

Updates on the neuropsychological evaluation of movement disorders

Elizabeth Ann Ćurko Kera

Department of Neurology, Hackensack University Medical Center, 650 From Rd, Paramus, NJ 07652, USA
Hackensack Meridian School of Medicine, Nutley, NJ 07110

Abstract: Movement disorders are amongst the most common forms of neurodegenerative diseases across the globe. However, there is significant variability among the cognitive and behavioral presentation of typical and atypical movement disorders. Prevalence rates are discussed along with the relevance of each disorder to clinical outcome. The importance of a tailored, yet thorough neuropsychological evaluation relevant to the differential diagnostic considerations is reviewed, including considerations during the clinical interview. The utility of neuropsychological evaluation in guiding appropriate treatment recommendations and improving quality of life are likewise discussed.

Keywords: neurodegenerative diseases, movement disorders, neuropsychological evaluation, clinical psychology

Movement disorders are amongst the most common neurodegenerative diseases across the globe, though there is considerable variability in prevalence rates among each type of disorder. Parkinson's disease (PD) is estimated to affect 8.5 million people worldwide (O'Shea & Shih, 2023), while atypical forms of Parkinsonism such as Progressive Supranuclear Palsy (PSP) and Corticobasal Syndrome (CBS) appear at a lower frequency worldwide (7.1 and 2.3 per 100,000 people worldwide, respectively; Swallow et al., 2022). Essential tremor (ET) is thought to be the most common movement disorder with a prevalence rate of more than 1.33% (i.e.,

1,330 per 100,000 people) worldwide (Louis & McCreary, 2021).

As recently as 20 years ago, the categorizations of movement disorders were much more simplistic. While diagnoses of PD, dystonia, tic disorders, PSP, and ET were identifiable previously, a lot of research has shed light on the specific delineations of physical and cognitive symptoms which differ not just between these classes of disorders, but also amongst the subvariants of each disorder. To date, for example, several subvariants have been identified in PSP, the most common form of atypical Parkinsonism (Krzosek et al., 2022; Respondek & Hoglinger, 2022). There

is considerable variability in the presentation of Lewy Body Dementia (LBD), with the classification of the diagnosis being revised as recently as 2017 (McKeith et al., 2017) and in 2020 when considering research applications (McKeith et al., 2020). These updates have not only led to increased knowledge about the disease itself, but also to updates in diagnostic labels. For example, ET was previously thought to be “benign”, a descriptor that was previously reflected in diagnostic terminology (i.e., “benign essential tremor”). However, in recent decades it has been shown that ET is associated with a higher risk of cognitive dysfunction, which in turn is associated with a greater risk of falls/imbalance (Louis et al., 2017). Additionally, an ET diagnosis has been shown to place individuals at a higher risk of developing neurocognitive deficits associated with both cortical and subcortical dementia (Berry et al., 2023). Moreover, individuals with ET can “cross over” and display symptoms suggestive of other disorders, such as Parkinsonian syndromes (Yoo et al., 2023). For this reason, the word “benign” was appropriately removed from diagnostic terminology given the potential implications of ET symptoms on an individual’s functioning and prognosis. In clinical practice, it is crucial to stay current with the changing landscape of key clinical symptoms associated with and pathognomonic of each neurodegenerative condition. The subtle differences in key features of clinical symptoms and cognitive profiles can lead to marked differences in treatment outcomes and the quality of life for individuals.

In neuropsychological assessment, this process often begins with a clinical interview that comprehensively addresses the concerns of patients. The thorough review of medical, developmental, and psychosocial history is imperative. The focus of the clinician should be on understanding ALL factors that contrib-

ute to an individual’s areas of concern. For example, it is important to consider the impact of any premorbid learning issues and educational attainment on an individual’s functioning. Identifying contributory medical factors such as issues with sleep, cardiovascular risk factors, urinary/bowel functioning, autonomic dysfunction, substance use/abuse history, or previous head injuries is crucial. The impact of psychological well-being and mental health should be considered, and along with this, the individual’s current psychosocial support network should be assessed.

Within the context of these contributory factors, assessing the timeline (onset of concerns from present day of evaluation) and the nature of symptoms (have they been intermittent, stable, or progressive from the time of onset) provides extensive information to the clinician. Lastly, obtaining specific information related to the specific cognitive concerns is key. While many patients attribute any cognitive change to a “memory issue”, often the concern is determined to be an issue with a cognitive domain that is separate from memory, despite the fact that these issues can have an impact on memory (Ferguson & Foley, 2024). Bradyphrenia, for example, or the slowed processing of information can certainly impact what we are capable of encoding to memory. Thus, an individual will likely not remember information if it was never committed to memory. However, if individuals are provided with enough time to process information, they may then, in fact, be able to remember the information that was previously encoded. This would be an indication that memory is intact, while highlighting processing speed as the main cognitive concern. Other times, an individual may indicate memory concerns, but when pressed will state that it is typically only an issue of remembering words or names, indicating a potential dysnomia or anomia rather than an amnesic profile.

