MOVEMENT DISORDERS

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What Are Movement Disorders?

- diseases/conditions/syndromes made up of one or more specific abnormal movements eg. Tremor vs PD
- are the result of many varied primary and secondary causes eg. HD vs drug exp.
- can be classified into hypokinetic vs hyperkinetic movements and neurodegenerative vs non degenerative

Describing Movement Disorders

- rate—fast or slow
- rhythm--- repetitive or unpredictable eg. tic vs tremor
- range---amplitude
- region---body part involved, proximal vs distal
- redirection---distract the patient eg. psychogenic
- present or not during sleep

Huntington's Disease

- prevalence 1/10,000 in Canada
- symptom onset from childhood to late 80's with mean age of 40
- Juvenile—onset before age 21 having higher CAG rpts typically from father
- disease duration about 20 yrs and juvenile about 10 yrs
- completely penatrant with 40 CAG rpts or higher (normal is less than 27)

Huntington's Disease--Pathology

- autosomonal-dominant neurodegenerative condition involving the huntingtin gene located on chromosome 4
- caused by expanded CAG (cysteine-adenosine-guanine) repeat lengths
- huntingtin protein widely expressed in brain
- mutant protein forms aggregates creating toxic environment for striatal neurons
- atrophy to neostriatum (caudate and putamen) as well cortex, brainstem, and cerebellum

Huntington Disease Presentation Motor Symptoms

- chorea, ocular abnormalities, motor impersistance
- chorea perceived by pt as clumsiness
- chorea incorporated into purposeful movement
- gait, balance and swallowing difficulties
- with disease progression axial rigidity and dystonia present, chorea decreases

Huntington Disease Presentation Psychiatric Symptoms

- can predate onset of motor symptoms resulting in misdiagnoses
- personality changes, impulsivity, irritability, aggressive behavior, social withdrawl and sexual disorders
- OCD—fixated thoughts
- depression and suicide risk higher
- paranoia and delusional thinking—lack of insight

Huntington Disease Presentation Cognitive Symptoms

- executive dysfunction, short term memory loss, inattention, planning and organizational difficulties
- information/visuospatial processing difficulties
- speech and language dysfunction—tangential speech
- sleep disturbances
- dementia

Huntington Disease Treatment

- symptomatic treatment using medications
- Tetrabenazine (Nitoman)12.5 – 25mg tid useful in treating chorea—dopamine antagonist/depletor—can cause depression
- mood stabilizers such as Valproate
- antidepressants and antipsychotics

Huntington Disease Management

- importance of thorough family history before genetic testing done
- genetic counseling and education for all family members involved
- end of life decisions should be made early
- safety measures w/r to home environment, swallowing (OT PT SLP)
- advocacy and support measures (CCAC, Huntington's Society)

Parkinson's Disease

- Second most common neurodegenerative disease
- Prevalence is 160/100,000 and is expected to rise
- Affects 1 – 2% of persons age 60+
- Mean age of onset is 55

Parkinson's Disease Risk Factors

- increases with age
- male gender
- 2 – 3x higher lifetime risk with family history of PD
- genetics, parkin mutations identified, younger onset
- environmental toxins—farming, welding, well water

Classification of Parkinson's Disease

- Tremor dominant—most benign
- Akinetic rigid dominant—most progressive
- Mixed—most common
- Postural instability dominant
- Young onset –age 40
- Juvenile onset –age 21

Pathology of Parkinson's Disease

- Degeneration of dopamine producing cells in the substantia nigra in BG resulting in cell death and formation of Lewy Bodies
- Braak's staging 1.dorsal motor nucleus of the vagus and olfactory tracts 2.spreads to other lower brainstem nuclei including sleep regulation 3.spreads upwards affecting SNc and motor symptoms appear 4. moves upwards to the cortex
- Also involved –cholinergic, norepinephrine, serotonin and neurons in cerebral cortex, brainstem, spinal cord, and peripheral ANS—multisystem disease

Motor Manifestations of PD

- Rest Tremor—70% of PD pts, asymmetrical, distal part of upper ext., worsens with stress, resolves with sleep
- Bradykinesia/Akinesia—slowness of all movement including spontaneous movement (swallowing, blinking) and gross/fine motor
- Rigidity—increased muscle tone during passive ROM, stiffness in neck/shoulder/LBP
- Postural instability and loss of postural reflexes—stooped and leaning to one side
- Gait dysfunction– tachykinesis, festination, and FOG

