

FTD: Fronto-temporal dementia(s)

Riedl et al., 2014, *Neuropsychiatric Dis. Treat.* 10, «Fronto-temporal lobar degeneration: current perspectives»

atrophy: mesio- and orbitofrontal, anterior insula

- 1) Behavioral variant of FTD: loss of former interests and social skills (disinhibition, lack of distance, poor empathy); stereotypes, hyperoral behavior, «exekutive deficits»

bv FTD

(behavioral variant FTD)

- 2) PPA: Primary Progressive Aphasias

language variants of FTD

- a) svPPA: semantic variant: fluid speech, spared grammar, loss of semantic knowledge, later on also personality changes
- b) nfvPPA: non-fluid aphasia variant: agrammatism, spared comprehension, spared semantic knowledge
- c) lvPPA: logopenic variant with spared grammar and semantics, but word finding deficits and inability to repeat words/sentences rather posterior temporal



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66-year-old woman with PPA

Right-handed. 12 years of schooling, commercial education. Owns an antiques shop in a small city. Signs of nonfluent verbal expression for 6 years. No other complaints.

Referred to by neurologist in private praxis to have a second opinion after a first neuropsychological evaluation (5 months ago) had diagnosed a functional speech disorder (first signs after husband ended marriage).

Early hx and family hx uneventful.

cMRI (5 months ago) discrete atrophy of the frontal lobes. Incidental finding: left frontal developmental venous anomaly (DVA)

Regular neurostatus

EEG right temporo-posterior focal abnormality without epileptic potentials

Liquor: cells 3/ul, protein 659 mg/l, lactate slightly reduced (1.5 mmol/l), no intrathecal Ig-Synthese, oligoclonal bands negative

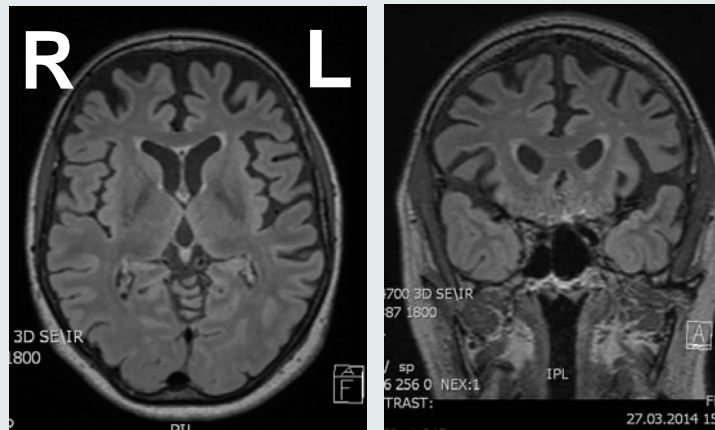


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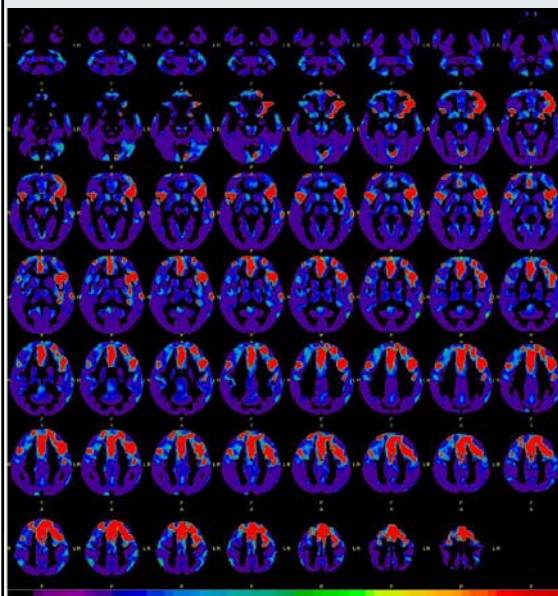
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66-year-old woman with PPA

cMRI (FLAIR):



66-year-old woman with PPA



FDG-PET:
(fluorodeoxyglucose, ^{18}F -FDG)

may show hypometabolism
long before any atrophy is
discernible in MRI

In present case:
Markedly reduced glucose
metabolism in bifrontal and,
less markedly, in bitemporal
regions (L > R)

66-year-old woman with PPA

Findings in neuropsychological examination:

Cooperative, fully oriented, cheerful.

