Treatment of Cervical Dystonia

Dystonia Educational Series

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Treatment is aimed at reducing symptoms and as a result improving quality of life. The first goal is usually to improve the abnormal head position and jerky movements of the head and neck. Since pain occurs in the majority of patients with cervical dystonia and reduction of these abnormal movements and reduction of muscle spasm can easily substantially improve pain. Secondary complications may occur as a result of the abnormal head posturing and jerking movements. Contractures may occur, which means that there can be limitation in range of motion of the neck due to permanent shortening of the muscles in the neck. Accelerated arthritis may occur, resulting in compression of the nerve roots which exit the neck resulting in pain and weakness in the neck and arms; this is referred to as a cervical radiculopathy. Accelerated arthritis may also result in compression of the spinal cord, resulting in weakness and numbness in the arms and the legs as well as difficulties controlling the bladder and bowel; this is referred to as cervical myelopathy. Therefore, another important goal of treatment is to reduce the risk of developing the secondary complications.
Oral medications are usually used only in patients with more severe cervical dystonia who do not have an adequate response to treatment with botulinum toxin injections alone. Only a small minority, perhaps 20 or 25% of patients, demonstrate significant improvement with use of oral medications.
Botulinum toxin injections are the treatment of choice for patients with cervical dystonia. The toxin is injected into the overactive muscles to the overactive muscles to result in selective weakening of the muscles just along the normally active muscles to bring the head to a more normal position. The effect comes on gradually over several days with peak effect in approximately two weeks. The benefit lasts on average for about 12 weeks.
When the botulinum toxin is injected into the overactive muscle, it is taken up by the nerve ending which supplies the signal to the muscle to contract. Once inside the nerve ending, the toxin breaks down a protein which is necessary for the release of acetylcholine by the nerve ending. Acetylcholine acts as a signal from the nerve ending to the muscle to contract. The end result is that the toxin prevents the communication from the nerve ending to the overactive muscle resulting in temporary weakening of the overactive muscle reducing involuntary movement and muscle spasm. The effect is not permanent and the nerve ending eventually regenerates the protein which has been broken down by the botulinum toxin in the nerve ending that prevents release of the acetylcholine signal from the nerve ending to the muscle. When this important signaling protein is regenerated, the nerve ending can communicate with the muscle and the improvement of symptoms goes away.
There are two major forms of botulinum toxin, Type A and Type B. Type A botulinum toxins are used more commonly and the name brands are Botox, Dysport, and Xeomin. The name brand of Type B botulinum injection is Myobloc. Type A and Type B toxins work similarly, except that the type A toxins break down a different protein in nerve endings than does Type B toxin. The different forms of botulinum toxin have similar benefits and adverse effects. The effect of botulinum toxin injection is temporary and as a result patients need periodic injections. A very small percentage of patients, perhaps 1 or 2%, may develop antibodies to one type of botulinum toxin so that injections are no longer effective. If this occurs, patients may be switched from one type of toxin to another type of toxin.
The selection of the muscles and the dose that each muscle receives when botulinum toxin injections are performed must be customized based on the pattern and severity of abnormal movements as well as the size of the involved muscles, and the patient’s response to previous sets of injections. Patients need to undergo careful examination and thereafter the choice of what muscles to inject and the specific doses should be determined.
Injections are distributed amongst the various muscles, customized and based on the patient’s pattern of abnormal movements. EMG guidance is often used to help localize the overactive muscles. EMG stands for electromyography and is a neurophysiologic method by which the electrical activity of muscle activity can be recorded.
After injections are performed, the onset of effect occurring gradually over a few days with a peak effect in approximately two to four weeks. The average duration of improvement of symptoms is 12 weeks, though there is substantial variability from patient-to-patient. About 80% of patients experience significant improvement in abnormal neck posturing and pain.
The main effect of the botulinum toxin injection is to produce selective muscle weakness. Excessive weakness may occur, resulting in a floppiness of the head and more difficulty bringing the head up to neutral when the head is flexed or bringing the head forward when the head is extended such as when lifting the head off the pillow when in bed. Dry mouth can occur due to spread of the toxin to the salivary glands. Occasional patients may experience transient flu-like symptoms. Pain may occur at the injection site due to the mechanical effect of placing a needle in different muscles. Injection site pain typically lasts only one or two days and is usually improved with taking simple pain relievers such as Tylenol or ibuprofen. Excessive muscle weakness and dry mouth typically also improves over a few weeks.
Surgery is reserved for patients with severe dystonia with significant impairment in quality of life. Usually, this means that patients have an inadequate response to aggressive treatment with botulinum toxin injections and oral medications. Surgery may also be used in patients who initially responded well, but who have subsequently developed resistance to both botulinum toxin Type A and Type B. Most commonly, surgery is especially helpful in patients who have marked anterocollis (neck flexion) or retrocollis (neck extension). Since it may be difficult to improve patients with severe anterocollis adequately without causing substantial trouble swallowing. It may be difficult to adequately improve patients with severe retrocollis since this may result in excessive weakness of the neck extensor muscles. Cervical dystonia may also occur as part of more widespread segmental and generalized dystonia in which case botulinum toxin injections may not be able to be applied throughout the entire areas affected.
Local surgery which involves cutting specific overactive nerves and muscles causing abnormal movements is referred to as selective denervation. This surgery is usually applied in patients who have predominantly abnormal neck turning, also known as torticollis, or neck tilting, also called laterocollis. It is usually used in patients who initially had a good response to botulinum toxin injections but subsequently developed resistance. Since the efficacy of this surgery is similar to that seen with botulinum toxin injections. It is usually not very effective for patients with predominantly retrocollis, where the head is pulling back, or anterocollis where the head is pulling forward. Since nerves and muscles are cut, the effect is permanent. Typical complications include tingling and burning in the areas of the skin where the nerves have been cut, trouble swallowing, or excessive weakness of the neck. Many of these adverse effects gradually resolve after surgery over a period of weeks or months.
Deep brain stimulation refers to planning electrodes in specific areas of the brain and hooking up the electrode to a pacemaker-like device, which is usually implanted just below the collar bone. The idea behind this surgery is to jam the abnormal brain signals responsible for generating the dystonia. The electrodes are usually placed in the globus pallidus or less commonly the subthalamic nucleus. In patients with cervical dystonia, surgery is usually performed bilaterally with electrodes placed on both sides of the brain. After surgery, the electrical parameters of stimulation need to be set which is a process which is typically done gradually over several weeks and begins soon after surgery.
The benefit usually occurs gradually over hours to weeks. Up to 90% of patients who undergo this surgery see significant improvement with average improvement of about 50%. Deep brain stimulation is usually performed in patients who have dystonia involving not only the neck but also other areas of the body or in patients who have severe complex dystonia with retrocollis, anterocollis, or complex jerking movements.
Deep brain stimulation is appropriate for patients who have an inadequate response to aggressive treatment with oral medications and botulinum toxin injections, those who have complex or difficult-to-treat cervical dystonia with retrocollis, anterocollis, or complex jerking, or those with more widespread dystonia. Deep brain stimulation may adversely affect cognitive abilities and as a result, patients should have intact memory and thinking abilities prior to surgery in order to minimize the risk of significantly adversely affecting cognition.

