rotate the tibia inwards. Spasticity of hip adductors can rotate the femur inwards. This inhibits the physiological derotation process of infantile femoral anteverision.

**Beneficial effects** Increased tone may be useful for the child. It helps maintain to keep the legs straight, thereby supporting the child’s weight against gravity. The child with increased tone in trunk extensors may stand and take a few steps. Spasticity may help preserve muscle bulk and bone density.

**References**


2002 Sheean G. ‘The pathophysiology of spasticity’ Eur J Neurol. 9 Suppl 1:3-9


Indications for treatment

Consider treating spasticity when it causes loss of function or produces contractures, deformities, pressure sores, or pain [A]. Additional indications include difficulty in positioning or caring for the total body involved child. Even though a wide range of treatments exist, none of them is fully satisfactory. Unwanted side effects limit the use of certain modalities. Some children do not respond to any of the antispasticity measures. The success of treatment depends on having specific goals in treatment, choosing the correct method according to the child’s problem and monitoring for side effects and complications.

Treatment methods

Treatment options are divided into reversible and permanent (surgical) procedures [B]. They can also be classified as systemic or local treatments. All treatment procedures aim to modulate the stretch reflex. In mild spasticity, basic measures such as positioning, exercises and bracing may be sufficient whereas in more severe cases, interventions can be more invasive. Often, treatments are combined to decrease side effects and to improve outcome.

Physiotherapy

Physiotherapy is a fundamental part of spasticity management. Muscle overactivity produces muscle shortening and muscle shortening increases spindle sensitivity. Muscle contracture and stretch sensitive muscle overactivity are intertwined. Therefore physical treatments aimed at lengthening the overactive muscles are fundamental. Address both shortening and overactivity. Consider applying various techniques such as positioning, ice, and exercises for these purposes.

Positioning

Position the child to stretch the spastic muscles and decrease the sensitivity of the stretch reflex and the brain stem reflexes that trigger spasticity [C]. The therapists should teach these positions to the family so that the child lies and sits this way most of the time at home. Head supports may improve tone in the trunk muscles by providing a sense of safety and inhibiting the tonic neck reflexes. Advise use of the tailor-sitting position to reduce adductor spasticity [D]. Good seating provides a stable platform and facilitates good upper extremity function.

Stretching exercises

Stretching muscles may prevent contractures and promote muscle growth. Spasticity decreases with slow and continuous stretching. This effect lasts from 30 minutes to 2 hours. Use stretching exercises before bracing and serial casting to obtain the necessary joint position.

Goals of spasticity treatment

<table>
<thead>
<tr>
<th>Increase function</th>
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<tbody>
<tr>
<td>to perform better in activities of daily living</td>
</tr>
<tr>
<td>to walk better</td>
</tr>
<tr>
<td>Increase sitting ability and balance</td>
</tr>
<tr>
<td>Prevent deformity &amp; decrease contractures</td>
</tr>
<tr>
<td>Pain relief</td>
</tr>
<tr>
<td>Improve hygiene and patient care</td>
</tr>
</tbody>
</table>

Treatment methods

- **Physiotherapy**
  - Positioning
  - Exercises
  - Stretching
  - Neurofacilitation
  - Electrostimulation

- **Splinting & Casting**

- **Oral medications**
  - Baclofen
  - Diazepam
  - Clonazepam
  - Dantrolene
  - Tizanidine

- **Intrathecal medications**
  - Baclofen
  - Morphine
  - Clonidine

- **Neuromuscular blocks**
  - Local anesthetics
  - Phenol
  - Botulinum toxin

- **Orthopedic surgery**

- **Selective dorsal rhizotomy**

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Have the child sit with legs in front, knees extended and ankles in neutral to stretch the hamstring and gastrocnemius muscles. This position is difficult to maintain for long periods.

Sitting in a cross legged position applies slow static stretch to the adductors and decreases spasticity.
Neurofacilitation techniques  Most neurofacilitation techniques are used to reduce muscle tone [A]. With the Bobath method, the therapist positions the child in reflex inhibitor positions and provides kinesthetic stimulation to inhibit the primitive reflexes and elicit advanced postural reactions to normalize muscle tone. With the Vojta method [B], different positions and proprioceptive stimulation are used for the same effect. Tone reduction lasts for a relatively short period of time with both methods.

Inhibitive (Tone Reducing) Casting and Bracing

Muscle relaxation after stretching exercises lasts for a short period of time. For longer duration the stretch on the muscle should be maintained for several hours every day. This is possible with the use of rigid splints or serial casting [C]. The effects are maximal if the cast or the splint is applied after the muscle is relaxed.

The tone-reducing effect of casts and splints is controversial. Some think that casts decrease muscle tone by creating atrophy in the already weak spastic muscle. Casts also cause pressure sores in children who are malnourished and have severe spasticity. Patient compliance may be poor because of difficulties of living with the cast.

Consider casting as an adjunct to treatment with local antispastic medications in the young diplegic or hemiplegic child with severe spasticity interfering with ambulation to delay orthopaedic surgery.

At present, the most common methods of spasticity management in cases of CP are oral medications, botulinum toxin, phenol or orthopaedic surgery [D].