History can be obtained, but examiner makes frequent proposals such that patient may answer with «yes» or «no».

Spontaneous speech almost limited to nouns. Telegram style with frequent set phrases («Ja, genau!», «dasch richtig»). Preserved comprehension.



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Spontaneous speech

Patient is asked whether she makes phone calls (yes), with how many people (about 4), what her strategy is, once somebody else is on the phone («I speak»)

(Use of phone call verified by examiner:
appointment for second visit over the phone)

this person still makes phone calls

Clip shows:

It is possible to communicate
as long as binary alternatives
are provided

-> yes/no responses

set phrases



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Spontaneous speech



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It is possible to communicate as long as binary alternatives are provided

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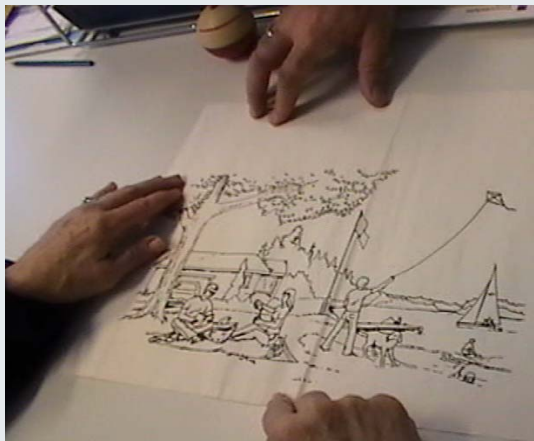
NOT a model examination ...



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Picture description



Clip shows:

- Spared perceptual integration («pic-nic»)
- Spared articulation
- Spared naming
- Spared comprehension

BUT:

- Impressive agrammatism with almost exclusive use of nouns

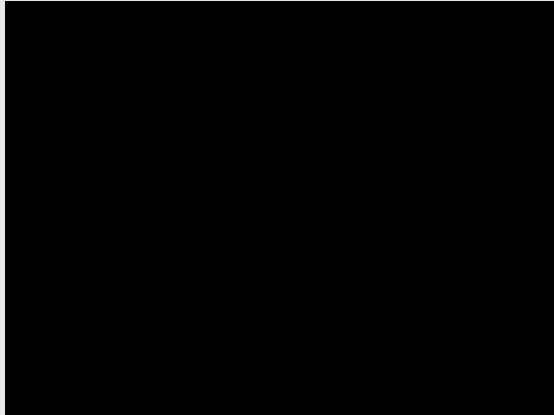
she cant construct meaningful sentences, lost the grammar



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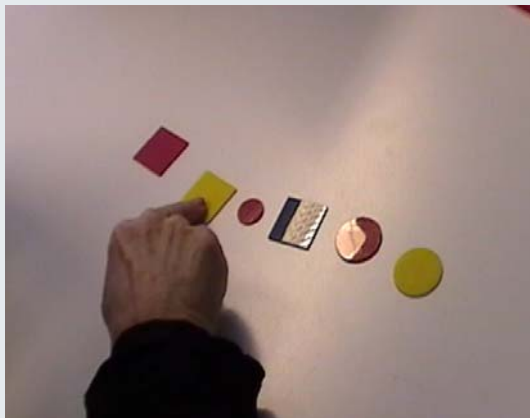
- Impressive agrammatism with almost exclusive use of nouns



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Language comprehension



Formal testing shows:

