INVOLUNTARY MOVEMENTS

(Extrapyramidal) Basal ganglia circuitry: modulates and enhances voluntary movement

- Pathways enter through striatum and exit through thalamus
- Direct (excitatory) pathway involves D1 receptors
- Indirect (inhibitory) pathway involves D2 receptors

Framework for involuntary/abnormal movements:

1) Characterize the abnormal movement – descriptors include:
    a. Hyperkinetic vs. hypokinetic
    b. Rhythmic vs. arrhythmic
    c. Degree of voluntariness
    d. Stereotypic vs. non-stereotypic
    e. Character (e.g., jerky vs. oscillatory vs. flowing, etc.)
2) Determine the phenomenology, followed by the syndrome associated with the phenomenology

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TREMOR: hyperkinetic, rhythmic, oscillatory, bidirectional; characterized by alternating/synchronous activity of agonistic and antagonistic muscles

Action (including kinetic, postural, & isometric) tremors

- Essential tremor: chronic, progressive, medium-frequency tremor – usually in the hands and arms bilaterally
    - May progress to head/voice, ± cerebellar signs and intention tremor later in disease course
    - Family history is frequently positive for ET
    - Test with spiral-drawing – tremor worsens on an axis
    - 1st-line treatment = primidone or propranolol
- Enhanced physiologic tremor: high-frequency, low-amplitude tremor that emerges in certain settings (e.g., stress, sympathetic stimulation) – usually in the hands/voice
    - No cerebellar signs or intention tremor
    - Transient once the underlying cause is reversed
- Cerebellar tremor: low-frequency tremor with intentional component (worsens when approaching target)
    - Seen in the setting of any cerebellar pathology
    - Associated with other cerebellar signs (eye movement and gait abnormalities, ataxia, dysdiadochokinesia)

Resting tremors: associated with parkinsonism (bradykinesia PLUS rigidity or tremor)

Parkinson's disease: bradykinesia + hypokinesia + low-frequency "pill-rolling" tremor + cogwheel rigidity

- Other signs: masked facies, stooped posture, shuffling gait & en bloc turns, decrement in repetitive movements (finger tapping)
- Postural instability appears later in course, seen on pull test
- Non-motor: cognitive complaints, mood disorders, REM sleep disorders, autonomic dysfunction, anosmia
- Diagnosis is clinical – DaT scan is helpful if unsure
- Treatment: dopaminergic meds (e.g., Sinemet); DBS for refractory cases

Parkinson-plus syndromes

- *Multiple system atrophy*: MSA-C (cerebellar) and MSA-P (parkinsonism; extreme autonomic dysfunction) variants
- *Corticobasal degeneration*: parkinsonism plus cortical signs (language dysfunction, apraxia, alien-limb phenomenon)
- *Progressive supranuclear palsy*: parkinsonism plus eye movement abnormalities and profound postural instability

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CHOREA: hyperkinetic, arrhythmic, non-stereotyped, flowing movements

- Part of a choreiform spectrum, along with:
    - Athethosis = slower flowing movement
    - Ballismus = large amplitude flinging movement
- Etiologies include Huntington's disease, Sydenham chorea (rheumatic fever), pregnancy, stroke, celiac, APLS, hereditary

Huntington's disease: cognitive impairment, chorea (choreoathetosis and hemiballismus), and psychiatric symptoms

- Autosomal dominant trinucleotide repeat (CAG) expansion in huntingtin gene, causing protein aggregates
- Ocular signs: slow eye movements, increased saccade latency
- Parkinsonism can appear later in course or in young patients
- Diagnosed with genetic testing – requires genetic counseling
- Manage symptoms with dopamine blockade

MYOCLONUS: hyperkinetic, arrhythmic, jerky, unidirectional; brief contractions or pathologic relaxation of specific muscle groups

- Positive myoclonus = abnormal contraction
- Negative myoclonus = asterixis; pathologic relaxation
- Causes include:
  - Physiologic (e.g., hiccups)
  - Toxic-metabolic (e.g., medications, uremia, hyperammonemia)
  - Hypoxic-ischemic brain injury
  - Epileptic (e.g., JME)
  - Genetic

DYSTONIA: hyperkinetic, arrhythmic; sustained muscle contraction resulting in abnormal posturing and/or twisting

- Generalized dystonia: usually idiopathic or genetic/inherited
- Cervical dystonia: focal dystonia of the neck; torticollis
- Blepharospasm: focal dystonia of the eyes
- Task-specific dystonia: focal dystonia due to overuse (e.g., musician's dystonia, dystonic writer's cramp)
- Secondary dystonia due to medications/reactions: seen with dopamine-blocking medications (e.g., acute dystonic reaction, tardive dystonia)

Treatment options for dystonia:

- Benzodiazepines, muscle relaxants, anticholinergics
- Consider botulinum toxin injection if focal
- Consider DBS if genetic

TICS: hyperkinetic, stereotyped, semi-voluntary (can be suppressed for short periods, but urge to perform the movement increases)

Tourette syndrome: characterized by both motor and vocal tics, with onset in childhood

- Motor tics include grimacing, shrugging, blinking, head jerking
- Vocal tics include sniffing, throat clearing, other utterances
- Frequently comorbid with other psychiatric disorders (e.g., OCD, anxiety)
- Management:
  - Mild tics can be managed with supportive care/counseling
  - Psychotherapeutic techniques = comprehensive behavioral intervention for tics (CBIT), habit reversal therapy (HRT)
  - More bothersome tics can be managed with dopamine blockers, alpha agonists (guanfacine, clonidine)

Key features of the examination in movement disorder patients:

- Movement exam: think on your feet!
  - Observation – beginning from the moment you encounter the patient!
  - Test for emergence of movements during distraction (e.g., cognitive load) or activity (e.g., spiral drawing)
  - Muscle tone and bulk
  - Repetitive movements and tests for ataxia
  - Gait assessment and pull test
- Mental status testing
- Extraocular movements, gaze, tracking and saccades
- Speech, including clarity and phonation

For additional resources/videos, check out Dr. Schaefer's modules at movementmodules.yale.edu.