References


<table>
<thead>
<tr>
<th>Treatment options in spasticity</th>
<th>Age</th>
<th>Patient group</th>
<th>Indication</th>
<th>Follow-up care</th>
<th>Result</th>
<th>Side-effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral medications</td>
<td>Any age 2-5 most common</td>
<td>Total body involved</td>
<td>Severe spasticity</td>
<td>Rehabilitation</td>
<td>Mild reduction</td>
<td>Sedation, weakness</td>
</tr>
<tr>
<td>Botulinum toxin A</td>
<td>Any age 2-10 most common</td>
<td>All spastic types</td>
<td>Focal spasticity too young for other interventions</td>
<td>Range of motion, stretching, strengthening exercises</td>
<td>Effective for 3-6 months</td>
<td>good results in walking and ADLs</td>
</tr>
<tr>
<td>Intrathecal baclofen</td>
<td>Above age 3 Abdomen large enough for pump insertion</td>
<td>Total body involved spastic or dystonic</td>
<td>Severe spasticity interfering with function or patient care</td>
<td>Range of motion exercises</td>
<td>Less need for orthopaedic surgery</td>
<td>easier care better sitting</td>
</tr>
<tr>
<td>Orthopaedic surgery</td>
<td>5-15 years</td>
<td>All spastic types</td>
<td>Contractures &amp; deformities</td>
<td>Strengthening</td>
<td>Better walking</td>
<td>Recurrence, weakness</td>
</tr>
<tr>
<td>Selective dorsal rhizotomy</td>
<td>3-7 years</td>
<td>Diplegic patient with pure spasticity</td>
<td>Spasticity interfering with walking</td>
<td>Intensive physiotherapy</td>
<td>Controversial</td>
<td>Increasing scoliosis, hip instability risk of incontinence</td>
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</tbody>
</table>
Oral Medications

Various pharmacological agents decrease spasticity. Baclofen, benzodiazepines (diazepam, clonazepam), dantrolene sodium and tizanidine are commonly used in children [A].

Indications

Consider systemic oral antispastic drugs in totally body involved nonambulatory children with generalized spasticity. They are also useful for short periods after orthopaedic surgery. Systemic side effects such as drowsiness, sedation, and generalised weakness are common, so they generally are not recommended for ambulatory children. Keep the initial dose low and gradually titrate to a level at which the effect is maximal and the side effects are minimal. The responses of the children to oral antispastic drugs are not consistent. Try different drugs to achieve a satisfactory clinical effect.

Oral antispastic drugs

Baclofen

Baclofen is an agonist of the main inhibitory CNS neurotransmitter gamma aminobutyric acid (GABA). It shows its effect mainly on the spinal cord. It decreases spasticity by increasing the inhibitory effect of the interneuron on the alpha motor neuron. The lipid solubility of baclofen is poor, so it cannot easily cross the blood brain barrier. High oral doses are necessary to achieve a therapeutic dose in the cerebrospinal fluid (CSF). The effect starts 1 hour after ingestion and lasts for 8 hours. The drug must be taken three to four times daily in divided doses. Daily dose for children between ages 2 to 7 is 10 to 15 mgrs per day with a maximum of 40 mgrs per day. After the age of 8 years, the dose may be increased to 60 mgrs per day. Maximum doses range between 80 to 120 mg. per day in adults. Side effects including sleepiness, sedation, fatigue, headache, nausea, and a decrease in seizure threshold are commonly associated with increasing doses. Baclofen also causes generalised muscle weakness. All side effects are dose dependent. Sudden withdrawal may cause hallucinations and seizures sometimes accompanied by extreme hyperthermia and increased spasticity called the baclofen withdrawal syndrome. The dose of the drug must be decreased gradually.

Diazepam

Diazepam is a benzodiazepine tranquillizer that works as a GABA agonist. It enhances the presynaptic inhibitory effect of GABA and decreases spasticity. It is absorbed faster than baclofen, acts faster, and has a longer lasting effect. Doses in children range between 0.12 to 0.8 mg/kg body weight with a maximum of 20 mg. daily divided into two or three equal doses. Diazepam decreases painful muscular spasms and improves sleep. Sedation and other CNS side effects are very common, so this drug is not recommended for treating ambulatory children except after orthopaedic surgery when it improves the child’s tolerance and participation in the rehabilitation program. CNS side effects are weakness, memory loss, ataxia, depression, and dependency.

Clonazepam

Clonazepam has an effect similar to that of diazepam, but it has a slightly longer half-life. It is preferred over diazepam because its side effects are fewer. Initial dose is 0.1 to 0.2 mg/kg/day. This dose is titrated for an optimal effect.

Dantrolene sodium

Dantrolene sodium inhibits muscle contraction by blocking calcium release from the sarcoplasmic reticulum in the muscle fiber. Initial dose is 0.5 mg/kg of body weight with a maximum dose of 3 mg/kg of body weight. Total daily dose should not exceed 12 mg per day administered in four divided doses. Side effects include muscle weakness, sedation, diarrhoea, and hepatotoxicity. CNS side effects are rare. Liver function tests should be performed two to four times a year, and the total treatment duration should not exceed 2 years.

Tizanidine

Tizanidine is an alpha adrenergic receptor agonist. It shows its effect at the brain and the spinal cord level. Tizanidine decreases the release of excitatory neurotransmitters and increases the release of inhibitory neurotransmitters. Guidelines for use in children are not well established. In adults the initial dose is 2 to 4 mg. administered at 4 hour intervals and increased to 36 mg. as needed. It may cause drowsiness, nausea, hallucinations, and is hepatotoxic.

References


A

<table>
<thead>
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<th>Oral antispastic agents in CP</th>
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<tbody>
<tr>
<td>Baclofen</td>
</tr>
<tr>
<td><strong>Mechanism</strong></td>
</tr>
<tr>
<td><strong>Dose</strong></td>
</tr>
<tr>
<td><strong>Duration</strong></td>
</tr>
<tr>
<td><strong>Side Effect</strong></td>
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</table>
Neuromuscular Blocking Agents

Local Anesthetics, Phenol, Botulinum Toxin

Consider using local anesthetics, alcohol, phenol and most recently, botulinum toxin as neuromuscular blocking agents [A] when treating focal spasticity.

Local anesthetics

Mechanism of effect
Local anesthetics block nerve conduction by changing membrane permeability to sodium ions. They affect both sensory and motor function in the area innervated by the nerve. This effect is completely reversible and causes no structural damage to the nerve. The effect starts within 3-15 minutes after the injection and lasts from 45 minutes to 8-12 hours depending on the type of drug used. Median nerve in the upper extremity and many nerves in the lower extremity are available for local anesthetic blocks [B].

