

5th Congress of the European Academy of Neurology Oslo, Norway, June 29 - July 2, 2019

Teaching Course 18

Testing of cognitive functions by the neurologist (Level 1)

Clinical dementia testing: methods and meaning

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Testing of cognitive functions by the neurologist (Level 1) Clinical Dementia testing: methods and meaning

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Clinical Dementia testing: methods and meaning

Nothing to disclose



Dementia is diagnosed when there are cognitive or behavioral (neuropsychiatric) symptoms that:

- 1. Interfere with the ability to function at usual activities;
- 2. Represent a decline from previous levels of functioning and performing;
 - 3. Are not explained by delirium or major psychiatric disorder;
- 4. Cognitive impairment is detected and diagnosed through a combination of (1) history-taking from the patient and a knowledgeable informant and (2) an objective cognitive assessment
 - 5. The cognitive or behavioral impairment involves a minimum of two of the following domains:
- **5.1.** Impaired ability to acquire and **remember** new information—symptoms include: repetitive questions or conversations, misplacing personal belongings, forgetting events or appointments, getting lost on a familiar route.
- 5.2. Impaired reasoning and handling of complex tasks, poor judgment— symptoms include: poor understanding of safety risks, inability to manage finances, poor decision-making ability, inability to plan complex or sequential activities.
- 5.3. Impaired visuospatial abilities—symptoms include: inability to recognize faces or common objects or to find objects in direct view despite good acuity, inability to operate simple spatial organisation, or orient clothing to the body.
- **5.4.** Impaired **language** functions (speaking, reading, writing)—symptoms include: difficulty thinking of common words while speaking, hesitations; speech, spelling, and writing errors.
- 5.5. Changes in personality, behavior—symptoms include: uncharacteristic mood fluctuations such as agitation, impaired motivation, initiative, apathy, loss of drive, social withdrawal, decreased interest in previous activities, loss of empathy, compulsive or obsessive behaviors, socially unacceptable behaviors.

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Memory is the most frequent complaint Different memory systems

Procedural memory (ex: biking)
Semantic memory (ex: second war)
Short term/working memory (ex: code for e-banking)
Episodic memory (ex: yesterday breakfast)

Robust versus fragile systems



Interview: Subjective Cognitive Decline

Do you complain about your cognition?
Do you worry about it?
Did you have a good cognition before?
Since when do you complain?
How do you behave compare to others?

Ask for daily examples of instrumental activities (IADL: administrative tasks, medication, transportation, phone)

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Interview: the judgment of a relative

Do you have other examples of cognitive difficuties?

Did you observe a change in behaviour?

The difference between patient's and relative's report allows to assess anosognosia for cognitive impairment



Clinical syndromes

Subjective Cognitive Decline

Mild Cognitive Impairment (objective cognitive decline with few repercussions on daily activities)

Dementia (objective cognitive decline with repercussions on daily activities)

There is a need to specify the underlying pathology to provide adapted recommendations

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Clinical Dementia testing: methods and meaning

Clinical Dementia testing

There a multiple short batteries to test cognition or daily activities (we made choices)



Mini Mental State Exam

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	ONE POINT FOR EACH ANSWER	}
ORIENT	ATION Year Month Day Date Time	/5
	real Month Day Date Time	/3
	Country Town District Hospital Ward	/5
REGISTI	RATION	
	Examiner names 3 objects (eg apple, table, penny) Patient asked to repeat (1 point for each correct).	
	THEN patient to learn the 3 names repeating until	
	соггест.	/3
ATTENT	TON AND CALCULATION Subtract 7 from 100, then repeat from result.	
	Continue 5 times: 100 93 86 79 65	/5
	Alternative: spell "WORLD" backwards - dlrow.	
RECALI		/2
	Ask for names of 3 objects learned earlier.	
LANGU		
	Name a pencil and watch.	
	Repeat "No ifs, ands, or buts".	/1
	Give a 3 stage command. Score 1 for each stage.	
	Eg. "Place index finger of right hand on your nose and then on your left ear".	/3
	Ask patient to read and obey a written command	
	on a piece of paper stating "Close your eyes".	/1
	Ask the patient to write a sentence. Score if it is sensible and has a subject and a verb.	/1
COPYT	NG	
	Ask the patient to copy a pair of intersecting pentagons:	

