PRACTICE GUIDELINE FOR DEEP BRAIN STIMULATION (DBS) FOR DYSTONIA

Policy:

ADHS/CRSA considers unilateral or bilateral deep brain stimulators as medically necessary durable medical equipment for the treatment of patients seven years of age or above with medically intractable/refractory primary dystonia, including generalized dystonia, hemidystonia, and cervical dystonia.

At this time, the U.S. Food and Drug Administration Humanitarian Device Exemption H020007 applies only to the treatment of medically intractable/refractory primary dystonia (USFDA, 2003). Therefore, the deep brain stimulator is not a CRS covered benefit for other diagnoses, and ADHS/CRS considers unilateral or bilateral deep brain stimulators for all other diagnoses experimental and investigational. These include, but are not limited to, trauma, hypoxic-ischemic encephalopathy, multiple sclerosis, degenerative disorders, metabolic disorders, Infectious diseases, drug-induced, epilepsy, chronic cluster headaches, Tourette’s syndrome, and secondary dystonia.

CRSA cannot authorize or provide payment for this procedure for any condition other than the treatment of patients who are seven years of age or above, with medically intractable/refractory primary dystonia.

Under the U.S. Food and Drug Administration Humanitarian Device Exemption provision, deep brain stimulators can only be placed at a clinical facility with Internal Review Board (IRB) approval for the use of the device for the FDA approved indication (USFDA, 2003).

Description of Product or Service:

Deep brain stimulation (DBS) is a neurosurgical procedure that involves the stereotactic implantation of an electrode into a specific region of the brain, delivering chronic, high frequency electrical stimulation/impulses to several deep brain nuclei. Deep brain stimulation therapy uses a device with three implantable components: a quadripolar brain lead, a computerized neurostimulator, and an extension wire that connects the two.

Generally, DBS lead implantation is done using local anesthesia while the patient is awake to optimize the mapping procedure and avoid stimulation-induced adverse effects during intraoperative test stimulation of the lead. For younger patients or those with extreme dystonia, this procedure can be done with general anesthesia. Leads can be implanted unilaterally or bilaterally depending on the distribution of the patient's symptoms.

Often, the electrode is attached to a temporary cable on the skin for short-term stimulation to validate treatment effectiveness. After effectiveness is determined, a permanent electrode is placed and the neurostimulator is implanted subcutaneously with the patient under general anesthesia. The neurostimulator is typically implanted in the subclavicular region, but can be located elsewhere.

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Days to weeks after device implantation, stimulation is activated using the DBS programmer, and the frequency and amplitude of the electrical pulses can be adjusted according to the patient’s symptoms. Setting the neurostimulation parameters involves a balance between optimal efficacy and avoidance of side effects of neurostimulation.

DBS is an adaptable procedure that provides an alternative to more permanent surgical procedures that are ablative or lesioning, such as thalamotomy and pallidotomy. Ideally, DBS should interrupt the pathways responsible for the abnormal movements, functioning to reduce the overactive brain regions without destroying them.

**Background:**

In August 1997, the U.S. Food and Drug Administration (USFDA) approved DBS for medically refractory essential tremor and medically refractory Parkinson’s Disease. Although these two diagnoses are not typically seen in the pediatric population, this information is included in this document to help the reader understand the different levels of FDA approval of DBS.

Effective October 1996, USFDA regulations regarding Humanitarian Use Devices (HUD’s) became effective as a result of provisions of the Safe Medical Devices Act of 1990. A HUD is defined by the FDA as “a device that is intended to benefit patients by treating or diagnosing a disease or condition that affects fewer than 4,000 individuals in the United States per year.” This regulation provides an incentive for the development of devices for use in these small patient populations.

For the USFDA, the HUD regulation provides for the submission of a Humanitarian Device Exemption (HDE) application, which is similar to a premarket approval (PMA) application, but is exempt from the effectiveness requirements of a PMA. An HDE application is not required to contain the results of scientifically valid clinical investigations demonstrating that the device is effective for its intended purpose, but must contain “sufficient information for FDA to determine that the device does not pose an unreasonable or significant risk of illness or injury,” and that the probable benefit to health outweighs the risk of injury or illness from its use.

In April 2003, the FDA granted approval for the Activa® Dystonia Therapy System (Medtronic, Minneapolis, MN) under the HDE process, for medically refractory intractable primary dystonia. This HDE approval authorizes marketing of the Activa® Dystonia Therapy System. However, according to USFDA guidelines, “an HDE may only be used after Internal Review Board (IRB) approval has been obtained for the use of the device for the FDA approved indication.” Furthermore, the labeling for an HUD must state that the device is a humanitarian device and the “effectiveness of the device for the specific indication has not been demonstrated.”

