Movement disorders and Deep Brain Stimulation: A treatment option

Lesley Johnson, MS, RN, FNP
University of Rochester Medical Center
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Movement Disorder
A neurological condition that affects speed, coordination, fluency, and ease of movement.
Symptoms vary among type of disorder.
The disorder may be progressive.
Severity of symptoms may fluctuate.

Movement Disorders
Abnormal movements may be rhythmical as in Essential tremor.
They may be irregular, rapid, and jerky as in Tics.
Slow and sustained as in Parkinson’s disease.
Combination of sustained muscle contractions that produce twisting and repetitive movements as in Dystonia.
Movement Disorders

Some disorders may cause

**Hyperkinesia** - Abnormal involuntary movement or excessive spontaneous movement.

**Hypokinesia** – Partial or complete loss of muscle movement.

**Dyskinesia**: Abnormal speed or fluency

**Ataxic**: Uncoordinated movements that appear as jerking motions

In most cases irregular movements cannot be consciously controlled or suppressed.

Causes and Risk Factors

Movement disorders occur as a result of damage or disease in a region located deep in the brain (basal ganglia).

Movement disorders can result from the following:

- Age-related changes
- Environmental toxins
- Genetic disorders (e.g., Huntington’s disease, Wilson disease)
- Medications (e.g., antipsychotic drugs)
- Metabolic disorders (e.g., hyperthyroidism)
- Stroke

Basal ganglia

A region of the base of the brain that consists of three clusters of neurons (caudate nucleus, putamen, and globus pallidus) that modulate motor control.

Comprised of clusters of nerve cells (neurons) that send and receive electrical signals and are responsible for involuntary movement.

Responsible for executing a **skill pattern** that you can do with very little thought.

Communicates with other regions of the brain to allow us to execute smooth and patterned movements-Motor and Premotor cortex.
Diagnosis

Family history
History of symptoms
Neurological examination
Laboratory tests may include CBC, CK, CSF, DNA analysis.
Imaging studies to detect structural abnormalities, damage in the basal ganglia, or stroke. May include CT scan and/or MRI.
EEG, EMG, muscle biopsies
Diagnosis may also be a process of exclusion sometimes utilizing medication trials as in PD.
Control of movement disorders depends on the underlying cause.
Goal of treatment is to relieve symptoms.
Medication side effects and doses may cause intolerable side effects.
Anti-seizure medications may cause a lack of coordination and balance (ataxia), dizziness, nausea, and fatigue.
Benzodiazepines may cause ataxia, drowsiness, and fatigue.
Beta-blockers include slowed heart rate (bradycardia), depression, light-headedness, and nausea.
Dopamine agonists may cause nausea, headache, dizziness, and fatigue.

Essential Tremor
- Nerve disorder characterized by uncontrollable shaking, or "tremors"
- Areas affected often include the hands, arms, head, larynx (voice box), tongue, and chin. The lower body is rarely affected.
- Cause is not clear
- Genetics is responsible in half of all people with the condition. A child born to a parent with ET will have up to a 50% chance of inheriting the responsible gene
- Most common movement disorder, affecting up to 10 million people in the U.S.
- Diagnosis based on reported symptoms, FH and a complete neurological exam.
Essential Tremor treatment options

**Medications**: Oral drugs can significantly reduce the severity of essential tremor. Medications include Propranolol, Primidone, Topiramate, and Gabapentin.
- Benzodiazepines such as Clonazepam, Diazepam, Alprazolam, and Lorazepam.
- Botox injections may also be a treatment option.
  - Effective for vocal and head tremors.

**Surgery**: Deep Brain Stimulation (DBS) is a surgical treatment option for people with severe symptoms despite medical therapy.

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Dystonia

Dystonia is a disorder characterized by involuntary muscle contractions
- Slow repetitive movements or abnormal postures.
- The movements may be painful, and some individuals with dystonia may have a tremor or other neurologic features.

Dystonia can affect many different parts of the body

Cause of dystonia is not known.