Episodic & Semantic memory

Working memory

Working memory, executive & Semantic

Episodic memory

Language, Semantic & Working memory

Visuoconstructive, Procedural & Working memory

Clinical interest: discussion on driving

Concerns about cognitive functioning & driving:

Procedural memory versus working and episodic memory

Automatic versus controlled behaviour

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Clinical Dementia testing: methods and meaning

Clinical interest: discussion on driving

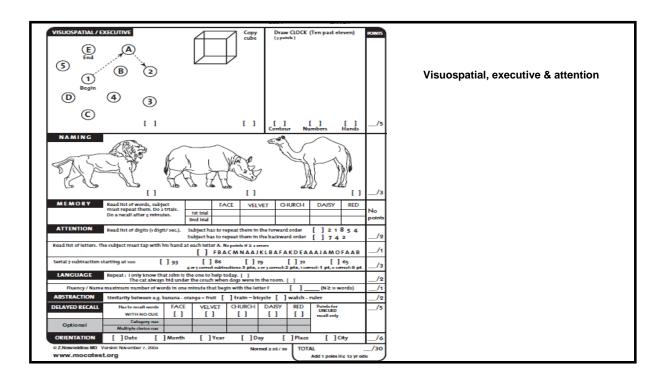
Orientation: could you complete an accident report
Working memory: could you immediately remember a traffic sign
Working memory & executive: could you keep your attention on
multiple informations

Semantic: could you complete an accident report Visuoconstructive: if you do not master 2 dimensions, could you master 3D?



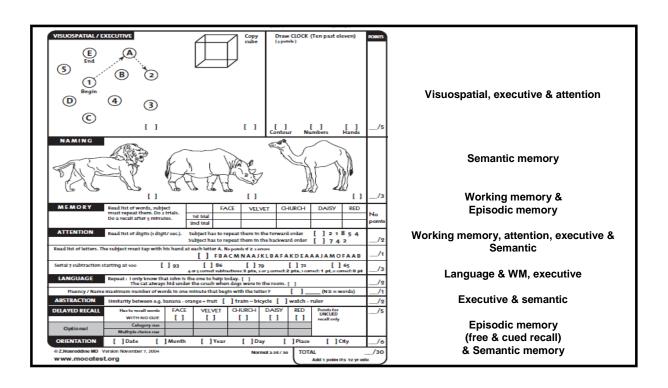
Montreal Cognitive Assessment Scale











Fluency: name a maximum number of words in one minute that begin with the letter « F » (> 11 words)

In the Frontal Assessment Battery: "Say as many words as you can beginning with the letter "S," any words except surnames or proper nouns."

Patient should be stimulated. The time allowed is 60 seconds.

> 9 words: 3 6 -9 words: 2 3 -5 words: 1 < 3 words: 0

[The FAB, Dubois et al, Neurology 2000;55:1621–162]

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Clinical interest: Subjective cognitive decline Decreased Verbal Fluency 12 years before dementia diagnosis (compared to Healthy controls) [Adapted from Amieva et al, Ann Neurol 2009] 5th EAN Congress Oslo, 2019

Multiple causes of cognitive or behavioral difficulties

Motivation
Fatigue & Sleep (sleep apnea, ...)
Stress, depression, psychiatric diseases
Medication & toxic substances
Metabolic & Systemic pathologies
Different types of dementia

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Clinical Dementia testing: methods and meaning

Probable clinical AD dementia (McKhann et al, 2011)

Insidious onset. Symptoms have a gradual onset over months to years, not sudden over hours or days;

a. Amnestic presentation is the most common syndromic presentation of AD dementia. The deficits should include impairment in learning and recall of recently learned information.

b. Nonamnestic presentations:

- Language presentation: The most prominent deficits are in word-finding
- Visuospatial presentation: spatial cognition, including object agnosia, impaired face recognition
 - Executive dysfunction: impaired reasoning, judgment, and problem solving.

The diagnosis of probable AD dementia **should not** be applied when there is evidence of (a) substantial concomitant **cerebrovascular disease**, defined by a history of a stroke temporally related to the onset or worsening of cognitive impairment; or the presence of multiple or extensive infarcts or severe white matter hyperintensity burden



Case 1

Men, 70y, former teacher;

Complaints about memory since 2 years, forgets what he read, less attentive when playing cards, asking for repetition of recent information; needs help for handling finances (spouse); Depressed since 1 year, poor sleep, irritability (spouse). His father was diagnosed with major depression.

