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Dementia as a motor disorder

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The structure of the talk

- Cognitive aspects of motor disorders
- Cognitive assessment in motor patients (ECAS)
- Motor aspects of cognitive disorders
- Motor assessment in cognitive patients:
 - As it is at the moment...
 - ... and as it could be

Example 1

- Presenting 1993, aged 68 y.
- Patient: 6/12 hx of morning fatigue, feeling stressed, forgetful
- Husband: 2y hx of personality change, ritualistic, stereotypic, apathy
- Normal neurological exam, dgn: "*progressive frontal lobe syndrome*"
- 1994: imbalance, falls, hypomimia, bradykinesia, dysarthria
- 1995: *Slowing of vertical gaze*, diagnosis revised to PSP
- Progressive deterioration, death 2000, pathological dgn: PSP

Example 2

- Presenting 1999, aged 58 (after husband's death from leukemia)
- 2000 - disinhibition (eating from other people's plates), staring gaze
- 2001 - obsession with funerals, digging up her husband ashes
- 2002 - spelling errors on Xmas cards
- 2003 - referred to Addenbrookes for dysphagia & loss of weight
- Nov 2003 - dgn. MND/dementia
- 2004 - death, pathological diagnosis: MND

Movement, cognition & behaviour

- **Converging evidence from:**
 - Clinical studies
 - Imaging, molecular biology, pathology & genetics
- **Motor, cognitive & behavioural changes as different expressions of the same underlying pathology**
- **"Why movement & cognition belong together"** (Bak, *Nature Reviews Neurology*, 2010)
- **Involvement of motor cognition as an integral phenotype of motor disorders**
- **"What wires together, dies together"** (Bak & Chandran, *Cortex*, 2012)

Motor/cognitive overlap: implications

- Diagnosis
- Prognosis
- Management
- Natural history => Treatment monitoring

Difficulties assessing cognitive functions in patients with motor symptoms

Speaking:

- Dysarthria
- Aphonia
- Mutism

Writing & drawing

- Rigidity
- Tremor
- Weakness
- Apraxia

And yet...

- Practically all major cognitive screening tools (MMSE, MoCA, ACE, DRS etc) have been designed assuming normal motor function

Edinburgh Cognitive Assessment (ECAS)

- Cognitive assessment for patients with motor deficits
- Tries to minimise the influence of motor dysfunction (e.g. pointing tasks, yes/no answers etc)
- Parallel written and spoken versions
- Multi-dimensional
- First applied in ALS, currently piloted in PD & PSP

ECAS – LANGUAGE

- Naming
- Comprehension:
 - nouns & verbs
- Spelling:
 - Irregular & regular
 - Compounds

EDINBURGH COGNITIVE AND DEPENDENCE ASSESSMENT - ECAS
English Version 2010

LANGUAGE - Naming

Look at the pictures and name the objects or people.

LANGUAGE - Comprehension

Read the questions and answer.

LANGUAGE - Spelling

Write the words in the correct order.

1. Cow	2. Sheep	3. Pig	4. Horse
5. Dog	6. Cat	7. Rabbit	8. Bird
9. Frog	10. Snake	11. Lizard	12. Insect
13. Fish	14. Turtle	15. Crab	16. Shell
17. Octopus	18. Jellyfish	19. Starfish	20. Seahorse
21. Dolphin	22. Whale	23. Shark	24. Manta ray
25. Stingray	26. Hammerhead	27. Sawfish	28. Shark
29. Whale shark	30. Great white shark	31. Tiger shark	32. Hammerhead shark
33. Bull shark	34. Lemon shark	35. Nurse shark	36. Tiger shark
37. Hammerhead shark	38. Tiger shark	39. Hammerhead shark	40. Tiger shark

ECAS – EXECUTIVE FUNCTIONS

- Reverse Digit Span
- Alternation
- Inhibition (Hayling)
- + verbal fluency index

EXECUTIVE - Reverse Digit Span

Look at the numbers and say them out loud in the order given.

1	2	3	4	5	6	7	8	9	0
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EXECUTIVE - Alternation

Look at the numbers and say them out loud in the order given.

1	2	3	4	5	6	7	8	9	0
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EXECUTIVE - Hayling

Look at the words and say them out loud in the order given.

1	2	3	4	5	6	7	8	9	0
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ECAS – SOCIAL COGNITION

SOCIAL COGNITION - Part A

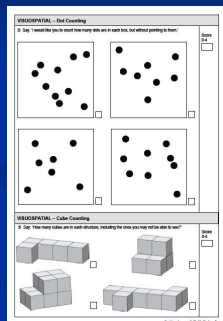
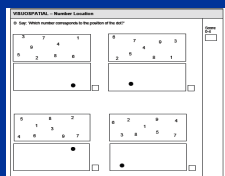
Look at the pictures and say them out loud in the order given.

SOCIAL COGNITION - Part B

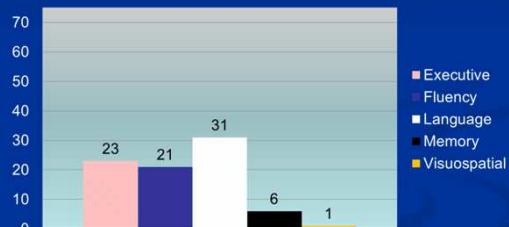
Look at the pictures and say them out loud in the order given.