- Impairment, but...
- ... due to interference among inter-phrase elements
- Disinhibition in responding (cannot withhold response)

Interpretation: deficient are the «executive function» components of the language process (note: digit span is 5)



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Confrontation naming

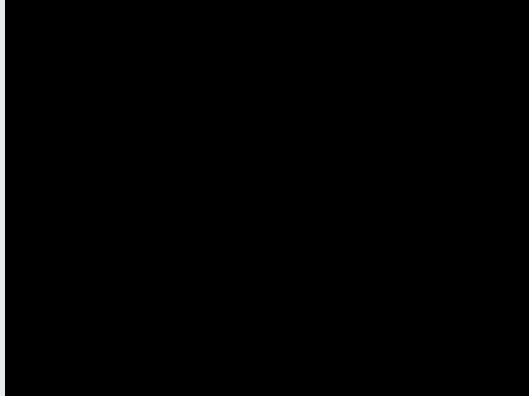


Correct and fast

(in accordance with picture description; but free naming and forced naming may show a striking dissociation!)

Confrontation naming

naming of single items no problem, uses one word descriptions, she is fast also



Correct and fast

Interpretation: nonfluent speech not because of a lack of the names or meaning of words, but because of the non-availability of linguistic relations (deficient syntax, grammar)



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Agrammatism

Clip shows abolished sense for simple linguistic relations

«Hans is taller than Otto. Who is the smaller of the two?»

«Hans is taller than Otto, but smaller than Sepp. Who is the tallest of all?»
(Not shown)



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Agrammatism



Clip shows abolished sense
for simple linguistic relations

Repetition



Flawless

Mnestic functions:

Learning

Storage: free recall («active memory»)
if insufficient: no conclusions
possible concerning storage!
(either *storage* or *retrieval* deficit!)

-> recognition («passive memory»)

Quality needs to be quantified!
(repetitions, intrusions, confabulations,...)

Mnestic functions: learning, recall, recognition

LÖWE

SMARAGD

PFERD

ZELT

SAPHIR

HOTEL

HÖHLE

OPAL

TIGER

PERLE

KUH

HÜTTE

4 animals

4 precious stones

4 'housings'

1) recalled after 1st reading

5

2) recalled after 2nd reading

8

3) recalled after 3rd reading

8

4) late recall (20 minutes)

5

5) Recognition: was «Pferd» in the list?

100%

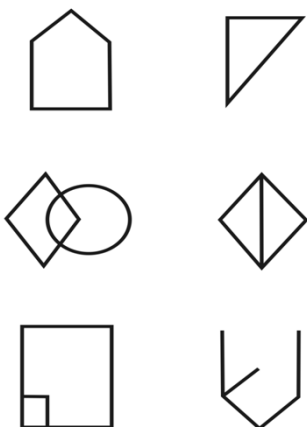
Rubin? Katze? Fahrrad? Opal?

confabulations and repetitions!





2 c., 3 rep.

Mnemonic functions: learning, recall, recognition

(correctly recalled figure scores 1, at correct place plus 1)



- 1) recalled after 1st reading
- 2) recalled after 2nd reading
- 3) recalled after 3rd reading
- 4) late recall (20 minutes)
- 5) Recognition:

(confabulations and repetitions!)

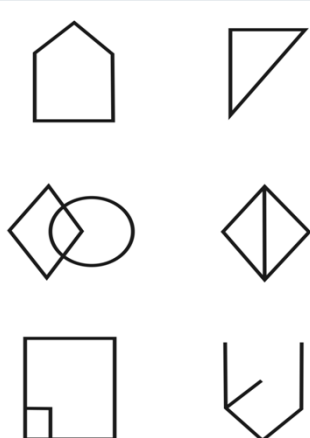



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Brief Visuospatial Memory Test: 6 variants for repeated exams 78/88

Mnemonic functions: learning, recall, recognition

(correctly recalled figure scores 1, at correct place plus 1)







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(compare DAT-case age 58, education 20+ 1/1/1/late1; rec.100%)

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Visual-perceptual functions



Spared, in all aspects.
(Shown for faces. Cave: not being able to name a face does not imply inability to recognize it!)

