How to examine a patient with parkinsonism – tips and tricks

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AN

ESSAY

ON THE

SHAKING PALSY.

CHAPTER I.

DEFINITION—HISTORY—ILLUSTRATIVE CASES.

SHAKING PALSY. (Paralysis Agitans.)

Involuntary tremulous motion, with lessened muscular power, in parts not in action and even when supported; with a propensity to bend the trunk forwards, and to pass from a walking to a running pace: the senses and intellects being uninjured.
How to define a patient with Parkinson’s disease?

• Patient 1
  • 65 years, mild tremor LUE, slightly changed walking with left leg, worsening of handwriting, feels more stiff and clumsy

• Patient 2
  • 54 years, several years of hyposmia, constipation, abnormal movements during sleep, hyperechogenic substantia nigra on USG, no parkinsonism

• Patient 3
  • 47 years, action dystonia left leg, no parkinsonism
  • Later abnormal DaT scan, development of PD

Erro, Ganos, Skorvanek 2014
What defines Parkinson's disease?

• Motor features?

• Other clinical symptoms?

• DaT scan?

• Genetics?

• Presence of pathological synuclein in body fluids / peripheral tissue biopsies?
The first essential criterion is parkinsonism, which is defined as bradykinesia, in combination with at least 1 of rest tremor or rigidity. Examination of all cardinal manifestations should be carried out as described in the MDS–Unified Parkinson Disease Rating Scale.
• 11 studies with pathological confirmation of PD diagnosis

• Non-experts accuracy 73.8%

• Experts accuracy at first visit – 79.6%

• Experts accuracy at follow-up – 83.9%

• UKPDBB criteria accuracy – 82.7%

• Accuracy in community-based study with subsequent pathological confirmation as low as 61.5%

• Most common reasons for misdiagnosis Essential tremor (community studies) and atypical parkinsonism (MSA, PSP) (clinic-based studies)
## Classification of parkinsonian syndromes

<table>
<thead>
<tr>
<th>Category</th>
<th>Percentage</th>
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<tbody>
<tr>
<td><strong>Idiopathic Parkinson’s disease</strong></td>
<td>~80%</td>
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<tr>
<td><strong>Symptomatic secondary parkinsonism</strong></td>
<td>~10%</td>
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<tr>
<td>• Drug-induced</td>
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<tr>
<td>• Toxic – M.Wilson, Mn, CO, MPTP</td>
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<tr>
<td>• Traumatic</td>
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<tr>
<td>• Vascular</td>
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<tr>
<td>• Normal pressure hydrocephalus</td>
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<tr>
<td><strong>Other neurodegenerative parkinsonisms</strong></td>
<td>~10%</td>
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<tr>
<td>• Multiple system atrophy</td>
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<tr>
<td>• Progressive supranuclear palsy</td>
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<tr>
<td>• Corticobasal degeneration</td>
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<td>• Lewy body dementia</td>
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<td>• Huntington’s disease, etc.</td>
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</table>
Pathological classification of synucleinopathies

<table>
<thead>
<tr>
<th>Synucleinopathies</th>
<th>Tauopathies</th>
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<tbody>
<tr>
<td>• Parkinson’s disease (+dementia)</td>
<td>• Progressive supranuclear palsy</td>
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<tr>
<td>• Lewy body dementia</td>
<td>• Corticobasal degeneration</td>
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<tr>
<td>• Multiple system atrophy</td>
<td>• Frontotemporal lobar degeneration</td>
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Slowness of movements

• Bradykinesia
• Paresis
• Dyssynergy related to cerebellar syndrome
• Apraxia/executive dysfunction
• „True bradykinesia“ vs. other cause of slowness
Increased muscle tone

