Movement Disorders

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Neurology

Classify

- Bradykinesia
- Tic
- Myoclonus
- Tremor

- Dystonia
- Athetosis
- Chorea
- Ballismus

This 64 year-old female has had progression of a tremor over the last two years. It affects her writing, eating and sewing.

Tremor: Definition

- Involuntary
- Rhythmic
- Sinusoidal movement

General types

- Action
 - » Kinetic
 - » Postural
 - Intention (cerebellar)
- Resting

Essential tremor

- Most common movement d/o
- Postural and kinetic
- Slowly progressive
- Prevalence increases with age
 - » Mean age of presentation 35-45
 - » Adolescence onset
 - » 0.5-5% population
 - » F:M 1:1
- Genetic AD
 - » FH (+) 50-60%

- Distribution
 - » Hands (symmetric)
 - » (30%) head, voice, jaw, face
 - » Legs
 - » Trunk
- Worsened by:
 - » Emotion
 - » Hunger
 - » Fatigue
 - » Temperature
 - » Drugs/toxins

Drugs/toxins

- Steroids
- Thyroid hormone
- Lithium
- Valproic acid
- Cyclosporine
- Tricyclics
- Beta agonists
- Theophyline
- Neuroleptics
- SSRIs

- Caffeine
- Nicotine
- Arsenic
- Lead
- Cocaine/amphetamines
- Alcohol withdrawal
- Benzo withdrawal
- Other drug withdrawal

Tremor work-up

- None if c/w essential tremor
- Resting tremor is the first distinction to make
- Review medications
- Ask about alcohol, illicit drugs
- Labs
 - » M7, LFTs, TSH, B12
 - » Ceruloplasmin and serum Cu
 - Wrine tox screen
 - » Rare: Vit E, heavy metals
- MRI
- Review psychiatric history

This 58 year-old male has a left hand tremor at rest and some trouble walking. His hand writing has become smaller, and he no longer smiles.

PD: Clinical

- Slow progressive course
- 4 cardinal features
 - » Resting tremor
 - » Bradykinesia
 - » Rigidity (cogwheel)
 - » Postural reflex impairment
- Other: dementia, depression, autonomic

PD: Levodopa

- Sinemet = carbidopa/levodopa
- Most potent drug for PD
- Preferred treatment age > 75
- Side effects
 - » GI
 - » Dyskinesias
 - » Dementia, hallucinations
 - » Orthostatic hypotension
- Start 25/100 QD, increase q 3d up 3 tid as needed

PD: Dopamine Agonist

- Act directly on dopamine receptors
- Synergy with levodopa
- Longer T1/2
- Reduced dyskinesias
- Same side effects as levodopa
- Initial treatment age < 65
- 3rd generation options(Pramipexole, Ropinirole)

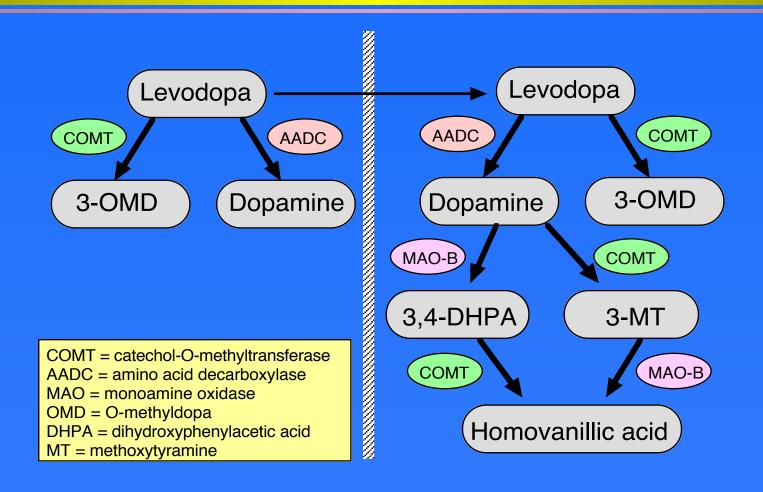
PD: MAO-B Inhibitors

- Rasagiline and Selegiline
- Reduction of motor fluctuations
- May have neuroprotective effects
 - Consider in early onset disease

PD: COMT inhibitors

- Block COMT metabolism of levodopa
- Dopaminergic SEs
 - » Reduce levodopa by 30%
- Entacapone (Comtan)
- Combination drug
 - » Carbidopa/levodopa/entacapone

Treatment Flow Chart



This 58 year-old male is having trouble walking in that he can't seem to keep up with his wife. Several falls have occurred. His hand writing has become smaller, and his wife wonders if he hasn't had a stroke because he no longer smiles. No tremor is present. Sinemet does not help.