Early symptoms may be very mild and may be noticeable only after prolonged exertion, stress, or fatigue.
- Eye spasms, neck pulling, hand or foot cramping.
Dystonia-Idiopathic, Genetic, and Acquired

**Primary Idiopathic dystonia**
- Genetic association
- Insidious onset: May appear initially during performance of a specific, repetitive, overlearned action

**Secondary Acquired dystonia** results from environmental or injury to the brain, or from exposure to certain types of medications.
- Infections, reactions to drugs, hypoxia/ischemia from trauma or stroke.
- Acute or delayed onset
- May plateau and not spread to other parts of the body.
- Dystonia as a result of medications often ceases if the medications are stopped

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Dystonia - Classifications

Based upon the regions of the body which they affect:

- **Generalized dystonia** affects most or all of the body.
- **Focal dystonia** is localized to a specific part of the body.
- **Multifocal dystonia** involves two or more unrelated body parts.
- **Segmental dystonia** affects two or more adjacent parts of the body.
- **Hemidystonia** involves the arm and leg on the same side of the body.

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Dystonia - Symptom management

**Botulinum Toxin (Botox) Injections**
- Toxin decreases muscle spasms

**Medications**
- Anticholinergic agents block the muscle contraction effects of the neurotransmitter acetylcholine.
  - Drugs in this group include trihexyphenidyl (Artane) and benztropine (Cogentin)
- GABAergic agents regulate the muscle tone activity the neurotransmitter GABA directly affects.
  - These include Benzodiazepines
- Dopaminergic agents act on the dopamine system that includes regulation of involuntary muscle movements.
  - Tetrabenazine (Xenasince)- presynaptic depleter of Dopamine

**Surgery**
Parkinson’s disease

Parkinson’s disease (PD) is a neurodegenerative brain disorder that progresses slowly in most people.

First described in 1817 by James Parkinson

Slow progressive decrease in the neurotransmitter dopamine leads to decrease in the ability to regulate movements and emotions.

Centers for Disease Control and Prevention (CDC) rated complications from PD as the 14th top cause of death in the United States.

Hoehn and Yahr staging. This scale, first introduced in 1967, is a simple rating tool to generally describe how motor symptoms progress in Parkinson’s.

Unified Parkinson’s Disease Rating Scale (UPDRS). It takes into account factors other than motor symptoms, including mental functioning, mood and social interaction.

Parkinson’s disease

Cause of Parkinson’s disease is unknown. No known cure

Maybe a combination of genetic and environmental factors

Insecticides: Permethrin and Beta-hexachlorocyclohexane (beta-HCH)

Herbicides: Paraquat and 2,4-dichlorophenoxyacetic acid

Fungicide: Maneb

Agent Orange used in the Vietnam War

MPTP, a synthetic neurotoxin (can cause Parkinsonism)

Head injuries

Parkinson’s disease symptoms

Four primary symptoms

• Tremor
• Rigidity
• Bradykinesia
• Postural instability

Typically symptoms occur > 60 yrs. 1% of the general population

Other symptoms may include depression and other emotional changes; difficulty in swallowing, chewing, and speaking; urinary problems or constipation; skin problems; and sleep disruptions.

Diagnosis is based on medical history, neurological examination, medication trials
Parkinson’s disease treatments

Medications may provide dramatic relief from the symptoms. Levodopa combined with carbidopa.

Carbidopa delays the conversion of levodopa into Dopamine. Nerve cells can use levodopa to make dopamine and replenish the brain’s dwindling supply.

Bradykinesia and rigidity respond best.
Tremor may be only marginally reduced.
Balance and other symptoms may not be alleviated at all.

Anticholinergics may help control tremor and rigidity.

Dopamine agonists such as bromocriptine, pramipexole, and ropinirole, mimic the role of dopamine in the brain.

Antiviral drug, amantadine, also appears to reduce symptoms by potentiating dopamine responses.

Rasagiline to be used along with levodopa for patients with advanced PD or as a single-drug treatment for early PD. Increases extracellular Dopamine.

Surgery

Hoehn and Yahr staging scale for PD

<table>
<thead>
<tr>
<th>Stage</th>
<th>Hoehn and Yahr Scale</th>
<th>Modified Hoehn and Yahr Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Unilateral involvement only</td>
<td>Unilateral involvement only</td>
</tr>
<tr>
<td>1.5</td>
<td>Unilateral and axial involvement</td>
<td>Unilateral and axial involvement</td>
</tr>
<tr>
<td>2</td>
<td>Bilateral involvement; without impairment of balance</td>
<td>Bilateral involvement; without impairment of balance</td>
</tr>
<tr>
<td>2.5</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>Mildly disabling</td>
<td>Mildly disabling</td>
</tr>
<tr>
<td>4</td>
<td>Severe disability; unable to walk or stand</td>
<td>Severe disability; unable to walk or stand</td>
</tr>
<tr>
<td>5</td>
<td>Confinement to bed or wheelchair or death</td>
<td>Confinement to bed or wheelchair or death</td>
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Unified Parkinson’s disease rating scale (UPDRS)