Who should have DBS?

- patients who have inadequate response to oral medications and botulinum toxin
- patients who have complex cervical dystonia anterocollis and/or retrocollis
- patients with jerky movements
- patients who have more widespread dystonia
- memory and thinking abilities intact
On average deep brain stimulation improves symptoms about 50%. The abnormal head position and tremulous movements may be improved, pain is typically markedly improved, and use of other medications may be reduced. Some patients who continue to respond to botulinum toxin injections prior to surgery may continue to receive injections but with an altered dose and pattern in order to maximize the benefit of the combined approach to treatment with both deep brain stimulation plus injection therapy. As a result of these improvements, patients may lead a more normal life and may have improvement in mood and anxiety.
The most severe complication seen from deep brain stimulation is hemorrhage in the brain during the surgical procedure. This risk is approximately 1 to 2% for each side of the brain operated. The result is similar to that seen with stroke and may result in trouble with language, vision, paralysis, or rarely even death. The implanted hardware may break or may become infected in up to 5 to 10% of patients at some time during their lifetime. Most of these hardware complications or infections typically occur in the first two or three months after surgery, but rarely may occur months or years later. During the course of programming the stimulation parameters, numbness and tingling may occur in the body due to current spread to close-by fibers conveying sensory information in the brain. Similarly, slurred speech may occur due to occurring spread to the motor fibers to the face or throat.
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