Neuropsychological Assessment and Differential Diagnosis in Young-Onset Dementias

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KEYWORDS

• Alzheimer's disease • Frontotemporal dementia • Primary progressive aphasia
• Progressive supranuclear palsy • Posterior cortical atrophy

KEY POINTS

- Most young-onset dementia syndromes belong to the spectrum of early-onset Alzheimer's disease or frontotemporal dementia.
- Patients with young-onset dementia usually present with early behavior, executive, or language changes.
- Episodic memory impairment is rarely seen at onset.
- Comprehensive neuropsychological assessment is crucial to the young-onset dementia diagnosis.

YOUNG-ONSET DEMENTIAS—THE ROLE OF NEUROPSYCHOLOGY

Young-onset dementias (YODs) are a heterogeneous group of disorders comprising mainly early-onset adult primary neurodegenerative diseases (in contrast to late onset), late-onset forms of childhood neurodegenerative conditions (eg, mitochondrial disorders), vascular dementia (VaD), various dementia syndromes with potentially reversible etiologies (eg, autoimmune, infectious diseases), and dementias related to substance abuse. The diagnosis of YOD is usually based on history (with the crucial

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Abbreviations

ACE-III	Addenbrooke's Cognitive Examination-III
AD	Alzheimer's disease
bvFTD	Behavioral variant of frontotemporal dementia
CBS	Corticobasal syndrome
DLB	Dementia with Lewy bodies
DRS-2	Mattis Dementia Rating Scale-2
ECAS	Edinburgh Cognitive and Behavioral ALS Screen
EOAD	Early-onset Alzheimer's disease
FTD	Frontotemporal dementia
HD	Huntington's disease
lvPPA	Logopenic variant of primary progressive aphasia
MMSE	Mini-Mental State Examination
nfvPPA	Nonfluent variant of primary progressive aphasia
PCA	Posterior cortical atrophy
PPA	Primary progressive aphasia
PSP	Progressive supranuclear palsy
svPPA	Semantic variant of primary progressive aphasia
VaD	Vascular dementia
VOSP	Visual Object and Space Perception Battery
YOAD	Young-onset Alzheimer's disease (also known as EOAD, early-onset AD)
YOD	Young-onset dementia

input from the informant), neuropsychiatric, cognitive and motor examination, neuroimaging, and laboratory findings, although for some diseases, genetic testing is becoming increasingly useful in the clinical practice.[1] The diagnosis of other disorders, such as normal-pressure hydrocephalus or mitochondrial disorder, is based mainly on physical, imaging, and laboratory findings. Thus, in this review, these disorders are not discussed in detail.

Neuropsychological assessment in YOD may serve several purposes:

1. Differentiating subjective cognitive complaints from cognitive impairment
2. Determining the presence and severity of cognitive impairment
3. Determining the impact of emotional factors (eg, anxiety, depressed mood) on cognitive performance
4. Determining the patient's cognitive profile to contribute to the differential diagnosis of dementia

Aims 1 through 3 have been extensively discussed in the literature[2] and exceed the scope of this review, which focuses on the role of neuropsychology in delineating the patient's cognitive profile. It is well documented that the integration of prospective clinical and neuropsychological data (eg, "anterior" vs "posterior" cognitive profile) associated with time course of illness is highly predictive of pathology on autopsy examination.[3]

For the sake of brevity, some specific aspects of neuropsychological examination, such as the assessment of reading, writing and calculation, were omitted in this review.

NEUROPSYCHOLOGICAL SCREENING ASSESSMENT IN YOUNG-ONSET DEMENTIA

Because YOD refers to individuals younger than 65, the cognitive screening tools aimed at this population need to be more sensitive to subtle cognitive alterations than coarse-grained measures, such as Mini-Mental State Examination (MMSE).[4]