Dosing and administration
Lidocaine, etidocaine and bupivacaine are used for nerve blocks. Prefer bupivacaine because it is more potent and its duration of action is longer. It can be injected in amounts up to 3 mg/kg of 0.25 to 0.75% of a solution. Do a perineural injection when you want to block the motor, sensory and autonomic fibers in the nerve. A motor point block affects the motor fibers only. A peripheral nerve stimulator that gives a low intensity electrical current through a needle electrode is used for blocks [C]. Use small needles and give short-lasting stimuli to localize the nerve more accurately. This makes the procedure less painful [D].

Local anesthetic blocks

<table>
<thead>
<tr>
<th>Block</th>
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<tr>
<td>Median</td>
</tr>
<tr>
<td>Tibial</td>
</tr>
<tr>
<td>Obturator</td>
</tr>
<tr>
<td>Femoral</td>
</tr>
<tr>
<td>Sciatic</td>
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</tbody>
</table>

Electrical stimulation technique

1. Locate the motor point or the nerve with the help of a stimulator. Charts exist for the location of each nerve.
2. Cleanse the skin. Choose the injection site and start stimulating the nerve. Adjust stimulation intensity first to a maximum, when the muscles innervated by the nerve begin to twitch, lower the intensity to 0.2-0.5 milliamperes.
3. If the muscle is still contracting, aspirate first and then inject the local anesthetic or phenol until the muscle is silent.
4. Increase the stimulus intensity to control the block. If there is no contraction at maximum stimulus intensity, the block is efficient. If not, inject more until the contraction stops.

Phenol denaturates the protein in the myelin and the axon. Injection into a mixed peripheral nerve causes a total nerve block for 2 - 12 months.

Botulinum toxin injected into the muscle inhibits acetylcholine release at the neuromuscular junction and causes a chemical denervation for 3 - 6 months.
Neuromuscular Blocking Agents

Indications for local anesthetic blocks

A

- Differentiate spasticity from contracture
- Predict functional changes
- Distinguish the muscles that contribute to spasticity
- Evaluate the presence of selective motor control

Advantages of local anesthetic blocks

B

- Reversible short duration effect
- Relatively painless
- Helps differentiate contracture from spasticity
- Unmasks activity in the antagonists by relaxing the spastic muscles.

Side effects and precautions

C

- Hypersensitivity reaction
- Hematoma at injection site
- Sudden weakness may cause injuries in the unprepared patient
- Systemic toxicity (dose related)

Indications

Local anesthetic blocks may be used as a diagnostic tool to differentiate spasticity from contracture and to predict functional changes with long term therapy [A]. The block may clarify which muscles contribute to spasticity and unmask selective motor control in the antagonist muscles if there is any. Block the median nerve at the elbow to evaluate the upper extremity. The hand relaxes completely a couple of minutes after the injection if the flexion in the wrist and fingers is because of spasticity. Bring the fingers into extension while holding the wrist in extension. The joint will not relax if there is a contracture. Thus, a local anesthetic block aids the physician in the decision making process of treatment of the spastic hand.

Advantages

Local anesthetics have a short and reversible effect, so they are useful for diagnosis of the problem and differentiating contracture from dynamic spasticity [B].

Chemical neurolysis: alcohol and phenol

Alcohol and phenol are chemical agents that block nerve conduction by creating a lesion in a portion of the nerve.

Alcohol

Ethyl alcohol acts as a local anesthetic by decreasing sodium and potassium conductance at the nerve membrane at low concentrations. It causes protein denaturation at higher concentrations such as 50%. Intramuscular injection of ethyl alcohol causes burning pain, therefore children must be injected under general anesthesia [D].

Even though alcohol has fewer adverse effects and is safer than phenol it has not been used as extensively in spasticity treatment possibly because of the pain it causes during the injection. Phenol blocks are generally used for lower extremity spasticity [E]. Recently botulinum toxin was added to the armamentarium of focal spasticity treatment [F].

Local Anesthetics

Phenol (6%)

Botulinum toxin A (Botox®)

<table>
<thead>
<tr>
<th>Mechanism</th>
<th>Onset</th>
<th>Duration</th>
<th>Dose</th>
<th>Precaution</th>
<th>Indication</th>
<th>Technique</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blocks sodium channels</td>
<td>Minutes</td>
<td>Hours</td>
<td>Bupivacain (0.25-0.75%) &lt;3mg/kg</td>
<td>Hypersensitivity</td>
<td>Differentiate spasticity from contracture</td>
<td>Stimulation - motor point</td>
</tr>
<tr>
<td>Denatures protein</td>
<td>Less than an hour</td>
<td>2-36 months</td>
<td>Less than 10 ml (1 gm)</td>
<td>Pain-dysesthesia</td>
<td>Proximal large muscles (no mixed nerve)</td>
<td>Stimulation - motor point</td>
</tr>
<tr>
<td>Inhibits acetylcholin release</td>
<td>Days</td>
<td>3-6 months</td>
<td>400 units at one single time</td>
<td>None</td>
<td>More for hygiene and comfort</td>
<td>In combination with BTX-A</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>All muscles accessible for injection</td>
<td>Active function</td>
<td>Combination with phenol</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Especially smaller muscles</td>
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<td>More for hygiene and comfort</td>
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<td>In combination with BTX-A</td>
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Phenol blocks for lower extremity spasticity

The rectus femoris motor point block

The hamstring motor point block

Adductor muscle motor point block

Tibial nerve block

The injection may be painful and is best performed under general anesthesia in young children.
Phenol

Mechanism of effect Phenol is benzyl-alcohol or carbolic acid with the old terminology. It has been used as a disinfectant and antiseptic. It causes protein denaturation and non-selective tissue destruction in the injected area. Wallerian degeneration of neurons occurs in the weeks following injection. Most axons regrow, over a period of time [A]. The effect of phenol starts rapidly because of its local anesthetic properties and lasts for up to 2 to 12 months.

Dosing and administration The usual dilution is 3 to 6% depending on the technique and the injection site. There are two techniques to apply phenol blocks: the motor point block and the motor nerve block. Motor point and motor nerve injection sites must be identified using electrical stimulation as explained in local anesthetic blocks. Electrically stimulating to find the motor points enables the physician to use very small quantities of the drug to obtain good clinical response [B].