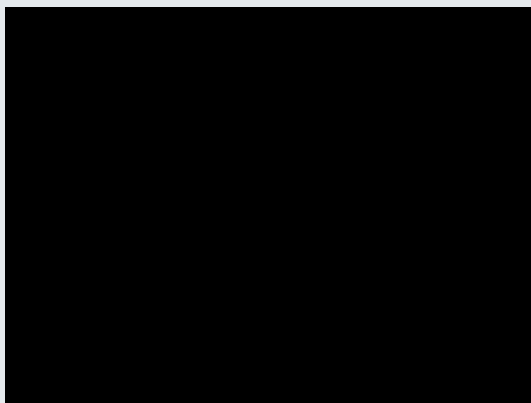
Visuo-constructive abilities insufficient because of planning deficits



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«Executive functions» PFC functions basically

Planning, monitoring and modifying action sequences.
Adaptation of automatized behavior to temporary affordances («flexibility»)

Initiating and sequencing of behavioral sequences, inhibiting inappropriate steps

Innovative generation of ideas according to (abstract or categorical) criteria

Conceptual thinking: finding concepts; shifting between them



Testing executive functions

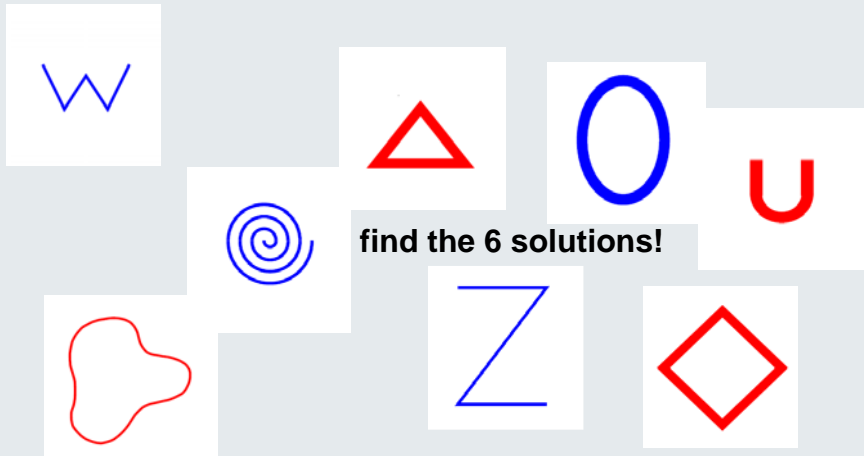
-> formal assessment of:

- 1) suppression of interferences (spared; not shown)
- 2) verbal fluency: letter criterion, category criterion (both impaired; not shown)
- 3) planning abilities in copying a complex meaningless drawing: impaired (not shown)
- 4) concept finding and conceptual flexibility most severely impaired



Executive functions

Concept finding and conceptual flexibility (Kramer)



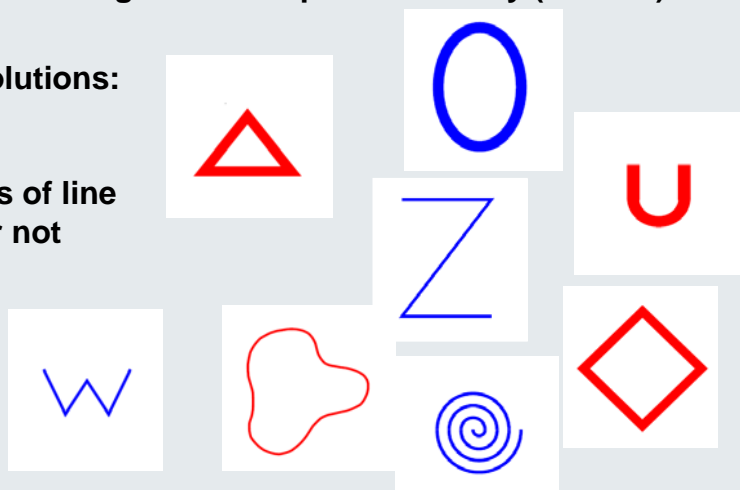
«Sort the 8 cards by making two groups (each with 4 cards), such that all 4 of a group have something in common» 84/88

