Introduction to Deep Brain Stimulation and Who’s a Candidate

Dystonia Educational Series

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Deep brain stimulation refers to implanting electrodes into specific areas of the brain and hooking the electrodes up to pacemaker-like devices in order to send signals into the brain to jam the abnormal firing of brain cells, which are part of the network causing the abnormal movements. Deep brain stimulation is used in a variety of movement disorders including Parkinson’s disease, essential tremor, and Tourette’s syndrome and may be used in other neurologic disorders including chronic pain syndromes and psychiatric disorders such as refractory depression.
The electrodes are implanted in the target area, usually the globus pallidus or the subthalamic nucleus in patients with dystonia. The electrodes are fixed to the skull and are connected to the pulse generator via an extension cable. The IPG or implantable pulse generator is usually placed below the collar bone. Stimulation may be performed on one side of the brain or on both sides of the brain, depending on the nature and distribution of the patient’s dystonia.
The pulse generator sends an electrical current to the electrodes that are implanted in the brain. The exact stimulation parameters can be programmed postoperatively in order to maximize benefit and minimize adverse effects. The electrical current is thought to jam the abnormal pattern of firing of the nerve cells sounding the electrode and in this way reduces the abnormal signal sent to cause the contraction of the affected muscles and the abnormal movements.
The most common site for deep brain stimulation for dystonia is the globus pallidus internus.
More recently, the subthalamic nucleus has been used as a surgical target for deep brain stimulation for dystonia. It is still not fully known which patients may respond better to deep brain stimulation in the subthalamic nucleus or globus pallidus. The thalamus may also be used as a site for deep brain stimulation for dystonia, predominantly in patients who have substantial associated tremor or myoclonus.
Deep brain stimulation is appropriate for those who have substantial disability and impairment in quality of life despite maximal treatment with nontypical treatments such as botulinum toxin injections and oral medication. Disability may be due to abnormal movements with or without significant accompanying pain.
Patients who have primary dystonia due to a genetic mutation or have idiopathic dystonia without any associated brain lesion on MRI scanning usually have a better outcome than those who have secondary dystonia. Patients who have secondary dystonia due to an acquired cause with a brain scan which demonstrates a lesion in the brain as the cause of dystonia, usually have a poorer outcome.

Therefore, in general, patients who have idiopathic focal, cervical, or generalized dystonia with a gene mutation tend to have a good result from deep brain stimulation. Patients with the mutation in the DYT1 gene almost uniformly experience excellent outcomes from deep brain stimulation.

Patients with secondary dystonia due to a variety of causes including head injury, stroke, or metabolic degenerative causes of dystonia tend to fare much more poorly.

Amongst acquired causes of dystonia, only patients who have tardive dystonia due to exposure to dopamine receptor blocking drugs tend to uniformly respond well.
There is some evidence to suggest that younger patients tend to respond better than older patients with deep brain stimulation. However, this does not mean that deep brain stimulation may not be effectively applied in patients who are middle aged or older. There is also significant data now suggesting that the shorter the duration of the symptoms prior to deep brain stimulation the more effective the ultimate outcome is. As a result, deep brain stimulation should not be delayed in patients who have demonstrated substantial impairment despite maximal treatment with more conservative therapies.

It is important to operate before patients develop thick skeletal deformities such as scoliosis, since even after the dystonic contractions have been improved, such skeletal deformities may not resolve. This is especially important in individuals with childhood or adolescent-onset dystonia.
DYSTONIA EDUCATION SERIES

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