Indications The advantages [C] include an early onset of action, longer duration of effect and low cost. In addition, there is no antibody formation to phenol so that larger, more powerful muscles may be treated without dosing considerations. Although the injection is painful at first, pain resolves in seconds because of its analgesic effects and injections are as easy as botulinum toxin injections for the experienced physician.

Side effects and precautions The main risks to be aware of when using phenol for spasticity management are permanent nerve injury, causalgia or neuropathic pain because of sensory fiber damage, tissue edema, venous thrombosis, and compartment syndrome resulting from large amounts of phenol in constrained space [B].

Avoid using phenol in the upper extremity because nerves in the upper limb are mainly mixed nerves and motor point blocks are difficult. Risks of dysesthesia, causalgia, venous thrombosis, and compartment syndromes are higher. Phenol is destructive to tissues, intramuscular administration in the small child may lead to unwanted and irreversible muscle fiber atrophy.

Combination treatment At present phenol has a rather small but useful place in spasticity treatment [D]. State-of-the-art treatment for focal spasticity relief is botulinum toxin. However, there is an upper limit to the amount of botulinum toxin that can be used in a single setting so a combination of phenol with botulinum toxin is preferred to better control multisegmental focal spasticity and to provide a longer duration of effect. Use phenol for large lower extremity muscles and botulinum toxin for smaller lower and all upper extremity muscles for multilevel injections whenever the necessary botulinum toxin dose exceeds the maximum amount you can use.

Botulinum toxin Botulinum toxin, produced by the anaerobic bacteria Clostridium botulinum, is one of the most potent poisons known to man. In the past two decades it has been transformed into one of the most useful antispastic agents. Of the seven distinct toxins from A to G, only type A and B are used for therapeutic purposes. The structure of all toxins and their mechanism of action are similar, only their site of action is different.
The mechanism of effect

The toxin inhibits acetylcholine release at the neuromuscular junction causing a reversible chemodenervation. Studies suggest that the toxin affects the muscle spindle and afferent nerve fibers as secondary actions.

Effect at the neuromuscular junction

The toxin must enter the nerve endings to exert its effect. It becomes fully active once inside the cholinergic nerve terminal.

When the impulse for contraction arrives at the axon terminal acetylcholine (Ach) vesicles fuse with the nerve membrane and the Ach is released into the synaptic cleft. This causes excitation in the muscle fiber and muscle contraction [A]. The various serotypes of botulinum toxin act on different portions of the acetylcholine vesicle complex. Botulinum toxin inhibits the fusion of acetylcholine vesicles at the pre-synaptic membrane. Ach cannot be released into the synaptic cleft, the impulse from the nerve to the muscle fiber is blocked and the muscle fibers innervated by that axon cannot contract. This is chemical denervation [B]. The extent of muscle weakness created by the botulinum toxin depends on the serotype, dose and volume of toxin used.

The effect of botulinum toxin is reversible. Nerve sprouts form at the unmyelinated terminal axon immediately proximal to the end plate. These sprouts innervate the muscle fiber [C]. Eventually, the original neuromuscular junction regains function [D]. This terminates the clinical effect in 3 - 6 months and spasticity reappears.

Afferent effect

The toxin may block the sensory afferents from the muscle spindle. This reduces spindle sensitivity and consequent reflex action.

Analgesic effect

There is an analgesic effect of the toxin explained by a couple of mechanisms. First, decreasing spasticity decreases pain. Second, botulinum toxin affects afferent transmission and inhibits the release of substance P. Substance P is the primary mediator of pain in the spinal cord and the brain. Inhibition of its release together with the block in afferent transmission result in pain relief.

Specific pharmacology

The potency of the toxin is defined by mouse units. One mouse unit is the amount required to kill 50% of a group of female Swiss-Webster mice. There are two different commercial preparations for botulinum toxin; Botox® (Allergan), and Dysport® (Spaywood) [B]. BTX-B is available as Myobloc™ in the United States and NeuroBloc® in Europe and elsewhere.

There are 100 units of botulinum toxin in one vial of Botox and 500 units in one vial of Dysport. The clinical potency of Botox and Dysport are influenced by numerous factors including the way they are produced. Therefore, the units are not interchangeable and there is no equivalence ratio between the two products [A on opposite page].

Indications

Botulinum toxin injections have been used as a safe and effective treatment for spastic CP for the past 10 years. Botulinum toxin B is also becoming commercially available.

The general indication for botulinum toxin injections in CP is ‘the presence of a dynamic contracture, interfering with function, in the absence of a fixed muscular contracture’. If botulinum toxin injections are started at an early age and repeated as necessary, they can help prevent the development of muscle contractures and bony deformities. This helps to delay orthopaedic surgery until the gait is mature. The need for extensive surgical procedures may be eliminated if bony deformities are prevented by botulinum toxin.
The success of botulinum toxin administration depends on many factors. Patient selection is critical [B]. Children with spasticity who do not have fixed contractures benefit a great deal from treatment whereas patients with dyskinesia have a variable response and athetoids do not benefit at all.

The timing of the injections is controversial. Most clinicians agree that the earlier the spasticity is reduced, the better the outcome. Botulinum toxin can be injected as early as 18 months of age. There is no upper age limit, however, once the muscle is shortened as occurs with age, the effect of spasticity relief will not be apparent because of contracture.

**Dosing and administration**

Botulinum toxin dosing depends on which preparation is used. Dysport dosing is different than Botox and there is no equivalence ratio between the two preparations in terms of clinical effect. The doses mentioned here refer to Botox injections [C,D]. The amount changes according to the number of muscles to be treated, prior response of the patient if there are any prior injections and functional goals.

The dose limits range from 2 units to 29 units/kg of body weight, most common range being between 10-20 units/kg of body weight. Avoid injecting more than 400 to 600 units of total dose at any one time, injecting more than 50 units at one injection site and exceeding 20 units per kilogram per muscle at any one time. If there is a need for more toxin because of multilevel involvement, combine treatment with phenol. Inject larger muscles with phenol and use botulinum toxin for more distal and smaller muscles [E].