Parkinsonism

- Parkinson's Disease
- Neuroleptic side effect
- Post-encephalitic
- Toxins (Mn, CO, MPTP)
- Dementia
- Wilson's disease
- Basal ganglia calcifications

- Parkinson-plus syndromes
 - » Progressive supranuclear palsy
 - Corticobasoganglionic degeneration
 - » Multisystem atrophy
 - Autonomic
 - Cerebellar
 - » Diffuse Lewy body disease

This 47 year-old woman who has trouble sitting still in the evenings. Her legs ache. Walking will temporarily relieve the problem.

Restless Leg Syndrome

- Characterized by an irresistible urge to move the legs, accompanied by uncomfortable sensations that often occur in the evening or when at rest.
 - » Creeping, crawling, tingling, aching, burning, pulling, itching, cramping, electric current-like

RLS - Prevalence

Very common

- Estimated that 2-15% of the population may have RLS symptoms.
- Age of onset varies widely, but onset of symptoms typically presents before age 50. Severity increases with age.
- Greater in women than men

RLS – Etiology

- Primary: Thought to be a CNS disorder, but exact mechanism unknown.
 - » Familial cases indicate potential genetic component.
- Secondary: Iron deficiency, neurologic lesions, pregnancy, uremia, drug induced, diabetes mellitus, venous insufficiency.

RLS - Diagnosis

Key diagnostic criteria	Supportive Features
Urge to move legs – usually with uncomfortable leg sensations.	Sleep disturbances
Temporary relief with movement – partial or total relief by walking or stretching.	Involuntary leg movements during sleep
Onset or worsening with inactivity	Family history
Worsening or onset of sx in the evening or at night.	Positive response to dopaminergic tx.

RLS - Treatment

- Non-Pharmacologic:
 - » Stretching/massage
 - » Long compressive socks in bed
 - » Hot/Cold compresses
 - » Hot bath before bed
 - » Avoid caffeine and alcohol before bed
 - » Daily exercise
 - » Mentally alerting activities

- Pharmacologic tx:
 - » Dopamine agonists: pramipexole, ropinerole
 - » Carbidopa-levodopa
 - » Clonazepam
 - Gabapentin, Carbamazepine.
 - Iron: indicated in all pts (if anemic), esp. pregnant women.
 - » Opioids

This 42 year-old female complains of a progressive inability to keep her arms and legs still. She has had depression for 10 years. Her father had the same movements starting at the age of 63.

Huntington Disease

- Genetics
 - CAG repeat
 - » AD
 - » Chrom 4
- Pathology
 - » Loss of GABA neurons in striatum
 - » Diffuse atrophy

- Clinical
 - » Chorea
 - » Dementia
 - » Psychiatric
- Treatment
 - » Neuroleptics
 - » Clonazepam
 - » Tetrabenazine
 - Antidepressants

This 35 year-old woman has trouble writing more than one sentence. It started 2 years ago and progressed. No weakness or sensory loss. She can play the piano and eat fine.

Focal dystonias

- Writer's cramp
- Spasmodic torticollis
- Blepharospasm
- Spasmodic dysphonia

- Treatment
 - » Physical therapy
 - » Botox
 - » Levodopa
 - » Anticholinergics
 - » Baclofen
 - » Surgery

This 19 year-old male complains of excessive blinking and the need to clear his throat. Onset was at the age of 8. In school he was always in trouble for not paying attention and making abnormal sounds.

Tourette Syndrome

- Clinical
 - » Tics: motor and vocal
 - Involuntary but suppressible
 - » Daily > 1 year
 - » Onset before age 21 (median age = 7)
 - » Lifelong, decline with age
 - » OCD
 - » ADHD

- Prevalence 40/100k
- Genetics
 - » 4:1 M:F
 - » AD with incomplete penetrance
 - » MZ twins 80%

Tourette Syndrome

Treatment

- » Counseling
- » Neuroleptics
- » Clonidine
- » Topiramate
- » Leviteracetam
- » Clonazepam
- » Dopamine agonists
- » Clomipramine
- » SSRIs
- » Nicotine patch/gum