Once the main concerns have been established, a tailored approach to the assessment of movement disorders is crucial. Many of the individuals in question are at risk for challenges with cognitive endurance to task due to multiple factors including fatigue, medication timing effects (on/off states), bradyphrenia, and challenges in arousal. Information gleaned from the clinical interview in addition to medical records (e.g., brain imaging, etc.) can assist the clinician in choosing an initial battery that serves as a basis for “screening” each cognitive domain, while still obtaining meaningful results within allowed time constraints with the patient. The Repeatable Battery for the Assessment of Neuropsychological Functions (Randolph et al., 1998) is a suitable battery for such screening in many cases as it includes the evaluation of immediate and delayed memory through multiple modalities (list learning, story, figure recall), language (naming, semantic fluency), visuospatial skills (figure copy, judgment of line orientation), and attention and processing speed (digit span, coding). Other executive measures are crucial additions to a movement disorder battery given the prevalence of executive dysfunction across virtually all forms of movement disorders (Dirnberger & Jahanshahi, 2013; Raimo et al., 2023; Dogan et al., 2024; Bermejo-Pareja & Puertas-Martín, 2012). Thus, the thorough assessment of cognitive set shifting (Trail Making Test A/B (Reitan, 1956) or Color Trails Test (D’Elia et al., 1996)), problem solving (Wisconsin Card Sort Task (Grant & Berg, 1948) or the Tower Task (Delis et al., 2001)), and inhibition (Stroop (Golden et al., 2021), Go/No-Go) are standard additions to the assessment. Motor tasks that assess not only graphomotor dysfunction but also motor planning (e.g., Luria motor sequences) are key components of a movement disorder battery given the sensitivity of motor planning tasks to fronto-subcortical dysfunction.

The specific cognitive profiles of typical and atypical Parkinsonism have been well-documented in literature (Santangelo et al., 2018). While bradyphrenia and executive dysfunction, for example, are core features in many movement disorders, including PD, PSP, LBD, and Huntington’s disease (HD), the severity of such dysfunction can differ early on in the disease course (Gerstenecker, 2017). Visuospatial deficits are often more pronounced in HD and LBD (Stucky et al., 2014). While PD and PSP often show impairment in complex attention, simple attention may be spared early on in these conditions, in contrast to simple attentional deficiencies initially evident in HD or LBD (Stucky et al., 2014). Moreover, PSP patients have been found to have more globally depressed scores in phonemic fluency and abstraction when compared to PD and MSA, whereas LBD patients exhibit more challenges with semantic fluency and visuospatial memory than other typical and atypical PD profiles, potentially due to the greater proportion of atrophy of posterior cortical regions (Raimo et al., 2023). Subtle differences within domains can also exist such as differences in performance on phonemic fluency, cognitive set-shifting (TMT-B), and WCST errors between PSP and PD patients (PSP>PD impairment, Gerstenecker, 2017). Therefore, thorough evaluation within cognitive domains is also crucial in differential diagnosis. Lastly, the evaluation of neuropsychiatric and sensorimotor features such as hallucinations (LBD), psychosis (HD), alien limb phenomenon (CBD), or applause sign and downward gaze palsy (PSP) (Stucky et al., 2014) is also informative.

Once a cognitive profile has been obtained, the utility of the evaluation results in such cases is multifold, and thus the referral reason for such an evaluation should be considered. Data obtained from neuropsychological evaluations can be diagnostic, quali-

tative/descriptive or both. In other words, in some instances, the information obtained in the neuropsychological assessment will be used in helping to establish a particular profile pathognomonic of a particular disorder. Once a diagnosis has been established, specific recommendations can be made that are tailored to the individual case. For example, the effectiveness of dopaminergic agonists in PD vs LBD is quite different (Drach, 2011). In other cases, the diagnosis may already be established, but due to inevitable changes in the individual's functioning, the clinical profile may be quite helpful in determining appropriate clinical recommendations.

One specific type of behavioral intervention that can be recommended is cognitive remediation, a common behavioral intervention that uses learning principles to improve specific cognitive abilities or daily living skills. It has been shown effective in treating the non-motor symptoms (i.e., cognitive deficits) typical of Parkinson's disease (Alzahrani & Veneri, 2018). However, the specific cognitive profile will determine the course of treatment and the goals in a cognitive remediation session. While, for example, a specific cognitive remediation protocol may be appropriate for an individual with dysexecutive or processing speed challenges, it may not be effective at all for a patient with a severely amnesic profile since challenges with memory will almost undoubtedly lead to difficulties in the acquisition of strategies or in carrying over new learning. Individuals may have exacerbated dysexecutive symptoms due to depression, leading to a recommendation for counseling or medication management for such symptoms. Speech/language therapy has been shown to be quite effective for patients with Parkinson's disease, particularly when provided through the modality of LSVT training to improve voice clarity, volume, and articulation (Pu et al., 2021). Accommodations can also be suggested for in-

dividuals based on their unique cognitive profile to optimize their performance at school, work, or home (e.g., flexible schedule to accommodate for medication timing).