Targeting the neuromuscular junction during the injection using electrical stimulation guide may result in more effect for less volume. Even though no serious complications have been reported, it is a good idea to apply high doses under general anesthesia in the operating theatre. Reduce the dose if the child is small and has atrophic muscles, if the treatment is going to be repeated for a number of times and if multiple muscles are being injected. Severely spastic and larger muscles should receive a larger dose whereas less spastic and small muscles receive a smaller dose [F].

The amount of toxin given to one muscle must be divided into more than two injection sites, depending on the dose. Put a safe distance between two injection sites with high doses. This increases the diffusion of the toxin in the muscle and prevents it from entering the systemic circulation. Divide the total dose per muscle over more sites as much as possible. For example, for a 20 kg child who has a very spastic gastrocnemius muscle, the dose should be 6 U/kg/muscle, 120 U total. This dose should be divided into 4 injection sites, 30 units per site in the muscle.

**Comparison of botulinum toxin A preparations**

<table>
<thead>
<tr>
<th>Preparation</th>
<th>Dysport</th>
<th>Botox</th>
</tr>
</thead>
<tbody>
<tr>
<td>Company</td>
<td>Ipsen</td>
<td>Allergan</td>
</tr>
<tr>
<td>1 ng toxsin-hemagglutinin</td>
<td>20 m.u.</td>
<td>&gt; 5 m.u.</td>
</tr>
<tr>
<td>Contents of one vial</td>
<td>500 m.u. (12.5 ng)</td>
<td>100 m.u.(40 ng)</td>
</tr>
</tbody>
</table>

The relative potency of these preparations has not been established yet.

**Specific goals for botulinum toxin A treatment**

To improve walking in the spastic diplegic and hemiplegic child
To minimise adductor tone in the child with early hip subluxation
To decrease the spasms and pain in the spastic-athetoid patients
To reduce tone in the psoas muscle in patients with back pain because of hyperlordosis
As a simulation for orthopedic surgery, to have a general idea of how the child will be when spasticity is reduced.

**General guidelines for upper extremity spasticity**

<table>
<thead>
<tr>
<th>Muscles injected</th>
<th>Dose range (units/kg of bw)</th>
<th>Number of sites per muscle</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biceps</td>
<td>2</td>
<td>2-3</td>
</tr>
<tr>
<td>Pronator teres</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Flexor carpi radialis</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Flexor carpi ulnaris</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Flexor digitorum superficialis</td>
<td>2</td>
<td>1-2</td>
</tr>
<tr>
<td>Flexor digitorum profundus</td>
<td>2</td>
<td>1-2</td>
</tr>
<tr>
<td>Flexor pollicis longus</td>
<td>0.5-1</td>
<td>1</td>
</tr>
<tr>
<td>Adductor pollicis</td>
<td>0.5-1</td>
<td>1</td>
</tr>
</tbody>
</table>

**General guidelines for lower extremity spasticity**

<table>
<thead>
<tr>
<th>Muscles injected</th>
<th>Dose range (units/kg of bw)</th>
<th>Number of sites per muscle</th>
</tr>
</thead>
<tbody>
<tr>
<td>Iliopsoas</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Quadriceps</td>
<td>3-6</td>
<td>4</td>
</tr>
<tr>
<td>Medial hamstrings</td>
<td>3-6</td>
<td>3-4</td>
</tr>
<tr>
<td>Lateral hamstrings</td>
<td>2-3</td>
<td>2</td>
</tr>
<tr>
<td>Adductors</td>
<td>3-6</td>
<td>2</td>
</tr>
<tr>
<td>Gastrocnemius</td>
<td>3-6</td>
<td>1-2</td>
</tr>
<tr>
<td>Soleus</td>
<td>2-3</td>
<td>1</td>
</tr>
<tr>
<td>Tibialis posterior</td>
<td>1-3</td>
<td>1</td>
</tr>
</tbody>
</table>

In general maximum of 50 U/site

**Botox® dose modifiers**

<table>
<thead>
<tr>
<th>Patient weight</th>
<th>Decrease dose if</th>
<th>Increase dose if</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low</td>
<td></td>
<td>High</td>
</tr>
<tr>
<td>High</td>
<td></td>
<td>Low</td>
</tr>
<tr>
<td>Duration of therapy</td>
<td>Chronic</td>
<td>Acute</td>
</tr>
<tr>
<td>Muscle bulk</td>
<td>Very small</td>
<td>Very large</td>
</tr>
<tr>
<td>Number of muscles injected simultaneously</td>
<td>Many</td>
<td>Few</td>
</tr>
<tr>
<td>Ashworth score</td>
<td>Low</td>
<td>Very high</td>
</tr>
<tr>
<td>Concern about weakness</td>
<td>High</td>
<td>Low</td>
</tr>
<tr>
<td>Results of previous therapy</td>
<td>Too much weakness</td>
<td>Inadequate response</td>
</tr>
</tbody>
</table>

Table reproduced with permission from WE MOVE New York www.mdvu.org.
General guidelines for spastic CP

<table>
<thead>
<tr>
<th>Type of CP</th>
<th>Muscles involved</th>
<th>Problem</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemiplegic</td>
<td>Rectus femoris</td>
<td>Stiff knee gait</td>
</tr>
<tr>
<td></td>
<td>Gastrocsoleus &amp; tibialis posterior</td>
<td>Pes equinovarus</td>
</tr>
<tr>
<td></td>
<td>Flexor – pronator spasticity</td>
<td>Thumb in palm deformity, flexion of the wrist and digits</td>
</tr>
<tr>
<td>Diplegic</td>
<td>Multilevel lower extremity injections</td>
<td>Adductor - flexor spasticity of the hip</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hamstring spasticity causing knee flexion</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Gastrocsoleus spasticity causing pes equinus</td>
</tr>
<tr>
<td>Quadriplegic</td>
<td>Hip adductors</td>
<td>Prevent hip subluxation</td>
</tr>
<tr>
<td></td>
<td>Hamstring spasticity</td>
<td>Sacral sitting</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sitting balance</td>
</tr>
</tbody>
</table>

**Patient Selection**

Botulinum toxin is useful in various upper and lower extremity problems in spastic cerebral palsy cases [A].

