Movement Disorders and Cognitive Decline

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DISCLOSURES

• None relevant to this talk

FINANCIAL CONFLICTS OF INTEREST

• None relevant to this talk
OUTLINE

• Introduction

• Neuroanatomic and neuropathology substrate/link between cognition & movement disorders

• DDx of movement disorders with “prominent” cognitive phenomenon

• Cognitive decline in parkinsonism
Early descriptions of MDs

“…..the sense and intellect being uninjured’

Unlike PD, Earliest description of HD included cognitive dysfunction
cognition ≠ memory. And when talking about cognitive decline, all DOMAINS of cognition must be interrogated and evaluated.

Why cognitive decline in movement disorders?

- Substantially adds to disease burden
- Negatively impacts QoL
- Contributes to caregiver burden
- Major challenge in management
- One of the unmet needs
Interplay Between Cognition & Movement

- Striatum - key contributor in the intersection of motor function, cognition, and emotion.

- Cognitive processes control movement including;
  - Plans for movement
  - Motor learning
  - Habit-formation
  - Goal-directed learning
  - Detection of errors

Movement disorders + *prominent* cognitive profile

<table>
<thead>
<tr>
<th>Common</th>
<th>Less Common</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Parkinson’s disease</td>
<td>• Chorea-acanthocytosis</td>
</tr>
<tr>
<td>• Atypical parkinsonism</td>
<td>• HDL-2</td>
</tr>
<tr>
<td>• DLB</td>
<td>• NBIA (PKAN)</td>
</tr>
<tr>
<td>• PSP</td>
<td>• DRPLA</td>
</tr>
<tr>
<td>• CBD</td>
<td>• Wilson’s disease</td>
</tr>
<tr>
<td>• MSA</td>
<td>• Aceruloplasminaemia</td>
</tr>
<tr>
<td>• Huntington’s disease</td>
<td>• Niemann-Pick disease</td>
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<td></td>
<td>• FXTAS</td>
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</tbody>
</table>
## Profile of Cognitive Dysfunction in Movement Disorders

<table>
<thead>
<tr>
<th>Domain</th>
<th>PD</th>
<th>DLB</th>
<th>CBD</th>
<th>PSP</th>
<th>HD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Memory</td>
<td>+</td>
<td>++</td>
<td></td>
<td></td>
<td>++</td>
</tr>
<tr>
<td>Language</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Visuospatial function</td>
<td>++</td>
<td>+++</td>
<td></td>
<td>+</td>
<td>++</td>
</tr>
<tr>
<td>Attention</td>
<td></td>
<td>+++</td>
<td></td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Apraxia</td>
<td>++</td>
<td>+++</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Executive function</td>
<td>+++</td>
<td>+++</td>
<td>++</td>
<td>++++</td>
<td>+++</td>
</tr>
<tr>
<td>Social cognition</td>
<td></td>
<td></td>
<td>++</td>
<td>+</td>
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</tbody>
</table>
Subjective cognitive dysfunction: presence of cognitive complaints in pts with objective normal cognitive performance on standardized neuropsychological evaluation/tests & preserved ability to carry out their ADLs. Important because it predicts future progression to PD-MCI or PDD.
**Diagnostic Criteria PD MCI**

**Inclusion Criteria**

- Diagnosis of PD as based on the UK PD Brain Bank Criteria
- Gradual decline, in the context of established PD, in cognitive ability (patient or informant-reported, or observed by clinician)
- Cognitive deficits on either formal neuropsychological testing or a scale of global cognitive abilities
- Cognitive deficits are *not sufficient to interfere significantly with functional independence*, although subtle difficulties on complex functional tasks may be present

**Exclusion criteria**

- Diagnosis of PD dementia based on MDS Task Force proposed criteria
- Other explanations for cognitive impairment
- Other PD-associated comorbid conditions that, in the opinion of the clinician, significantly influence cognitive testing (e.g., motor impairment or severe anxiety, depression, excessive daytime sleepiness, or psychosis)
Diagnostic Criteria PDD

I. Core features
• Diagnosis of PD according to Queen Square BBC
• Dementia syndrome developing within the context of established Parkinson’s

II. Associated features
• Cognitive: at least 2 of 4 domains
• Behavioural:

Probable PD-D
A. Core features: Both must be present
B. Associated clinical features:
• Typical profile of cognitive deficits including impairment in at least 2 of the 4 core cognitive domains (attention which may fluctuate, executive functions, visuospatial functions, and memory which usually improves with cueing)
• The presence of at least one behavioral symptom (apathy, depression or anxiety, hallucinations, delusions, EDS) supportive; lack of behavioral symptoms, however, does not exclude the diagnosis

None of the group III features OR group IV features present

Possible PD-D
A. Core features: Both must be present
B. Associated clinical features:
• Atypical profile of cognitive impairment in one or more domains, such as prominent or receptive-type (fluent) aphasia, or pure storage-failure type amnesia (memory does not improve with cueing or in recognition tasks) with preserved attention
• Behavioral symptoms may or may not be present OR 1 or > group II features

None of Group IV
# Neuropsychological Tests

<table>
<thead>
<tr>
<th>Global Cognition</th>
<th>Executive function</th>
<th>Attention &amp; Working memory</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Montreal Cognitive Assessment (MOCA)</td>
<td>• Clock drawing test</td>
<td>• Digit span (Forward &amp; backward)</td>
</tr>
<tr>
<td>• Frontal Assessment Battery</td>
<td>• Symbol Digit Modalities Test</td>
<td>• Trail-making Test</td>
</tr>
<tr>
<td>• Mattis Dementia Rating Scale (MDRS)</td>
<td>• Semantic verbal fluency</td>
<td>• Stroop test</td>
</tr>
<tr>
<td>• Mini-mental State Examination (MMSE)</td>
<td>• Phonemic verbal fluency (F, A, S)</td>
<td></td>
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<tr>
<td></td>
<td>• Wisconsin CST</td>
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</tbody>
</table>