Used to follow the longitudinal course of PD
The UPDRS is made up of these sections:
Part I: evaluation of mentation, behavior, and mood
Part II: self-evaluation of the activities of daily life (ADLs) including speech, swallowing, handwriting, dressing, hygiene, falling, salivating, turning in bed, walking, and cutting food
Part III: clinician-scored monitored motor evaluation
Part IV: complications of therapy
Part V: Hoehn and Yahr staging of severity of Parkinson’s disease
Part VI: Schwab and England ADL scale
These are evaluated by interview and clinical observation.

History of Functional Neurosurgery

Medications often have limited efficacy and often cause intolerable side effects.
Lesioning or surgical destruction of small areas of brain tissue has been done for years. Long before the development of drugs such as Sinemet (Carbidopa/Levodopa).
Early 1990’s, Deep Brain Stimulation (DBS) became an accepted and effective treatment.
It creates a lesioning effect. Allows for alterations in treatment through programming.

Neuromodulation

Advances in imaging and safety of stereotactic surgery.
Improvements in targeting techniques with neuroimaging and microelectrical recordings.
Better understanding in the organization and function of the basal ganglia.
DBS is now directed at modulating activity within the basal ganglia output by stimulating nuclei.
What is Deep Brain Stimulation?

DBS involves surgical implantation of electrical leads into the thalamus. May include high frequency, pulsatile, mono- or bipolar stimulation. Can be activated, inactivated or adjusted with external control device. Reversible, with ability to change parameters over time. Stimulation-related complications are potentially reversible.

Targeting in DBS

As a result of functional organization of the basal ganglia surgical treatment has been aimed at 3 targets:

- **Vim**: Ventral intermediate nucleus of thalamus
- **GPI**: Globus pallidus pars internal
- **STN**: Subthalamic nucleus

Targeting in DBS

Basal Ganglia

The first area used was VIM but was only effective in alleviating tremor.

- Thalamus (VIM) site of choice for tremor control
- GPI exerts the largest effect on dyskinesia
- STN seems to allow more tapering off of medications and requires less electrical stimulation but may contribute to depression or other cognitive effects.
Preoperative testing & selection

DBS team
- Evaluation by movement disorder neurologist
- Neuro-psychological evaluation
- Nurse Coordinator
- Family/social support assessment
- MRI with thin cuts (BrainLAB protocol)
- Neurosurgical evaluation

MRI

DBS surgery

Preadmission testing
Stereotactic headframe placement
CT scan (BrainLAB)
Intra-op microelectrode recording
IPG placement
Programming
Pre-operative planning CT scan

Trajectory planning with BrainLab
Post-operative nursing considerations

Seizures <1%
Intracranial hemorrhage 3%
Confusion/hallucinations 20% may increase with age
Wound healing problems 8%
Speech difficulties
Gait instability
Pain usually is not significant, Tylenol if there are narcotic induced cognitive issues.
• Close neurological and vital sign monitoring (esp. BP control) in the step down unit.
Post procedure & ongoing concerns

Depression
Device infection, can be secondary to compulsive picking.
Skin erosions
Dementia- can be surgical acceleration of pre-operative cognitive deficit.
Long term: eyelid apraxia, weight gain, hypomania, disinhibition, dysarthria, and apathy.
Battery exhaustion.
Imaging

Implantable pulse generator (IPG) placement

Usually placed 1-2 weeks after DBS electrode placement.
Typically an outpatient procedure
Placed subcutaneously near clavicle
IPG can accommodate 1 or 2 leads (surgeon selection)
Programming usually begins 2 weeks later

IPG programming

Goal of programming: Set stimulation to optimize symptom control with minimum side effects.
Patient off medications at least 12 hrs prior to programming.
May take 2-4 hrs
MD neurologist adjusts medications as indicated.
Often takes several sessions
Tremor and balance response seen quickly. Rigidity and bradykinesia responses can take days.
Patient education important.
Thank you

References
http://thalamus.wustl.edu/course/cerebell.html
www.stanford.edu/.../basics/brain/ab6.html