Medications Used in PD

- Levodopa/Carbidopa (Sinemet) 100/25, 100/10, 250/25, CR 100/25, CR 200/50
- Levodopa/Benzeride (Prolopa) 100/25, 50/12.5
- Stalevo=Levodopa/Carbidopa+Comtan (COMT Inhibitor)
- Azilect (Rasagaline)—MAO-B Inhibitor—FOG, possible neuroprotection
- Amantadine—dyskinesia, DBS, livedo reticularis, ankle edema, confusion
- Anticholinergics—Parsitan, Artane—to treat tremor

Medications Contraindicated in PD

- Maxeran (Metaclopramide)
- Stematil (Prochlorperazine)
- Demerol
- Zofran (Ondansetron) is a good antiemetic to use in PD
- narcotics typically cause hallucinations

Proper Levodopa Administration

- Similar construction as amino acid protein molecule
- *MUST BE GIVEN 1 HOUR BEFORE MEALS OR 2 HOURS AFTER MEALS* for maximum efficacy
- May be given with SMALL NON PROTEIN snack only
- Absorbed by active transport in proximal small bowel—need for regular BM

Side Effects of Levodopa

- nausea and GI upset initially—titrate slowly, add domperidone
- dyskinesia—involuntary movement more pronounced with higher doses
- constipation, postural hypotension, hallucinations/confusion
- motor fluctuations—unpredictable on/off
- BP lowering tendency over time

Medications Used in PD—Dopamine Agonists

- Mirapex (Pramipexole) 0.5-1.5mg tid
  Requip (Ropinerole) 2-4mg tid
- Synthetic dopamine used in early PD and in conjunction with Levodopa and in RLS
- Confusion, hallucinations, leg/ankle edema
- Impulse Control Disorder (ICD) gambling/shopping/eating/hypersexual, hypomania
- Dopamine Dysregulation Syndrome (DDS)

Non Motor Manifestations of PD – Autonomic--GI

- Swallowing—drooling, coughing/choking with food/saliva, silent aspiration
- Delayed gastric emptying and GI motility—nausea, bloating
- Constipation and denervation of anal sphincter—high fibre diet, restrict caffeine, add stool softeners/lactulose

Non Motor Manifestations of PD—Autonomic--Cardiovascular

- Orthostatic Hypotension—high risk for falls, levodopa contributes to this
- Importance of sitting and standing BP
- Adequate hydration—esp. in a.m., assess caffeine intake
- Assessment of antihypertensives and proper administration of antihypotensives (Amatine)
- Cardiac sympathetic denervation

Non Motor Manifestations of PD—Autonomic--Genitourinary

- frequency/urgency/nocturia—risk for falls
- prone to chronic UTI's
- incontinence later on--immobility
- meds to treat often contain anticholinergic properties—confusion
- use of botulinum toxin
- erectile dysfunction

Non Motor Manifestations of PD—Autonomic--Thermoregulatory

- Hyperhidrosis—excessive sweating mainly in face and trunk
- often occurs in "off" state
- temperature dysregulation

Non Motor Manifestations of PD—Sleep Dysfunction

- Impaired initiation and maintenance
- REM sleep behavior disorder (RBD)
- RLS and PLMS
- Excessive daytime somnolence--meds
- Sudden onset of sleep—dopamine agonists
- Nocturnal akinesia and urination
- Sleep Apnea

Non Motor Manifestations of PD—Sensory Symptoms

- Hyposmia—impaired olfactory function resulting in loss of smell and taste—weight loss
- Pain—numbness/tingling/burning and muscle cramping, toe curling (dystonia), rigidity and tremor

Non Motor Manifestations of PD—Anxiety and Depression

- Neurochemical imbalance
- Often occurs in “off” state
- Panic attacks and fear of being alone
- Mild to moderate—suicide rare
- Apathy—loss of motivation and interest in pleasurable activities

Non Motor Manifestations of PD--Psychosis

- Hallucinations--presence/passage/formed—UTI/URI can trigger, meds, dementia
- Paranoia/delusional thinking/illusions
- Visual disturbances—loss of colour discrimination/contrast sensitivity, diplopia, blurred vision, cataracts, floaters
- Treat source/give antipsychotic--Seroquel