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ORIEN	TATION	
	Year Month Day Date Time	/5
	Country Town District Hospital Ward	/5
REGIS	TRATION	
	Examiner names 3 objects (eg apple, table, penny) Patient asked to repeat (1 point for each correct).	
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	sensible and has a subject and a verb.	
COPY		
	Ask the patient to copy a pair of intersecting pentagons:	

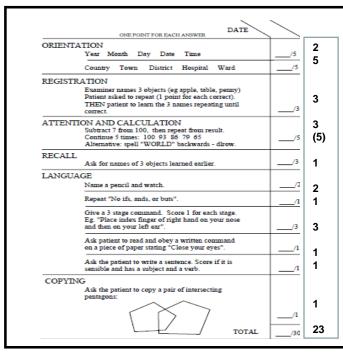
Depression
Versus
Early stage dementia?

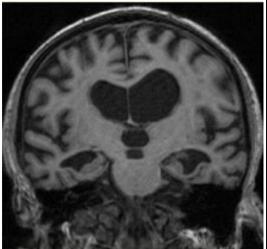
Case 1

Men, 70y, former teacher;

Complaints about memory since 2 years, forgets what he read, less attentive when **playing** cards, **asking** for repetition of recent information; **cannot remember bad recent information**; Depressed since **1 year (first episode)**, poor sleep, irritability. His father was diagnosed with depression.







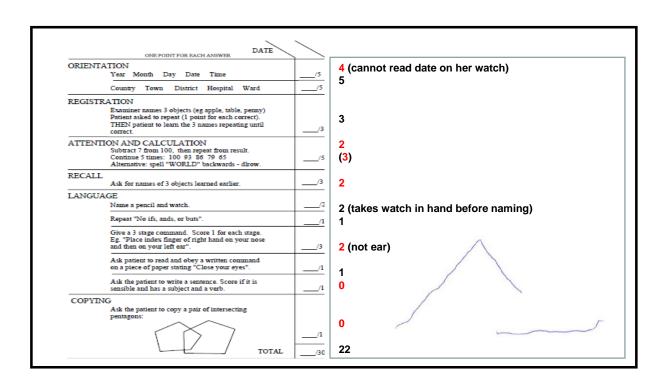
Medial temporal lobe atrophy as a biomarker of AD neurodegeneration

Case 2

Woman, 60y, working in public administration
Progressive difficulty for putting objects at the right place, loss of usual objects, forgets information, difficulties using home appliance, difficulties with spatial orientation (stopped driving).

No toxic, no neurological story, spectacles;
No familial story;
Hesitation for sitting down, no parkinsonian sign





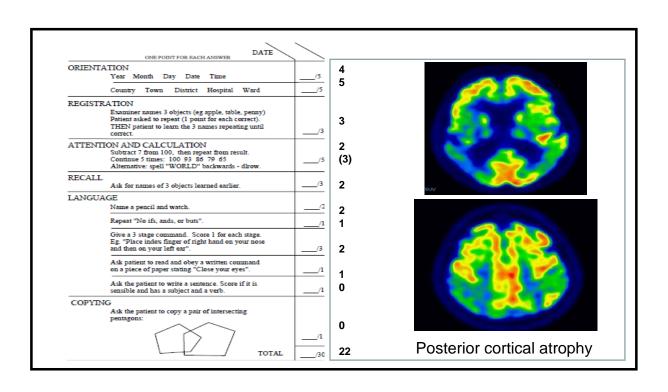
Case 2

Woman, 60y, working in public administration
Progressive difficulty for putting objects at the right place, loss of usual objects, forgets information, difficulties using home appliance, difficulties with spatial orientation (stopped driving).