Executive functions

Concept finding and conceptual flexibility (Kramer)

Six valid solutions:

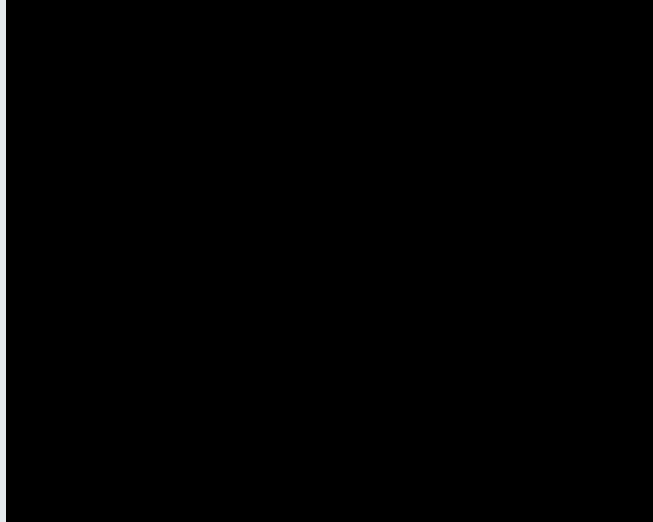
- 1) color
- 2) form
- 3) thickness of line
- 4) letters or not
- 5) size
- 6) closure



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Executive functions (concept finding and conceptual shifting)



Note:

* manual performance superior to verbalization!

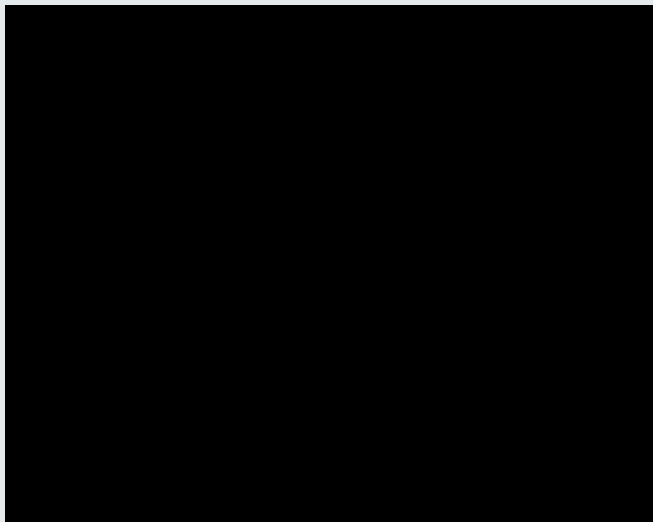
* perseverative quality of conceptual thinking (in the absence of motor perseveration)



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Executive functions (concept finding and conceptual shifting)



Note again:

manual performance dissociates from verbalization



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To sum up

PPA = Primary Progressive Aphasia,
a form of a fronto-temporal dementia

This case of a degenerative aphasia («PPA») illustrates:

(Relative) preservation of mnestic functions for years after onset of first signs

«Language» is very broad construct! Some aspects can be entirely preserved, others abolished

Surprising discrepancy between integration in everyday life (taking phone calls verified!) and actual test performance

Cognitive assessments should not unilaterally quantify deficits, but also delineate personal strengths and cognitive reserve!



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Questions?



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