GUIDELINE STATEMENT

This guideline outlines the management of patients are being considered for a Deep Brain Stimulator as required by the Children’s Rehabilitative Services Program (CRS), Arizona Health Care Cost Containment System, State of Arizona.

PURPOSE

Clinical Practice Guidelines represent the minimum requirements for providing care for individuals with CRS conditions that might respond to a Deep Brain Stimulator and treatment which should be provided in a manner that includes adherence to and consistency with the following Guideline.

DEFINITIONS:

Children’s Rehabilitative Services (CRS): An AHCCCS program for children with certain diagnoses which provides services using an integrated family-centered, culturally competent, multi-specialty, interdisciplinary approach.

Deep Brain Stimulator: is a device which delivers an electrical pulses to the brain via electrodes surgically implanted in the internal globus pallidus interna (GPI), subthalamic nucleus (STN) or ventral intermediate nucleus (VIM) of the thalamus. The mechanism of action is not completely understood, but the goal of DBS is to interrupt the pathways responsible for the abnormal movements associated with movement disorders.
1. PROCEDURAL GUIDELINES for POLICY COMPLIANCE

A. CRS Enrollment:
Children diagnosed with dystonia must be seen at a CRS Multispecialty Interdisciplinary Clinics (MSIC) site with a Neurology or Neurosurgical Specialty Clinic with additionally a Pediatric Comprehensive Assessment.

B. Interdisciplinary Team Membership:

<table>
<thead>
<tr>
<th>Clinics</th>
<th>Interdisciplinary Team Members</th>
<th>Interdisciplinary Team Members Available During Specialty Clinics As Needed</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neurology</td>
<td>Pediatric Neurologist Registered Nurse, Educator, Social Worker, Translator, Advocate</td>
<td></td>
</tr>
<tr>
<td>Neurosurgery</td>
<td>Neurosurgeon Registered Nurse, Educator, Social Worker, Child Life Specialist, Translator, Advocate</td>
<td></td>
</tr>
<tr>
<td>Pediatrics</td>
<td>Pediatrician LPN or MA (Medical Assistant) Pediatrics, Child Life Specialist, Educator, Social Worker, Translator, Audiologist, Registered Nurse, Advocate</td>
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</tbody>
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E. 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 lesion procedures, cervical rhizotomy, and now functional brain surgery with DBS for medically intractable primary dystonia.
2. GUIDELINES FOR MANAGEMENT AND TREATMENT OF HEARING LOSS WITH IMPLANTABLE HEARING AIDS INCLUDING BAH A AND COCHLEAR IMPLANTS

The purpose of this guideline is to promote a uniform level of care at CRS MSIC sites for members with dystonia and to provide a general framework for treatment of dystonia. The relevance to specific situations will depend on individual variations in clinical course and professional judgment. In addition, this document should serve as a tool to assess programs, secure resources needed to enhance patient care and education, and guide the future development and treatment of Hearing Loss.

A. Goals:

1. To maintain maximum individual functioning for members who would be candidates for deep brain stimulator implants.
2. To improve quality of life for members with dystonia

B. Objectives:

1. To maintain a network with specialists able to anticipate and dystonia related physical and psychosocial problems for effective management and outcomes. This implies easy accessibility to all team members experienced with the comorbidities and educational needs of members with dystonia. Specialists must be able to provide accurate and timely diagnosis as well as be knowledgeable of appropriate treatments for this condition. This will allow appropriate community/social integration including transition to adulthood. Nurses at the MSIC must also be experienced or have additional training to be competent in supporting the member’s treatment and educational needs.

2. Services will be provided with multispecialty, interdisciplinary clinics in a member centric, family friendly, culturally sensitive manner.

F. Clinical Indications for Deep Brain Stimulation Implant for Dystonia:

1. Deep brain stimulation is proven for treating the following:
   a. Primary dystonia* (occurs apart from any other identifiable illness) including generalized and/or segmental dystonia, hemidystonia and cervical dystonia (torticollis) when used according to the U.S. Food and Drug Administration (FDA) indications

2. Patient Selection:

   a. The patient must have medically intractable/refractory primary dystonia, with adequate documentation of previous medical failure.

   b. The patient must be an appropriate candidate for stereotactic procedures.

   c. The patient must be physically able to endure the surgical procedure.

   d. The patient must be able to cooperate by answering questions and following directions during the surgery.

   e. The patient must understand the nature of therapy and be able to operate the neuro-stimulator control magnet or therapy controller.

   f. The patient must be available for periodic follow-up visits, and must have demonstrated exceptional compliance with previous medical therapies.
g. 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.

h. 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.

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

j. 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.

k. Patients treated with DBS systems must never receive Diathermy.

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

m. 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.

n. The patient must not have other operating pacemakers.

Bibliography:


