Benign essential tremor or Neurodegenerative Brain Disease?
Clinical pearls in history and assessment for the adult patient presenting with tremor.

Tremors

Intended Audience

- Nurse Practitioners (NP) or Physician Associates (PA) specializing in movement disorders, neurology, neurosurgery
- NP/Pas in primary care who are interested in appropriate diagnosis and initial management of tremor prior to and/or while awaiting specialty consultation

Disclosures

- No relevant financial disclosures

Educational goals

By the end of this lecture, the NP/PA shall

- Be confident in identifying etiology and common characteristics of essential tremor (ET) and Parkinson disease (PD)
- Identify basic differential diagnosis in tremor
- Be successful in meaningful history taking in assessment of tremor
- Have several physical assessment strategies to diagnose tremor
- Be confident is identifying how ET and PD present differently

Do I have Parkinson disease?

- Take a thorough history including family history, medications current and PAST, psychiatric care, environmental exposures, drug history, head trauma, etc.
- EXAMINE your patient.
- When the story doesn't make sense, dig further. Labs, imaging.
- REASSESS.

Essential tremor

- A disorder of the nervous system that causes involuntary, rhythmic shaking
- No other significant associated neurologic signs or symptoms
- Can involve any part of the body. Most common in hands. Trunk, face, head, neck, voice. Legs not as common

Also known as

- Familial tremor
- Benign essential tremor
- Benign familial tremor
- ET

Clinical features of ET

- Moderate frequency, Rhythmic oscillations
- Most often bilateral and symmetric
- Begins gradually (often years or even decades of history)
- Worse with movement/intention
- Mild or absent at rest
- Aggravated by emotional distress, fatigue, caffeine, temperature extremes

Diagnosis is CLINICAL

- Clinical history
- Family history (genetic?)
- Examination
- Absence of abnormal lab findings (TSH)
- Medications (psych meds, seizure meds, cardiac meds, many!)
- Effect of alcohol (stereotypically will suppress temporarily)
- Archimedes spiral – can see the rhythmic tremor in the writing.
- Handwriting test

Inclusion criteria

- Bilateral, largely postural or kinetic tremor involving the hands or forearms
- Tremor is persistent and visible (not distractable)
- Additional or isolated tremor of the head (midline) in the absence of abnormal posturing (dystonia)

Exclusion criteria

- Other neurologic signs
- Presence of known causes of enhanced physiologic tremor
- History or clinical evidence of psychogenic tremor (variable, distractible)
- Convincing evidence of sudden onset or stepwise deterioration (stroke, lesion, metabolic)
- Isolate voice tremor, position or task specific tremor, tongue or chin tremor, leg tremor

Clinical history intake ideas:

- Where is the tremor?
- When did it start?
- How has it changed/has it changed?
- Does drinking alcohol affect the tremor?
- Does drinking caffeine affect the tremor?
- Any trouble eating/drinking?
- What activities are affected by your tremor?
- Have you ever been/are you taking antidepressants or antipsychotic medications?
- Any recent illnesses/physical ailments?
- Does anyone in your family have tremor?
- Does the tremor bother you?

Pearls for physical examination

- Postural and kinetic tremor assessment
- Bradykinesia assessment, gait assessment, reflexes, eye movements (should all be NORMAL)
- Archimedes spiral, handwriting
- Hold a piece of paper
- Pouring water from cup to cup

Differential diagnosis

- Metabolic disorders: thyroid disease, alcohol withdrawal, excessive caffeine
- Drug induced:
  - Amiodarone
  - SSRIs/SNRI
  - Seizure medications
  - Many others!
- Zebras: pheochromocytoma, Wilson's disease

Prognosis

- This is NOT considered a neurodegenerative disease
- NOT treating is an acceptable option, especially if the tremor is not bothersome to patient or disabling

Pearls to remember

- BOTH hands/arms and possibly head, rarely legs involved
- Postural and kinetic, minimal if at all at rest
- TREMOR is the disease/brain signaling issue
- Alcohol tends to temporally suppress tremor
- Caffeine/agitation/sleep deprivation will exacerbate tremor
- Think hands and head. (Jaw tremor tends to be PD.)

Treatments

- Stress reduction and relaxation techniques
- Weighted silverware
- Beta Blockers, esp propranolol
- Seizure medications
- Deep Brain Stimulation (VIM) and High focused ultrasound.
- New on horizon: CalaTrio

Parkinson disease

- Degeneration of substantia nigra pars compacta dopaminergic neurons = NOT ENOUGH DOPAMINE in the brain
- Causes dysfunction of the basal ganglia function
- HYPOkinetic movement disorder
- Tremor CAN BE a SYMPTOM
- Why? Genetic and environmental.
- Diagnosis PEARL: must be LEVODOPA responsive

Disease of the basal ganglia

- Disruption of basal ganglia circuits impairs automatic execution of **learned motor plans**
- Involuntary Movements/Tremor (hyperkinetic)
- Muscle Rigidity (hypokinetic)
- Immobility without Paralysis (hypokinetic)
- Complex mood, cognitive, and behavioral disturbances

Differential diagnosis

- Essential tremor (ET)
- Progressive supranuclear palsy (PSP)
- Multiple system atrophy (MSA)
- Corticobasal degeneration (CBD)
- Diffuse Lewy Body Dementia (LBD)
- Alzheimer disease (AD)
- Drug induced Parkinsonism
- Vascular parkinsonism (Lower body parkinsonism)

Zebras: rare and rarer

- Wilson's disease (copper)
- Huntington's disease (careful family history)
- DRPLA
- Spinocerebellar ataxia
- Metabolic disorders
- Structural lesion (this is why MRI is NOT part of routine diagnosis)
- Hydrocephalus
- Infectious encephalitis

Clinical Pearls to History Taking

- Any problems with fine motor skills (buttoning shirts, tying shoes, cutting food, etc) (stiff and slow)
- Do you feel like it is taking you longer to get ready and move around? (stiff and slow)
- Any trouble with balance? **Getting up out of chairs**, turning, bending, reaching above head? Have you had any near falls? (gait instability)
- Loss of smell or taste (nonmotor)
- Trouble rolling over in bed or "adjusting bed linens" (stiff and slow)
- Acting out dreams (?precursor)

History taking continued.

- Changes in handwriting (smaller and smaller until illegible)
- Loss in facial expression (slow)
- Softer, raspier voice
- Numerous nonmotor symptoms: constipation!!! Apathy, mood changes, intense or vivid dreams, and many more.

Flags for ATYPICAL Parkinsonism

- Early hallucinations – Lewy Body
- Multiple Autonomic complaints (constipation, urinary incontinence, orthostasis, erectile dysfunction, inappropriate sweating or lack thereof) – MSA
- Early and frequent falling, severe postural instability – PSP (we talk about double vision and gaze palsy but often that comes LATER)
- "it just won't do what I tell it to do" - CBD

Clinical Exam in Neurodegenerative Parkinsonism

- REMEMBER DIAGNOSTIC CRITERIA
  - 1. MUST HAVE BRADYKINESIA (without this, no PD)
  - 2. MUST HAVE RIGIDITY AND/OR TREMOR
  - 3. ONCE PARKINSONISM NOTED (1 AND 2 ACHIEVED)
    - EXLUDE RED FLAGS AND ABSOLUTE EXLUSIONS
    - DOCUMENT SUPPORTIVE CRITERIA
    - TIME IS ON YOUR SIDE! "PARKINSONISM" IS OFTEN A TOTALLY APPROPRIATE DIAGNOSIS!

EXAM PEARLS

- 1. FIND THE BRADYKINESIA (LOSS OF AMPLITUDE AND/OR PAUSES/BREAKS)
- 2. TREMOR AND RIGIDITY (NEED ONE OR BOTH)
- 3. GAIT INSTABILITY
- STOOPED POSTURE, MASKED FACE, HYPOPHONIA, SHUFFLING, TURNING EN BLOC, TREMOR WITH AMBULATION **ARE NOT NECESSARY**
- SEARCH FOR RED FLAGS: EYE MOVEMENTS SHOULD BE NORMAL, REFLEXES SHOULD BE NORMAL, STRENGTH SHOULD BE NORMAL, MENTAL STATUS SHOULD BE NORMAL

Clinical Exam findings and associated diagnosis:

- SEVERE assymetric rigidity – CBD
- Early frequent falls – PSP
- Multiple autonomic complaints – MSA
- Symmetric bradykinesia – drug induced, Lewy Body
- Timing of movement and memory issues at or around the same time – Lewy Body Dementia
- Only LOWER body symptoms – vascular
- Dementia and movement at/around the same time – Lewy Body

Textbook differences SUMMARY

Essential tremor

- Bilateral
- WITH action
- Improved with alcohol
- Disease limited to tremor associated disability
- NEGATIVE for other neuro deficits

Parkinson disease

- One sided (72-75%)
- At REST
- Alcohol not beneficial
- Levodopa responsive (94-100% accurate on autopsy)
- Symptom of larger neurological disease: stiff, slow, tremor, gait instability
- Often associated with NONmotor symptoms as well

Both folks will report

- family history of disease (genetic % in both)
- Worse with stress, fatigue (can be worse in both)
- sex/age (not sex linked diseases)
- Changes in handwriting (often complaints in both)

Listen for the primary complaint

- “weakness” – PD
- Food falls off the fork – ET
- Speaking soft – PD
- Voice shaky – either
- Leg cramping – PD
- Drooling – PD
- Can’t turn over in bed – PD
- Trouble getting out of the chair – PD
- Handwriting sloppy – either

I STILL DON'T KNOW

- THAT'S OKAY. EVEN IN MOVEMENT DISORDER CLINICS IT CAN TAKE YEARS FOR A CONFIDENT DIAGNOSIS
- SOME PEOPLE HAVE BOTH ET AND PD, ALSO CALLED A MIXED TREMOR
- Longstanding ET can develop resting component and coordination issues
- DATscan may help, but make sure you have a radiologist who is comfortable reading them and doing QUALitative and QUANTitative analysis in their reading.
- Referral to a Movement Disorder fellowship trained MD or specialized NP/PA – training and experience matters!