

MOVEMENT DISORDERS GENERAL OVERVIEW



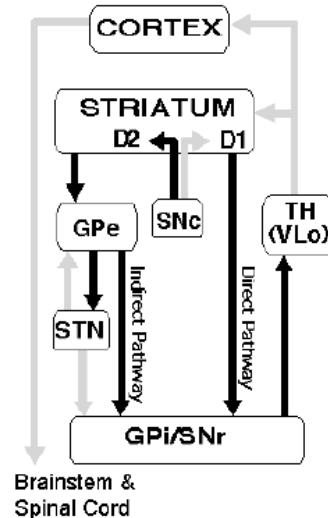
Leili Shahgholi M.D.
Johns Hopkins University

Physical Medicine and Rehabilitation specialist
Movement disorders and Neurorestoration
fellowship
Geriatric Fellowship
Interventional Pain Management Fellowship

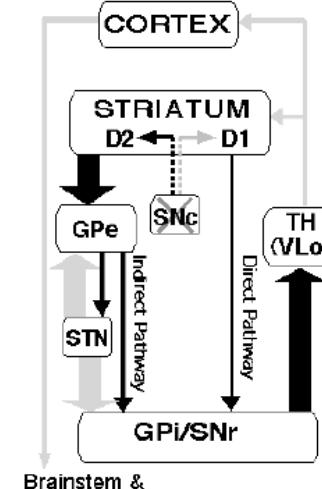
BASAL GANGLIA CIRCUITRY

The Model for Understanding Basal Ganglia Function in PD

A. NORMAL



B. PD



— excitatory
— inhibitory

Movement disorders

- Hypokinesia (Decreased movements)
- Hyperkinesia (Increased movements)

Hypokinesias

- **Parkinsonism**
 1. Akinesia/Bradykinesia
 2. Rigidity
 3. Motor Freezing
- Spasticity
- Apraxias
- Tics (Blocking & Holding Tics, Obsessional Tics)
- Psychiatric
 - Catatonia
 - **Psychomotor Depression**
 - Psychogenic Parkinsonism
- Cataplexy/Drop Attacks
- **Hypothyroidism** or other Endocrinologic Disorders
- **Hesitant Gait of the Elderly**

MSK RELATED

- **1- DJD**
- **2- Rheumatologic disorders**
- **3- Fractures**

Hyperkinesias and Dyskinesias

- **Tremor**
- Chorea
- Dystonia
- Myoclonus
- **Ataxia and Dysmetria**
- Stereotypies and Tics
- **Akathisia**
- Myokymia and Synkinesias
- **Restless Legs**
- Periodic Hypnagogic Movements (PLMS)

Movement Disorders -- Parkinsonisms

- **Parkinson's Disease**
- Diffuse Lewy Body Disease
- Multiple Systems Atrophy (+/- Ataxia)
- Progressive Supranuclear Palsy
- Corticobasal Degeneration
- **Vascular Parkinsonism**
- Post Traumatic Parkinsonism
- **Secondary (Toxin, Medication, Metabolic)**

PARKINSONISMS

Idiopathic Parkinson's Disease

- Usually Asymmetric
- Cardinal Signs
 - Tremor
 - Rigidity
 - Bradykinesia
- Levodopa Responsive
- Later Findings
 - Postural Instability
 - Levodopa-unresponsive gait disorder
 - Non Motor Features (Depression, Sexual Dysfunction, Cognitive Dysfunction of Variable Severity)

Diffuse Lewy Body Disease

- Unlike PD, Lewy Bodies rapidly spready throughout the brain, including the cerebral cortex
- Levodopa Responsive Parkinsonism
- Rapidly Progressive Dementia
 - Hallucinations

Multiple Systems Atrophy

- Cardinal Findings
 - Parkinsonism that is poorly responsive to Levodopa
 - Autonomic Failure (Low Blood Pressure)
 - Cerebellar Signs
 - Long Tract Signs (Spasticity)
- Striato-Nigral Type (Parkinsonism First)
- Shy-Drager Syndrome (Autonomic Failure First)
- Olivo-ponto-cerebellar Type (OPCA – Ataxia First)

Progressive Supranuclear Palsy

- Early Postural Instability and Falls
- Parkinsonism (Unresponsive to Levodopa)
- “Stone Face”
- Dementia
- Ocular Signs

Corticobasal Degeneration

- Asymmetric Parkinsonism with Poor Response to Levodopa
- Apraxia and Alien Limb
- Spasticity, Rigidity, Dystonia
- Gait and Balance Problems
- Dementia always occurs, but may be a late feature