No toxic, no neurological story, spectacles;
No familial story;

Hesitation for sitting down, for taking the pencil, no parkinsonian sign





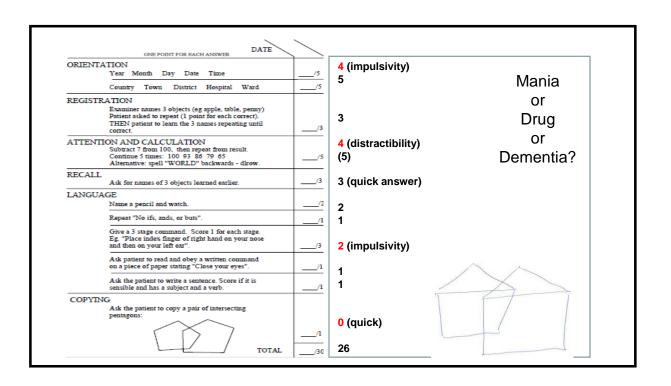
Case 3

Woman, 58y, bank employee

2 hypo-manic episodes, changes in her social behavior, changes in her personality, inadapted emotion, unaware of the behavioral anomaly

Drug treatment (carbamazepine), forgetfulness, lack attention, cannot organize her daily activities, cannot go back to work.





Case 3

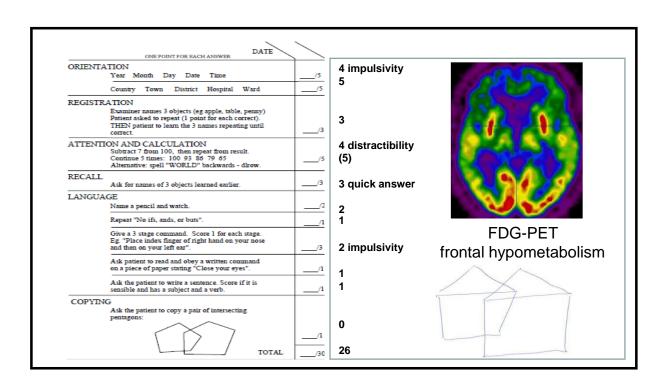
Woman, 58y, bank employee

2 hypo-manic episodes, changes in her social behavior, changes in her personality, inadapted emotion, unaware of the behavioral anomaly

Drug treatment (carbamazepine), forgetfulness, lack attention, cannot organize her daily activities, cannot go back to work.

Impaired sequences of tapping, impaired fluency (errors), loss of social inhibition during testing





Frontotemporal dementia – behavioural variant

Raskovski et al. Sensitivity of revised diagnostic criteria for the behavioural variant of frontotemporal dementia. Brain 2011; 134: 2456-2477

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Possible bvFTD

Three of the following behavioural/cognitive symptoms (A–F) must be present to meet criteria.

A. Early behavioural disinhibition

Socially inappropriate behaviour or Loss of manners or decorum or Impulsive, rash or careless actions
B. Early apathy or inertia

C. Early loss of sympathy or empathy

Diminished response to other people's needs and feelings or Diminished social interest, interrelatedness or personal warmth

D. Early perseverative, stereotyped or compulsive/ritualistic behaviour

Simple repetitive movements or Complex, compulsive or ritualistic behaviours or Stereotypy of speech

E. Hyperorality and dietary changes

Altered food preference or Binge eating, increased consumption of alcohol or cigarettes or Oral exploration or consumption of inedible objects

F. Neuropsychological profile: executive/generation deficits with relative sparing of memory and visuospatial functions



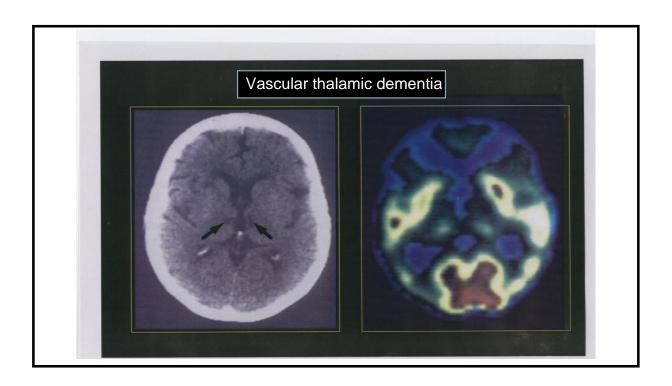
Case 4

Woman, 66y, merchant
Sudden vertigo followed by comatose
Back to consciousness at the hospital, agitation, euphoria, manic behaviour

During the **next weeks**, disoriented, memory impairment and confabulations, apragmatism, disorganised, fluctuating alertness

Dementia? Pathology?