**Muscle selection**

Choosing the right muscles to inject depends on a good clinical evaluation [B]. Evaluate passive range of motion at the ankle, knee and hip; measure spasticity using the modified Ashworth or the Tardieu scale and determine strength and selective motor control of different muscle groups of the lower limbs. Gait analysis using dynamic EMG may be helpful in complex cases.

**Injection technique**

- **Needle size** depends on site of injection and physician preference. 1.0 ml tuberculin type syringes and 26-30 gauge, 1/2 inch (1.5 cm) needles are used. Teflon-coated monopolar injection needles are necessary for stimulation and injection with EMG or electrical stimulation guide [C].

- **Targeting** Botulinum toxin dosing and injection technique is relatively easy. For optimal results the physicians must be experienced in managing children with CP. Difficult-to-localize muscles often require adjunctive methods to confirm injection sites and to target the region of the neuromuscular junctions. Electromyography (EMG), electrical stimulation [D], computerized tomography (CT), fluoroscopy, and ultrasound have been used to target the region of maximum muscle activity. The technique of electrical stimulation is the same as in local anesthetic blocks. Efficacy is maximal and adverse effects minimal if the muscles are targeted properly.

- **Sedation** The injection is not painful, but may be a cause of distress in young children and in multilevel injections. It is rather difficult to inject certain muscles such as the hamstrings or iliopsoas in a fully awake and frightened child in the outpatient setting. Consider a simple sedative like diazepam or chloral hydrate when injecting single muscles in the outpatient clinic. Using EMG or ES guide and injecting multiple muscles is a considerable stress on the child so perform these under local anesthesia, conscious sedation using midazolam or general anesthesia.

- **Preparation** Keep the toxin frozen in vial. Dilute with normal saline to the desired concentration prior to usage [E]. The toxin is in a vacuumed vial, when diluting hold the piston of the syringe steady because sudden inflow of saline into the vial may cause protein denaturation and loss of pharmacological activity. Then put a second needle through the lid to balance the negative pressure inside the vial before drawing back the diluted toxin.

- **Injection** Clean the area, put sterile gloves on, localize the target muscle [A - L on opposite page], inject the desired amount into the muscle belly. You may need to inject at two or more sites depending on the dose and muscle size.

**Injection Dilutions**

For 100 units of Botox preparation

<table>
<thead>
<tr>
<th>Aimed final dilution</th>
<th>Saline added to vial</th>
</tr>
</thead>
<tbody>
<tr>
<td>2.5 U/0.1 ml</td>
<td>4 ml.</td>
</tr>
<tr>
<td>5.0 U/0.1 ml</td>
<td>2 ml.</td>
</tr>
<tr>
<td>10.0 U/0.1 ml</td>
<td>1 ml.</td>
</tr>
</tbody>
</table>
Adductor longus muscle: Patient lies supine. Abduct the leg to 15°. Palpate the tendon arising from the pubic tubercle and insert the needle 2-4 finger breadths distal to the tubercle into the muscle belly.

Adductor magnus muscle: Patient lies supine. Abduct and externally rotate the leg. Insert the needle midway between the medial femoral epicondyle and the pubic tubercle.

Rectus femoris: Patient lies supine. Insert the needle on the anterior aspect of the thigh, midway between the superior border of patella and the anterior superior iliac spine.

Medial hamstring muscles: Patient lies prone. Insert the needle at the midway on a line between the medial femoral epicondyle and the ischial tuberosity.

Lateral hamstring muscles: Patient lies prone. Insert the needle at the midway on a line between the fibula head and the ischial tuberosity.

Gastrocnemius, medial head: Patient lies prone, leg extended. Insert at the most prominent point of the medial muscle mass (approximately 3 fingers to one handbreadth below the popliteal crease.)

Gastrocnemius, lateral head: Patient lies prone, leg extended. Insert at the most prominent point of the lateral muscle mass (approximately 3 fingers to one handbreadth below the popliteal crease.)

Soleus: Patient lies prone, the leg is extended. Insert the needle deep just distal to the belly of the gastrocnemius muscle, medial and anterior to the Achilles tendon.

Tibialis posterior: Patient lies prone, with the leg in internal rotation. Draw a line from the popliteal crease to the medial malleolus. Inject one finger breadth off the medial edge of the tibia, directly obliquely through the soleus and the flexor digitorum longus, just posterior to the tibia.

Coronal view of commonly injected thigh muscles

Coronal view of commonly injected calf muscles

Ultrasonographic guidance can be helpful especially when injecting deep muscles.  

Courtesy D. Ganjwala
**Neuromuscular Blocking Agents**

**Resistance**

<table>
<thead>
<tr>
<th>Primary nonresponder</th>
<th>Secondary non responder</th>
</tr>
</thead>
<tbody>
<tr>
<td>No response to initial injection</td>
<td>Relative or complete loss of effect after second injection</td>
</tr>
<tr>
<td>Presence of antibodies</td>
<td>Low dose</td>
</tr>
<tr>
<td></td>
<td>Poor technique</td>
</tr>
<tr>
<td></td>
<td>Change in spasticity</td>
</tr>
<tr>
<td></td>
<td>Inappropriate reconstitution</td>
</tr>
<tr>
<td></td>
<td>Inappropriate storage</td>
</tr>
<tr>
<td></td>
<td>Antibody formation</td>
</tr>
</tbody>
</table>

**Contraindications & precautions**

- Aminoglycoside antibiotic use
- Pregnancy (for adult CP patients)
- Lactation (for adult CP patients)

**Post-injection treatment**

The antispastic effect appears within 24 hours to 3 days after injection and becomes maximum at 10 days to a month. It lasts for 3 to 6 months. Some patients are golden responders in whom the antispastic effect lasts for over a year. Proper exercises, splinting and casting may increase the number of golden responders.