Drug Related Parkinsonism

- Haldol and other antipsychotic medications cause symmetric findings that are indistinguishable at times from Idiopathic Parkinson's Disease
- **Reglan (Metoclopramide)** is a dopamine blocker and is an important cause of Parkinsonism in elderly patients (currently used as an anti-nausea medication)

HYPERKINESIAS

Movement Disorders -- Tremor

- **Essential Tremor**
 - Limbs, Head, Voice
- **Dystonic Tremor**
 - Task Specific Tremors
- Midbrain Outflow Tremors
 - Cerebellar Tremor
 - Rubral Tremor
- Rest Tremor (Parkinsonism)
- Orthostatic Tremor
- **Exaggerated Physiologic Tremor**
 - Caffeine, Medication Withdrawal
- Other (Drug Related Tremor)

Essential Tremor

- Upper Extremity Tremor with Posture and/or Action
- Bilateral, usually roughly symmetric
- Typically of Long-Standing Duration
- Other Areas May Be Involved
 - Head or Jaw
 - Voice
 - Lower Extremities
- Tremor may produce disability, but often doesn't
- No clear association with other diseases or disorders

Myoclonus

- Not a Tremor
- Sudden, shock-like muscle contractions
- Focal, Multifocal, or Generalized
- May be regular and rhythmic, but usually irregular and jerky
- Epileptic or Non-Epileptic
 - Cortical (Epileptic)
 - Brainstem or Spinal Segmental (Non-Epileptic)

Chorea

- Excessive Spontaneous Movements
- Irregular, Random, Brief and Abrupt
- Non-repetitive
- Distal Predominance
- Flitting randomly from one body part to another
- Purposeless, but may be disguised (parakinesias)
- Facial grimacing and abnormal respiratory sounds

Chorea -- Causes

- MEDICATIONS (e.g. tardive dyskinesia)
 - Haldol, other antipsychotics
 - Reglan is an important cause of tardive dyskinesia
- Huntington's Disease
- Hemiballism (Stroke)
- Post-Infectious (Strep Infection)
- Pregnancy
- Other Disorders

TiCS

- Semi-voluntary (e.g. suppressible), rapid, non-rhythmic movements or sounds
- Background of normal activity
- Associated Compulsions
- May be associated with OCD
- Occasionally tics are disabling, but the OCD is usually the more disabling condition, if it exists

Akathisia

- Motor restlessness with a “desire to move”
- Difficult for patients to describe
 - “I feel like I’m always crawling out of my skin”
- Often with associated vocalizations and grunting
- Often very disabling
- Cause: MEDICATIONS – antipsychotics and Metoclopramide (Reglan)

Dystonia

- Sustained, patterned muscle contractions
- Agonists and Antagonists
- Clinical Findings
 - Repetitive Twisting or Squeezing Movements
 - Fixed Postures
- Localization
 - Focal
 - Face (Blepharospasm and Meige Syndrome)
 - Neck (Cervical Dystonia)
 - Limbs (Task Specific Dystonias)
 - Segmental
 - Generalized

Causes of Dystonia

- Idiopathic (Most Cases)
- DRUG RELATED
 - Antipsychotics and Reglan
- Genetic (DYT-1 Gene)
- Structural
 - Trauma
 - Stroke
 - Multiple Rare Diseases

Ataxia

- Symptomatic Interruption of the Cerebellar Pathways
 - Dysmetria (Poor targeting of planned movements)
 - Dysdiadochokinesia (Poor sequencing of planned movements)
 - Intention Tremor (Past Pointing)
- Gait Abnormalities
 - Wide based gait
 - Poor balance
 - “Drunken Sailor’s Walk”

Causes of Ataxia

- Lesion in the following areas of the brain
 - Cerebellum
 - Pons
 - Midbrain (Red Nucleus)
 - Olivary Nucleus
 - Thalamus (Cerebellar Inflow Nucleus)

Causes of Ataxia

- Acute intoxication (Alcohol, Dilantin, others)
- Chronic Substance Abuse (including Alcohol)
- Stroke
- Multiple Sclerosis
- Brain Tumors
- Genetic Ataxias
 - Autosomal Dominant Ataxias
 - Autosomal Recessive Ataxias
- Metabolic
 - Vitamin E, Thiamine,
- Multiple Systems Atrophy

Restless Legs

- Bothersome urge to move legs at night
- Prevents sleep

Treatment of Parkinson's Disease

- Levodopa and Dopamine Agonists
- Amantidine, MAO-B Inhibitors
- Deep Brain Stimulation (Subthalamic Nucleus, Globus Pallidus Interna)

Treatment of Dystonia

- Artane, Valium, occasionally Baclofen
- Botulinum Toxin Injection
- Deep Brain Stimulation (Globus Pallidus Interna)