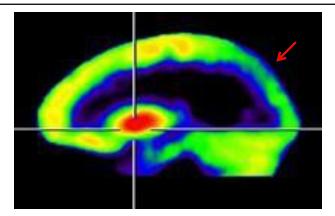
Woman, 68y, grandmother
Fluctuating forgetting of recent events; fluctuating attention to
discussion; fluctuating difficulties in preparing meals or cakes,
episodes of visual hallucinations (criticized).
Poor sleep (frequent movements) > benzodiazepine
MMSE from 25 to 13 (on different days, GP versus neurologist)
Micrography

Dementia? Characteristics? Pathology?

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PET-FDG: occipital hypometabolism



Revised criteria for the clinical diagnosis of probable and possible dementia with Lewy bodies (DLB) (McKeith et al, 2017)

Dementia. Prominent or persistent memory impairment may not necessarily occur in the early stages but is usually evident with progression.

Deficits on tests of attention, executive function, and visuoperceptual ability may be especially prominent and occur early.

Core clinical features (The first 3 typically occur early and may persist throughout the course.)

Fluctuating cognition with pronounced variations in attention and alertness.

Recurrent visual hallucinations that are typically well formed and detailed.

REM sleep behavior disorder, which may precede cognitive decline.

One or more cardinal features of parkinsonism: bradykinesia, rest tremor, or rigidity.

Supportive clinical features

Severe sensitivity to antipsychotic agents; postural instability; repeated falls; syncope or other transient episodes of unresponsiveness; severe autonomic dysfunction, e.g., constipation, orthostatic hypotension, urinary incontinence; hypersomnia; hyposmia; hallucinations in other modalities; systematized delusions; apathy, anxiety, and depression.

Indicative biomarkers

Reduced dopamine transporter uptake in basal ganglia demonstrated by SPECT or PET.

Abnormal (low uptake) 123iodine-MIBG myocardial scintigraphy.

Polysomnographic confirmation of REM sleep without atonia.

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Take home message

Interview, observation and relative

Short cognitive testing (> neuropsychological evaluation)

Biomarkers



References

Folstein et al. "Mini-mental state". A practical method for grading the cognitive state of patients for the clinician. Psychiatr. Res 1975:12(3):189–198.

Nasreddine et al. The Montreal Cognitive Assessment, MoCA: a brief screening tool for mild cognitive impairment. J Am Geriatr Soc 2005;53:695–9

Tsoi et al. Cognitive Tests to Detect Dementia. A Systematic Review and Meta-analysis. JAMA Intern Med. 2015;175(9):1450-1458. doi:10.1001/jamainternmed.2015.2152

Costa et al. The need for harmonisation and innovation of neuropsychological assessment in neurodegenerative dementias in Europe: consensus document of the Joint Program for Neurodegenerative Diseases Working Group. Alzheimer's Research & Therapy (2017) 9(1):27.

DOI 10.1186/s13195-017-0254-x

Albert et al. The diagnosis of mild cognitive impairment due to Alzheimer's disease: recommendations from the National Institute on Aging Alzheimer's Association workgroups on diagnostic guidelines for Alzheimer's disease. <u>Alzheimers Dement</u>. 2011 May;7(3):270-9. doi: 10.1016/j.jalz.2011.03.008

McKhann et al. The diagnosis of dementia due to Alzheimer's disease: Recommendations from the National Institute on Aging-Alzheimer's Association workgroups on diagnostic guidelines for Alzheimer's disease. <u>Alzheimers Dement</u>. 2011 May;7(3):263-9. doi: 10.1016/j.jalz.2011.03.005

McKeith et al. Diagnosis and management of dementia with Lewy bodies. Fourth consensus report of the DLB Consortium. Neurology 2017;89:88–100

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Useful information

Dementia Revealed Toolkit, NHS England (https://www.england.nhs.uk/wp-content/uploads/2014/09/dementia-revealed-toolkit.pdf)

NICE guidelines: Dementia: assessment, management and support for people living with dementia and their carers (https://www.nice.org.uk/guidance/ng97/resources/dementia-assessment-management-and-support-for-people-living-with-dementia-and-their-carers-pdf-1837760199109

Alzheimer Society cognitive assesment toolkit (https://www.alzheimers.org.uk/sites/default/files/migrate/downloads/alzheimers_society_cognitive_assessmen t toolkit.pdf