**Dystonia:**

The term “dystonia” covers a diverse group of movement disorders, all of which are characterized by involuntary muscle contractions that may cause twisting and repetitive movements or abnormal postures. Dystonia can be classified by its distribution on clinical examination or its known or inferred etiology. Regarding the former, focal dystonia affects a single body part, segmental dystonia involves adjacent body parts, hemidystonia involves one side of the body, and generalized dystonia is more widespread, affecting two or more body
segments. Regarding etiologic classifications two broad categories occur, primary and secondary dystonia. Marks (2005) defines these classifications as follows:

- **Primary Dystonia**: (idiopathic dystonia) is diagnosed when dystonia is the only sign present and there is no identifiable exogenous cause, degenerative disorder, or inherited condition underlying the dystonia. Several genetic mutations have been linked to some of the primary dystonia syndromes such as DYT1.

- **Secondary Dystonia**: is associated with acquired or exogenous causes and frequently has other neurologic findings in addition to dystonia. Acquired or exogenous causes may include: perinatal-hypoxic-ischemic insult, stroke, encephalitis, neurodegenerative disorders, or traumatic brain injury. Hereditary neurologic syndromes may also cause secondary dystonia, and can include Huntington’s disease, Wilson’s disease, pantothenate kinase-associated neurodegeneration, and many others.

Those with dystonia usually have normal intelligence and no associated psychiatric disorders. Dystonia is the most severe form of a group of movement disorders called dyskinesias. At this time, the FDA approved indication for the use of DBS in the treatment of dystonia is for medically refractory, intractable primary dystonia in patients seven years of age or older. In addition, the clinical facility implanting DBS for primary dystonia under the HDE provision must have IRB approval for this FDA HDE indication.

**Treatment of Dystonia**

The primary goal of all treatments for dystonia is to provide symptomatic relief, improve function, and reduce disability. Currently, there is no cure for dystonia. Treatment options include medical treatment and surgical procedures, with surgery reserved for medically intractable dystonia.

Medical treatment includes oral medications such as dopaminergic and anticholinergic medications, and Baclofen. Chemodenervation is the use of chemicals such as botulinum toxin injection therapy to interrupt the flow of nerve impulses to the abnormal muscle. Botulinum toxin injection for the treatment of focal dystonia has been used for over a decade, and is considered the current standard of care for focal dystonia.

Surgical treatment for dystonia includes ablative/lesioning procedures, cervical rhizotomy, and now functional brain surgery with DBS for medically intractable primary dystonia.

**Patient Selection:**

- The patient must have medically intractable/refractory primary dystonia, with adequate documentation of previous medical failure.
- The patient must be an appropriate candidate for stereotactic procedures.
- The patient must be physically able to endure the surgical procedure.
- The patient must be able to cooperate by answering questions and following directions during the surgery.
- The patient must understand the nature of therapy and be able to operate the neurostimulator control magnet or therapy controller.
- The patient must be available for periodic follow-up visits, and must have demonstrated exceptional compliance with previous medical therapies.

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The patient should not be suffering from advanced dementia or have other independent diagnoses that could explain the failure to respond to medical treatment. There should not be any focal lesion of the basal ganglia, such as a space-occupying lesion or lacunae, at the target site that would nullify the result of the DBS.

The patient will be fully aware of the risks and benefits of the surgery, including mortality and morbidity experience of the center and the performing surgeon.

The patient must not have a medical condition that requires repeated magnetic resonance imaging (MRI).

Various medical or environmental devices may generate enough electromagnetic interference to change the parameters of a neurostimulator; to turn it on or off, or it may cause a surge, shock, or jolt.

Patients treated with DBS systems must never receive Diathermy.

The use of cerebellar stimulation or pacing is considered investigational or not medically necessary.

There will be no coverage for patients who have had previous thalamotomy, or when simultaneous DBS of the Ventralis Intermediate Nucleus of the Thalamus (VIM) is planned.

The patient must not have other operating pacemakers.

Patients will be advised to avoid games, sports, and other pastimes where a strain to the lead connector assembly or a percussive injury to system components may be likely to occur (soccer, football, rugby, etc).

References and Recommended Reading:


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U.S. Food and Drug Administration (FDA) and Center for Devices and Radiological Health (CDRH). (2003). Letter to Mr. Kenneth Jensen, Principal Regulatory Affairs Specialist at Medtronic Neurological from Daniel Schultz MD., Director, Office of Device Evaluation at CDRH. Rockville, MD.


Clinical Policy Bulletins:

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