**Casting** for 2 to 3 weeks after injections may improve the results. Botulinum toxin relieves dynamic spasticity whereas casting addresses fixed contracture. Consider casting for two weeks beginning on the third day after the injection in severe cases. If injecting under conscious sedation or general anesthesia, put the cast on when the child is sedated or asleep [A].

Problems related to casting are psychological trauma of putting the cast on and taking it off and muscle atrophy.

**Physical therapy** Perform range of motion and strengthening exercises in an intensive manner to obtain maximum benefits from the injection. Intensive exercises and electrical stimulation after the injection may increase toxin uptake by the nerve terminal and potentiate the effect.

**Orthotic management** Continue bracing as prior. Brace tolerance generally increases after the injection.

**Resistance**

A small percent of children may not respond to initial injection of botulinum toxin. Consider one or more treatments before classifying patient as a “non-responder”. A secondary non-responder is a child who shows a relative or complete loss of effect after a second injection. The reasons are too low a dose, poor injection technique, a change in the spastic muscles during treatment, inappropriate reconstitution or storage of toxin and the presence of neutralizing antibodies.

Development of resistance to botulinum toxin therapy is characterized by absence of any beneficial effect and by lack of muscle atrophy following the injection. Antitoxin antibodies are presumed responsible for most cases of resistance. Use the smallest possible effective dose and extend the time interval between treatments to at least 3 months to reduce the likelihood of antibody development. Botulinum toxin B or F may benefit those who have developed antibody resistance.

**Advantages and dysadvantages**

Side effects are few, mild and rare. The injection is relatively easy compared to phenol. There is no permanent tissue injury and all the effects are reversible. The cost is the only factor limiting toxin use [C].

**Contraindications**

Side effects are extremely few [D]. Slight weakness at injection site, local pain, fever, generalised weakness and fatigue presenting as a flu-like syndrome, respiratory tract infections, temporary incontinence and constipation have been reported with an incidence of 2-3%.

Contraindications include patients who are hypersensitive to any ingredient in botulinum toxin, who are using aminoglycoside antibiotics, pregnant or may become pregnant, or in lactation [E]. These contraindications are not absolute and not really relevant for children with CP. Patients who have a neuromuscular junction disease such as myasthenia like syndrome are not appropriate candidates for botulinum toxin therapy.
Conclusion
Botulinum toxin has an established place in the treatment of spasticity in cerebral palsy. Consider botulinum toxin treatment as early as two years of age and combine with other treatment options as the child grows older and spasticity begins to cause contractures and deformities [A]. The only factors limiting its use are high cost and restriction on the maximum dose per treatment session. The most common indications are young diplegic [B] and hemiplegic [C] children.

References
2004 Berweck S, Heinen F ‘Use of botulinum toxin in pediatric spasticity (cerebral palsy)’ Mov Disord. 19 Suppl 8:S162-7

Table modified from Allergan training module 4

<table>
<thead>
<tr>
<th>Muscles to be injected</th>
<th>Dose</th>
<th>Total dose per muscle</th>
<th>Number of injection sites</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right medial hamstring</td>
<td>4</td>
<td>72</td>
<td>2</td>
</tr>
<tr>
<td>Left medial hamstring</td>
<td>4</td>
<td>72</td>
<td>2</td>
</tr>
<tr>
<td>Right lateral hamstring</td>
<td>3</td>
<td>54</td>
<td>1</td>
</tr>
<tr>
<td>Left lateral hamstring</td>
<td>3</td>
<td>54</td>
<td>1</td>
</tr>
<tr>
<td>Right gastrocnemius</td>
<td>4</td>
<td>72</td>
<td>2</td>
</tr>
<tr>
<td>Left gastrocnemius</td>
<td>4</td>
<td>72</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>396 units</td>
<td></td>
<td>10</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Muscles to be injected</th>
<th>Dose</th>
<th>Total dose per muscle</th>
<th>Number of injection sites</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right quadriceps</td>
<td>4</td>
<td>60</td>
<td>4</td>
</tr>
<tr>
<td>Right gastrocnemius</td>
<td>5</td>
<td>75</td>
<td>2</td>
</tr>
<tr>
<td>Right tibialis posterior</td>
<td>2</td>
<td>30</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>165 units</td>
<td></td>
<td>7</td>
</tr>
</tbody>
</table>
Intrathecal Baclofen (ITB)

Baclofen is one of the most potent antispastic drugs. It cannot easily cross the blood brain barrier because of its poor lipid solubility. This makes it difficult to reach therapeutic doses in the CNS. A novel method of introducing the baclofen directly into the CSF through an implantable pump and catheter system has been devised in the past decade and has become increasingly popular. Intrathecal administration enables the drug to reach the receptor site quicker with a much lesser side effect profile.

**Indications for ITB**

ITB is useful for the severely involved spastic, dystonic or mixed child [A]. The aim is to enable sitting in the wheelchair, make transfers easier, decrease spinal deformity, increase the comfort level and ease of care through a decrease in spasticity. ITB pumps have been used in severe spastic diplegia, but more research is needed before one can definitely recommend this form of therapy for this particular problem.

**Factors to consider**

Consider several factors before the implantation [B]. Look for spasticity interfering with function and patient care. Define the type of involvement and the expected outcome after the intervention. Family cooperation is absolutely essential because complications of ITB pumps are potentially life threatening. The pump can be inserted in cases above the age of three, with an abdomen large enough for implantation. Check for hydrocephalus. It should be under control if present, otherwise it increases the chance of CSF leak. Get appropriate medical treatment for seizure activity because baclofen decreases the seizure threshold. Examine the skin of the back. It must be intact, there must be no pressure sores or active infection anywhere in the body. Financial resources must be sufficient because both the implantation and maintenance cost a substantial amount.

**Performing the test dose**

After the initial decision to implant a baclofen pump, perform a test to evaluate the effect of the drug when given intrathecally. Introduce 50 micrograms of baclofen into the intrathecal space by bolus injection through a lumbar punction in the spastic total body involved child. Implant the pump if the child responds to this dose. If the child does not respond, use 75 to 100 micrograms in the consecutive trials on the following days. The effect of intrathecal baclofen starts at 1-2 hours after the injection, reaches a maximum at 4-6 hours and gradually diminishes after 8 hours. Perform the test with an intrathecal catheter placed at the level of the 9th thoracic vertebra for the dystonic child. Give a continuous infusion of baclofen. Children who show a decrease of one or more in the Ashworth scale for a six to eight hour period are good candidates for pump implantation.