Memory – HVLTs; Visuospatial – Benton JLO; Language – Boston NT, MINT
Global Scales for Cognitive Screening in Parkinson’s Disease: Critique and Recommendations

Matej Skovranek, MD, PhD,1,2* Jennifer G. Goldman, MD, MS3, Marjan Jahanshahi, PhD,4 Connie Marras, MD, PhD,5 Irena Rektorova, MD, PhD,6 Ben Schmand, PhD,7 Erik van Duijn, MD, PhD,8 Christopher G. Goetz, MD,3 Daniel Weintraub, MD,9 Glenn T. Stebbins, PhD,3 Pablo Martinez-Martín, MD, PhD,10 and the members of the MDS Rating Scales Review Committee

• “Recommended”: Montreal Cognitive Assessment, Mattis Dementia Rating Scale 2nd ed, Parkinson’s Disease-Cognitive Rating Scale

• “Recommended with caveats”: the Mini-Mental Parkinson, Scales for Outcomes in Parkinson’s Disease-Cognition

• “Suggested” or “Listed”: MMSE, ADAS-Cog, PANDA, CAMCOG-R, PDD-SS, RBANS

Visit
www.mocatest.org/training-certification
IDEA COGNITION SCREENING INSTRUMENT

Please note the total time taken to complete the entire questionnaire (including the preparation for Question 5)

STEP 1: Preparation for ten-word list item (Question 5)

Interviewer instructions:
- Read out the instructions to the participant.
- There are 3 attempts in all.
- For each attempt, let the participant know that you will require him/her to repeat the words after you.
- Tick the words the participant remembers on the grid at each attempt.
- After completing all attempts, move to Step 2 (completion of the IDEA instrument).

Interviewer to participant: I am going to read out a list of words. Please listen carefully and I will ask you to repeat them back to me once I have finished (read out the words slowly).
1. First attempt: Now tell me all the words you can remember (Tick the words remembered).
2. Second attempt: Now I will read out the words again. Listen carefully and I will ask you to repeat as many as you can (Read out the words slowly). Now tell me all the words you can remember (Tick the words remembered).
3. Third attempt: Now I will read out the words one last time. Listen carefully and I will ask you to repeat as many as you can (Read out the words slowly). Now tell me all the words you can remember (Tick the words remembered).

<table>
<thead>
<tr>
<th>Vernacular word list</th>
<th>First attempt</th>
<th>Second attempt</th>
<th>Third attempt</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Butter</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 Arm</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3 Letter</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4 Queen</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5 Ticket</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6 Grass</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7 Corner</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>8 Stone</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9 Book</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>10 Stick</td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

IDEA SCREENING INSTRUMENT QUESTIONS

<table>
<thead>
<tr>
<th>ITEM</th>
<th>INSTRUCTION</th>
<th>SCORE</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 I will tell you the name of something and I want you to describe what it is. What is a bridge? (Correct answer: something that goes across a river, canyon or road) 0 if incorrect, 2 if correct.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 I want you to name as many different animals as you can in one minute. Number of animals named. 0 for 0-3 animals named, 1 for 4-7 animals named, 2 for 8 or more animals named.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3 Who is the chief/head/leader of this village? 0 if incorrect, 1 if correct.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4 What day of the week is it? 0 if incorrect, 2 if correct.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5 Can you tell me the ten words we learned earlier? Try to remember as many as you can. 0 for no words remembered, 1 for 1 word, 2 for 2 words, 3 for 3 words, 4 for 4 words, 5 for 5 or more words.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6 Can you make the design shown below using these four matchsticks. I will show you once and then you have to copy exactly. Score 1 for each part of the design that is performed correctly. 1 Middle two matchstick heads pointing in the same way, 1 Outside two matchsticks pointing at an angle, 1 Matchstick heads are oriented correctly.</td>
<td></td>
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</tr>
</tbody>
</table>

TOTAL SCORE/15

Interviewer Date:

Approximate time taken to complete the test (including preparation for Q 5) Minutes: 
Cognitive dysfunction is common in PD

• Pooled PD MCI prevalence: 40% (n=7053 pwPD) (95% CI 36–44)
• Point prevalence of PDD: 30%
  • After 5 years: 15 – 20%; After 10 years: 46%; At 20 years: 80%
• Incidence of PDD: 24.3/1000 per year
• Risk of dementia 5 – 6x normal aging population
• Moderated by stage of PD

• Associations (PD MCI): *older age, lower education, longer PD duration, higher LEDD, more severe motor symptoms, PIGD motor subtype, poorer quality of life, higher apathy, depression.*

• Associations (PDD): *older age, more severe parkinsonism (particular rigidity, PIGD), MCI at baseline*

Take-home points

• Movement disorders are heterogenous syndromes reflecting degeneration in multiple circuits & neurotransmitters systems

• Neurodegenerative movement disorders are not just motor syndromes

• Cognitive dysfunction accompanies parkinsonism – PD, DLB and common in PD where it predicts QoL

• Testing at initial presentation and periodically important

• Choice of screening and neuropsychological tests important (consider level of education; is the test culturally appropriate?)
References/Resources


• Special Issue: Cognition and Movement Disorders. Mov. Disord. 2014; 29(5)


References/Resources