Treatment of Essential Tremor

- Primidone
- Propanolol and other Beta Blockers
- Deep Brain Stimulation (Thalamus)
- Tremors of some patients are quite responsive to alcohol, and patients may self-medicate

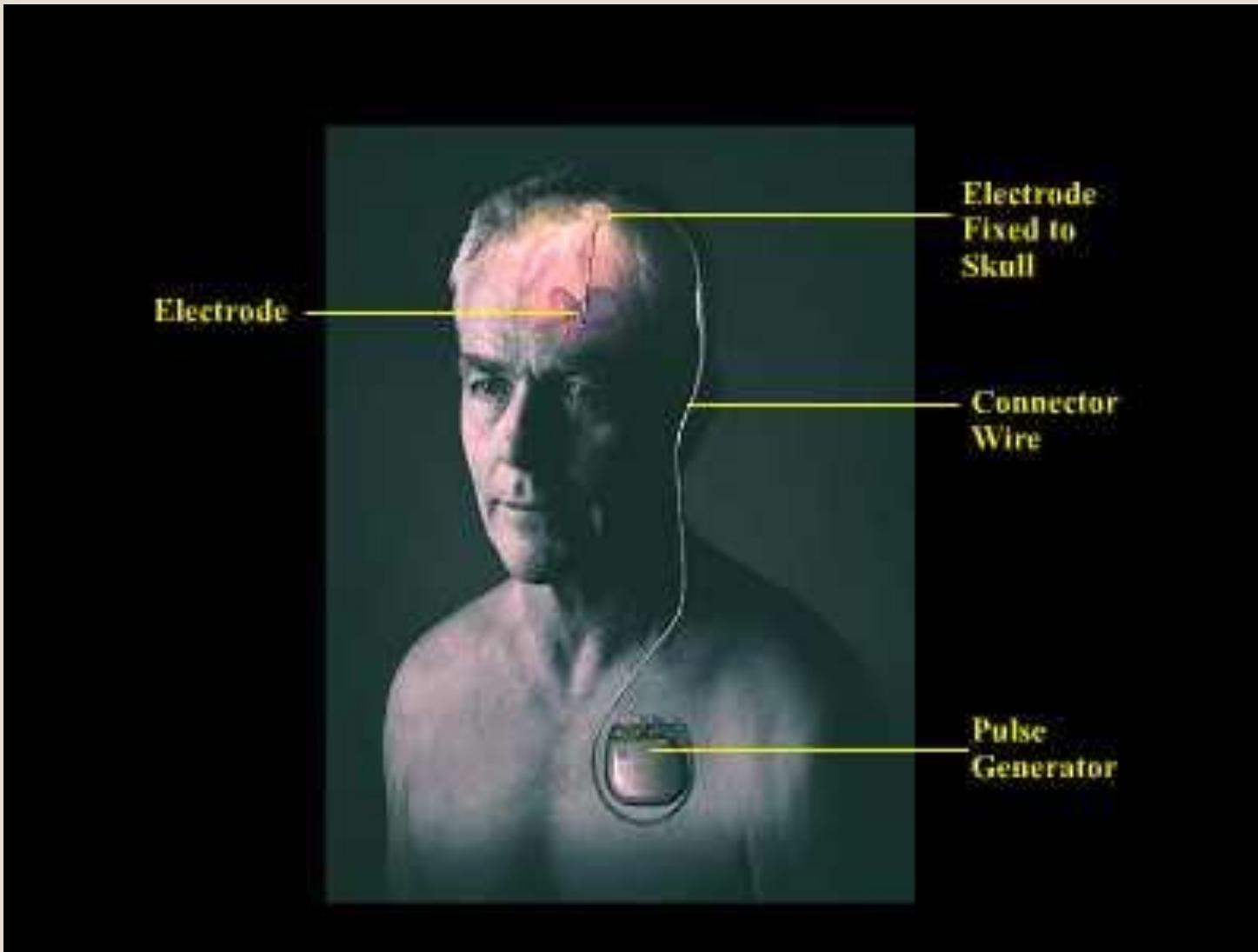
Treatment of Tardive (Post Dopamine-Blockade) Movement Disorders

- PREVENTION
- Dopamine Depleting Agents (Difficult to Obtain in the U.S.)
- Ramp up dopamine blockade (often worsens the underlying pathologic process)
- Life Long Botulinum Toxin Injections
- Deep Brain Stimulation

Complications of Dopamine Blocking Agents

- Reversible? Sequelae Include
 - Tremor
 - Parkinsonism
- Permanent and Difficult to Treat Sequelae Include
 - Tardive Dyskinesia (Chorea-like syndrome)
 - Tardive Akathisia
 - Tardive Dystonia (Long Term Botox, may need DBS)

DBS



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