Another possible consideration in choosing appropriate tests may be whether the individual is planning on undergoing neurosurgical treatment of said movement disorder. Increasingly, patients are choosing options such as high intensity focused ultrasound and deep brain stimulation to treat tremor when medication becomes less effective and/or causes unpleasant side effects. In such cases, evaluating an individual's comprehension skills (are they understanding the procedure and medical risks/benefits), judgment/decision-making ability (can they effectively synthesize all of this information to make an educated decision), and memory (can we rule out secondary progressive conditions such as a cortical dementia which would impact their ability to fully participate in such a decision) is essential. Additionally, ruling out a secondary progressive cortical condition has important implications for making predictions about prognosis after surgical treatment and serves to further improve sensitivity and specificity of candidate selection. While some clinicians are tempted to perform extremely comprehensive batteries in such cases, it is important to remember that going on extensive fishing expeditions will often lead to at least one "catch of the day." That is, even in individuals from normative populations without neurological or cognitive concerns, if enough tests are administered, eventually some relative weakness will be found in an individual's profile. It behooves the clinician to be mindful of the fact that the aim is to rule out larger cognitive issues that preclude the patient's candidacy for surgical intervention rather than finding a candidate with a normative cognitive profile. By virtue of the fact that most individuals considering neurosurgical intervention have

been diagnosed with a particular movement disorder for some time (i.e., neurosurgical procedures are not typically the first line of treatment for most individuals but are rather considered after multiple medication options have failed to remain effective), they will likely experience some change to their cognitive profile as well. Most commonly, issues with slowed processing and executive functioning are present, and likewise can impact other aspects of functioning, influencing verbal fluency and memory (Amunts et al., 2020; Saikia & Tripathi, 2024). A skilled clinician should be able to tease apart these distinguishing characteristics of a patient's profile. Other aspects of cognitive assessment, such as intelligence testing, are not a distinguishing factor in such cases. Individuals of higher or lower intellectual capabilities should have the same access to such treatment options and intra-individual change will be particularly important to assess in such cases. The same holds true for the assessment of such skills in cortical dementias such as Alzheimer's disease. One can be of higher or lower intelligence while still exhibiting signs of cortical degeneration, so intelligence is not the key feature that will help to distinguish the diagnosis or appropriateness of clinical treatment options in such cases.

Once the evaluation process is complete, the recommendations of the clinician will only be useful if the individual/patient in question fully understands them and their utility/benefits. Delineating the specific intricacies of abstract cognitive skills is likely to be overwhelming to the lay public (Gruters et al., 2022; Baum et al., 2018). Identifying a few key areas of weakness/deficit and explaining the nature of them and their impact on the patient's life is often much more helpful as a take-away message than going through every aspect of the individual's scores. Moreover, explaining why certain recommendations could be beneficial will likely have a much higher

impact on the individual's likelihood to follow through on that recommendation than providing them with just a summary of results. While clinical treatment options are the gold standard, at times individuals could benefit from additional integrative treatment options as well for multiple reasons (Bloem et al., 2020). First, clinical treatment options are often not as available in one's geographic area (given the level of expertise, training, and equipment needed for such treatments) as are integrative options in one's community. Second, the cost of such clinical treatment options may be too great for the individual to bear for prolonged periods of time. Third, the implementation of skills acquired in clinical treatment in integrative care settings can be carried over into different settings in addition to increasing the frequency of utilization. Lastly, family members and friends often can be included in such integrative care settings more easily, again increasing the probability that the carrying over of skills can be supported by others in an individual patient's daily environment. Programs such as PingPongParkinson, Rock Steady Boxing, and Dance for PD are ideal examples of integrative care programs that not only complement traditional treatment approaches, but incorporate activities that appeal to a larger age range of affected individuals.

As the diagnostic classifications and features of movement disorders become more complex, the same applies to the considerations that are key to a useful neuropsychological evaluation and its applications. There is much room for improvement in the understanding of cognitive similarities and differences of each distinct disorder as well as within the subvariants of a particular disorder. It will be crucial for neuropsychologists to not only stay abreast of such changes in the field, but also to be a driving force in furthering the understanding of the cognitive implications of these disorders and their impact on a patient's quality of life.

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Novosti u neuropsihološkoj procjeni poremećaja kretanja

Sažetak: Poremećaji kretanja među najčešćim su oblicima neurodegenerativnih bolesti diljem svijeta. Međutim, postoji značajna varijabilnost u kognitivnom i bihevioralnom prikazu tipičnih i atipičnih poremećaja kretanja. U radu se raspravlja o stopama prevalencije zajedno sa značajnosti svakog poremećaja za klinički ishod. Prikazuje se važnost prilagođene, ali temeljite neuropsihološke evaluacije relevantne za diferencijalno-dijagnostička razmatranja, uključujući razmatranja tijekom kliničkog intervjua. Također, raspravlja se o korisnosti neuropsihološke procjene u davanju odgovarajućih preporuka za liječenje i poboljšanju kvalitete života.

Ključne riječi: neurovegetativne bolesti, poremećaji pokreta, neuropsihologijska evaluacija, klinička psihologija

Korespondencija: Elizabeth Ann Ćurko Kera
elizabeth.kera@hmhn.org

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