Non Motor Manifestations of PD—Cognitive Impairment

- Bradyphrenia—slowed information processing
- Executive dysfunction similar to ADD
- Poor short term memory
- Speech and language dysfunction
- Visuospatial dysfunction—MOCA—replication of cube/clock drawing

Non Motor Manifestations of PD--Dementia

- occurs in 30 – 40% of PD pts later in disease process, PD-D vs DLB
- confusion/disorientation/unintelligible speech, excessive somnolence
- Risk Factors: age, akinetic/rigid type
- hallucinations without insight
- Treatment with cholinesterase inhibitors—Exelon (Rivastigmine)-GI

Surgical Treatment in PD—Deep Brain Stimulation

- Criteria: Response to Levodopa, age, no significant cognitive impairment, MRI
- Indications: Motor fluctuations, dyskinesias, young onset
- Target: Subthalamic Nucleus (STN)—PD meds reduced
- Adverse Effects: Worsening speech and balance, depression and apathy, personality change

Dystonia

- involuntary, sustained, patterned, repetitive muscle contractions of opposing muscles resulting in abnormal postures
- dysfunction of BG
- continuous or paroxysmal
- task specific or spontaneous
- sensory trick (geste antagoniste)

Dystonia--Classification

- Age of onset
- Affected body region—focal, segmental, hemidystonia (lesion in BG), multifocal, generalized
- Primary (Idiopathic)(CD), Dystonia Plus—with parkinsonism or with myoclonus (DRD), Secondary, Inherited

Cervical Dystonia (Torticollis)

- Most common focal, primary, adult onset (female)
- Involves muscles of neck region
- Often associated with pain
- Subdivided into laterocollis (head tilt to side), anterocollis (head flexion), and retrocollis (head extension)

Treatment of Cervical Dystonia

- Botulinum toxin A—takes 7-10 days to take effect and repeated q3months
- Botox/Xeomin—interferes with release of acetylcholine at the level of neuromuscular junction
- Side effects: dysphagia
- Deep Brain Stimulation in GPi

Dopa Responsive Dystonia

- Secondary, inherited, dystonia-plus, non degenerative condition
- Onset in childhood age 5-6, female
- Involves lower extremity first affecting gait
- Symptoms worsen later in day and improve after sleep
- Excellent response to small dose levodopa

Tourette Syndrome (TS)

- Neurodevelopmental/neuropsychiatric disorder with complex genetic link
- Tics begin age 6-8 and occur in 1% of all children
- Tics—sudden, rapid, recurrent, nonrhythmic, stereotyped movement
- urge to make movement/vocalization and ability to suppress
- Must have motor tics and one vocal tic for at least one year for diagnosis of TS

Tourette Syndrome—Simple and Complex Tics

- Simple Motor Tic: eye blinking, eyebrow raising, grimacing
- Simple Vocal Tic: sniffing, grunting, throat clearing, clicking
- Complex Motor Tic: head shaking, jumping
- Complex Vocal Tic: Coprolalia (foul language), whistling, panting, barking

Tourette Syndrome

- 80 – 90% have ADHD, OCD, depression, anxiety, emotional dysregulation, autoaggression, and conduct disorders
- Tics spontaneously improve in adolescence in most

Treatment Options in TS

- Provide opportunity for children to relieve tic at school and education to caregivers
- Forcing child to stop making movement will exacerbate tic
- Only use medications if child is functionally impaired or socially isolated—bullying
- Adult TS—use of botox, mariguana, dopamine blockers (Orap, Zyprexa, Risperdal)
- Behavioural Therapy—Habit Reversal Training
- Deep Brain Stimulation

Normal Pressure Hydrocephalus

- CSF reabsorption issue seen in elderly
- Symptoms due to enlarged ventricles putting pressure on underlying structures
- Symptom Triad: Gait disturbances, cognitive decline, and urinary incontinence

NPH Treatment Options

- Lumbar Puncture—30-50cc CSF removed with gait and cognitive assessment pre and post
- Ventroperitoneal Shunt—divert CSF from ventricles to peritoneum for absorption
- CSF pressure low or normal—resevoir pressure must also be low