• Rigidity
• Spasticity
• Interposition of tremor
Tremor

• Resting vs. action tremor
• Asymmetrical vs. Symmetrical
• Where?
  • Head – more likely ET or dystonic!
  • Chin, lips, tongue – PD more likely
• Postural vs. Reemergent - transient tremor supression
• Myoclonus (more likely multiple system atrophy, corticobasal degeneration)
<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Frequency</th>
<th>Activation by</th>
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<tr>
<td></td>
<td></td>
<td>goal-directed</td>
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<td></td>
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<td>movement</td>
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<td>Physiologic tremor</td>
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<td>Enhanced physiologic tremor</td>
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<tr>
<td>Essential tremor syndromes</td>
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<td>Classic essential tremor</td>
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<td>Undetermined tremor syndrome</td>
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<td>Orthostatic tremor</td>
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<td>Task- and position-specific tremors</td>
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<tr>
<td>Dystonic tremor</td>
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<td>Parkinsonian tremor</td>
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<tr>
<td>Cerebellar tremor</td>
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<td>Holmes tremor</td>
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<td>Palatal tremor</td>
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<tr>
<td>Palatal tremor</td>
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<tr>
<td>Neuropathic tremor syndrome</td>
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<tr>
<td>Drug-induced and toxic tremors</td>
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<td>Psychogenic tremor</td>
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Deuschl 1998
Motor features

Responsive
• Bradykinesia
• Rigidity
• Tremor
• Off freezing
• Off dystonia

Poorly responsive
• Postural instability
• Speech disorders
• Dysphagia
• On freezing
• On dystonia
TABLE 1. MDS Clinical Diagnostic Criteria for PD—Executive Summary/Completion Form

The first essential criterion is parkinsonism, which is defined as bradykinesia, in combination with at least 1 of rest tremor or rigidity. Examination of all cardinal manifestations should be carried out as described in the MDS–Unified Parkinson Disease Rating Scale.\textsuperscript{30} Once parkinsonism has been diagnosed:

Diagnosis of Clinically Established PD requires:
1. Absence of absolute exclusion criteria
2. At least two supportive criteria, and
3. No red flags

Diagnosis of Clinically Probable PD requires:
1. Absence of absolute exclusion criteria
2. Presence of red flags counterbalanced by supportive criteria
   - If 1 red flag is present, there must also be at least 1 supportive criterion
   - If 2 red flags, at least 2 supportive criteria are needed
   - No more than 2 red flags are allowed for this category
Supportive criteria

- Clear and dramatic beneficial response to dopaminergic therapy
  jednoznačné a dramatické zlepšenie po dopaminergnej liečbe.

- Presence of L-dopa induced dyskinesia

- Rest tremor of a limb documented on clinical examination

- Presence of either olfactory loss or cardiac sympathetic denervation on MIBG SPECT
Typical course of PD

Early stage
- Good response to dopaminergic treatment

Wearing-off
- short off periods

Wearing-off with dyskinesias

On-off fluctuations
- Unpredictable fluctuations
- Very narrow therapeutic window

Normal mobility
Parkinsonism
Dyskinesias

Therapeutic window

Time (years)

~ 0-4
~ 4-7
~ 7-10
~ >10
Imaging - MRI

- Standard MRI finding in PD normal “age-related”
- Done to exclude other disorders
Imaging - MRI

- Standard MRI finding in PD normal “age-related”
- Done rather to exclude other disorders
Imaging – DaT scan

• Does not differentiate PD from other forms of neurodegenerative parkinsonism!

• Differentiates
  • Drug-induced parkinsonism
  • Essential tremor
  • Dystonic tremor
  • Functional parkinsonism
  • Lewy body dementia vs. Alzheimer’s disease

• SWEDD – scans without evidence of dopaminergic deficit
Imaging – MIBG + IBZM SPECT

• To differentiate PD vs. atypical parkinsonism

• MIBG SPECT – cardial sympathetic denervation in PD, not in MSA
  • PD – pathological (usually dramatic) reduction of radiotracer uptake
  • MSA, PSP normal radiotracer uptake

• IBZM SPECT – examination of postsynaptic D2 receptors
  • Normal uptake in PD (only presynaptic deficits)
  • Reduced uptake in atypical parkinsonism
Therapeutic test

• L-dopa challenge (single dose administration) – not reliable
  • Positive might support PD
  • Negative (single dose administration) doesn’t mean anything

• Apomorphine – unreliable similar to L-dopa challenge

• Necessary to administrate high doses for longer time (~1000mg for at least 1 month, if therapeutic benefit not observed on lower doses)

• Usually no/minimal response to chronic L-dopa administration in atypical parkinsonism (MSA may be an exception in first few years)
Conclusions

• Parkinson’s disease has many faces

• However, some faces are very atypical and these need to be distinguished...
2nd Kosice Course of Movement Disorders
17. - 19. 5. 2018, Double Tree by Hilton, Košice

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