**Implanting the pump**

A minor surgical procedure is necessary for pump implantation [C]. Introduce the catheter into the intrathecal space at the distal thoracic or lumbar spine. Push the catheter tip to upper thoracic levels in cases of upper extremity spasticity and dystonia. The catheter is attached to an externally programmable pump implanted into the abdominal wall. The pump is filled transcutaneously every 2-3 months depending on the dosing schedule.
Intrathecal Baclofen Follow-up Dosing and clinical evaluation

Intrathecal administration of baclofen provides a continuous infusion of the desired amount of baclofen into the CSF. A computer based remote control system makes it possible to regulate the daily dose [A]. The antispastic effects of intrathecal baclofen are obtained at 1% of the daily oral dose.

Begin with an initial daily dose of 25 micrograms and titrate up until there is a satisfactory reduction in spasticity. The dose is usually between 100 to 500 micrograms per day. A static dose is generally achieved within a year after implantation. The pump should be refilled at 1-3 month periods. Refills are made through a transcutaneous injection. The battery life of the pump is approximately 4-5 years.

Begin an intensive physiotherapy program after pump implantation to reach functional goals [B,C]. Muscle weakness becomes prominent after a decrease in spasticity. Strengthening is important.

Complications

ITB pump implantation is expensive and the complication rate is moderately high. Complications include CNS infections, CSF leaks, and catheter related problems. Acute baclofen withdrawal syndrome [D] characterized by hallucinations, seizures, psychosis and rebound spasticity occurs if the baclofen flow to the CSF is interrupted. Signs of overdose are drowsiness, dizziness, somnolence, seizures, respiratory depression and loss of consciousness progressing to coma.

References


The intrathecal baclofen pump is remotely controlled by a computer. This enables the physician to increase or decrease the dose if necessary. Bolus injections may also be given.

![Image A](https://example.com/image1)

The child’s abdomen must be large enough for the pump. Sometimes the pump protrudes from under the skin and becomes vulnerable to trauma or infection.

![Image B](https://example.com/image2)

![Image C](https://example.com/image3)

Symptoms of acute baclofen withdrawal

- Acute increased tone
- Spasms
- Paresthesias
- Profuse sweating
- Dysphoria
- Hallucinations
- Seizures

![Image D](https://example.com/image4)
Selective Dorsal Rhizotomy

Selective Dorsal Rhizotomy and Other Neurosurgical Treatment Modalities

Selective dorsal rhizotomy (SDR) involves sectioning of the dorsal column rootlets to interrupt the spinal reflex arc [A]. This inhibits the afferent input from the muscle and tendons and reduces the efferent activity at the level of the spinal cord. The advantage of SDR is a global muscle tone reduction in lower extremities without producing weakness. All the lower extremity muscles are affected. The effects are permanent and weakness is not a major issue, however, there is loss of superficial and deep sensation.

Indications

Patient selection is important for success of the intervention. The ideal patient [B] is an independent ambulatory diplegic child between the ages of 3-10 with pure spasticity, no fixed contractures, good strength and balance with spasticity being the major limitation to function. Family commitment is essential for success because there is a need for long term intensive physiotherapy after the procedure. The extent of functional improvements cannot always be related to SDR itself because the patients also receive long and intensive hours of physiotherapy after the procedure for at least a year.

Technique

A laminectomy is done under general anesthesia and the posterior roots are exposed. EMG monitorization is recommended to determine which rootlets should be cut. The rootlets are stimulated electrically and the response from the muscles are observed. This way, the most active rootlets are localized. Up to 30-50% of the dorsal rootlets at each level from L2 to S1 are cut. In some centers, the L1 rootlets are also cut to assist in reduction of psoas activity. S2-S4 rootlets must be spared to preserve bladder function.

Follow-up

Expected results of the procedure are a loss of deep tendon reflexes, decrease in muscle tone, an improved gait pattern and smoothness of gait. Energy consumption may improve if walking is very inefficient prior to surgery. Sensory loss is usually transient though long term effects are not clear.

There is a need for extensive postoperative rehabilitation. After surgery, the therapy must focus on strengthening. Orthopaedic surgery is still necessary usually for foot instability (excessive valgus), rotational abnormalities and contractures. Continued gait improvements are minimal between 1 and 2 years after surgery.

Contraindications

SDR is contraindicated in patients who have extrapyramidal findings, significant weakness or contractures, spinal abnormality and poor family support and commitment.

Side effects & Precautions

There are concerns regarding the development of hip instability and spinal deformity after SDR. Proprioceptive sensory loss is common and the long term effects are unknown.

Other neurosurgical treatment modalities

Deep brain stimulation and magnetic repetitive stimulation have all been tried in the CP patient with limited success [C]. Certain neurosurgical procedures such as thalamotomy and stereotaxic surgery have not produced satisfactory results.

The ideal SDR candidate

- Diplegic child
- Age 3-10
- Independent ambulator
- Pure spasticity
- No fixed contractures
- Good strength and balance
- Reasonable selective motor control
- Family commitment

Neurosurgical procedures in spasticity

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Target</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stereotaxic encephalotomy</td>
<td>Globus pallidus</td>
<td>Variable-poor</td>
</tr>
<tr>
<td>Cerebellar stimulation</td>
<td>Cerebellum</td>
<td>Poor</td>
</tr>
<tr>
<td>Cervical rhizotomy</td>
<td>C1-C3</td>
<td>Variable-complications</td>
</tr>
<tr>
<td>Selective dorsal rhizotomy</td>
<td>L2-S2 selected rootlets</td>
<td>Variable-good</td>
</tr>
<tr>
<td>Neurectomy</td>
<td>Peripheral nerves</td>
<td>Variable, may cause chronic pain</td>
</tr>
</tbody>
</table>

References