ECAS – VISUOSPATIAL FUNCTIONS (WITHOUT DRAWING)

- Number location
- Dots
- Cubes



ECAS Subdomains: Frequency of Abnormal Performance (75 ALS patients)



Motor features in dementias

- Cognitive/behavioural features often precede the motor:
 - bvFTD, followed by ALS or PSP
 - Progressive aphasia, followed by CBD
- In these initial stages patients often seen by psychiatrists and their assessment focused on cognitive symptoms
- Are patients with dementia/cognitive symptoms examined for the presence of motor symptoms?

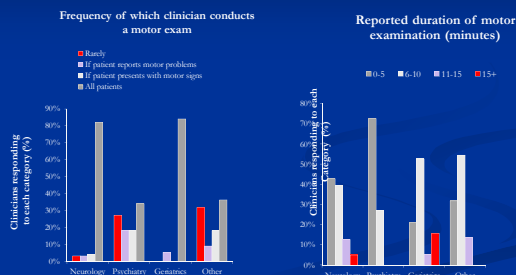
“Cognitive” disorders are not just cognitive

- 20-50% of AD patients have motor signs (*Parkinsonism*)
- Present in early stages (inc. MCI), progressing with time
- Associated with faster cognitive & functional decline
- PM: motor symptoms with pure AD pathology
- Atypical AD – PCA associated with motor symptoms

Current practice of motor examination in cognitive clinics: an on-line survey

- 10 brief questions (< 2 minutes), available in 10 languages
- Sent to mailing lists, organisations, individual contacts
- 323 responses (166 to non-English), from 32 countries
- UK: 48, Eur: 115, N America: 50, S America: 33, Asia: 73
- Neurology 185, Psychiatry: 65, Geriatrics: 28

Motor examination in dementia clinics: how often? how long?



Cognitive screening in dementia clinics

92% use regularly screening tools, often more than one:

- 62% MMSE (9% alone, 53% together with other tests)
- 36% MoCA
- 20% ACE (UK 86%, South America 26%, RotW ca 10%)
- 14% CDR
- 5% Clock drawing
- 3.2% DRS
- 3% FAB

Motor screening in dementia clinics

34% use motor screening tests (mostly neurologists)

- 23 % UPDRS
- 4% MRC power scale
- 2% Golbe PSP scale
- No general (non-disease specific) screens used

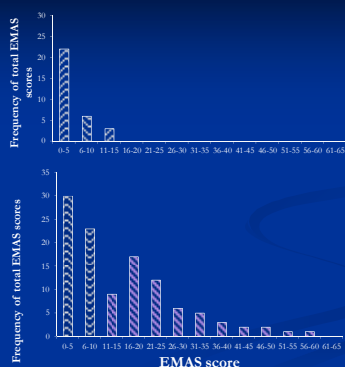
Edinburgh Motor Assessment (EMAS): Objectives

- Brief (5-10 mins)
- Easy to conduct:
 - No need for special equipment (tendon hammer etc)
 - No need for special rooms (e.g. bed)
- Easy to score
- Should focus on symptoms particularly relevant in dementias

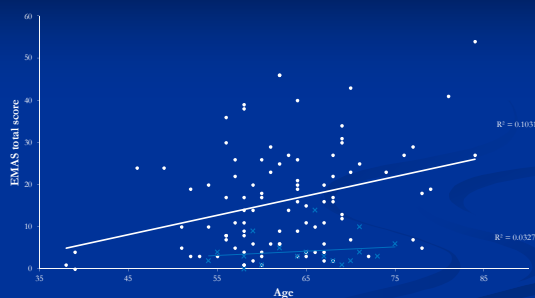
EMAS – Structure & scoring

- 33 Items, 4 dimensions:
 - Extraparamidal, amyotrophic, cerebellar, complex
- Grouped by descending body region:
 - Eye movements, mouth, upper limbs, lower limbs/gait
- 0-3 scoring system (as used from pathology to NPI):
 - 0 = normal function/no abnormal signs
 - 1 = equivocally abnormal/slightly reduced function
 - 2 = unequivocally abnormal/significant reduction of function
 - 3 = severely abnormal/lack of function

The first results: impairment in ca. 50% of patients

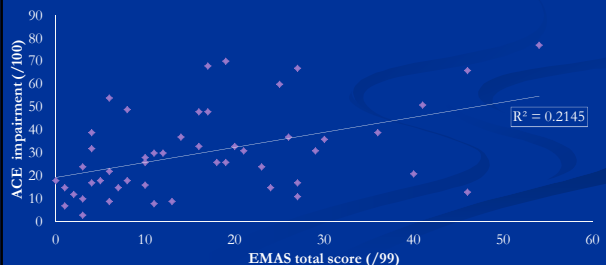


The influence of age on EMAS score



EMAS vs ACE Impairment

n=49	Unimpaired ACE	ACE impairment (<88)
Unimpaired EMAS	6%	16%
EMAS impairment (>12)	8%	70%



EMAS & ACE

ACE	Attention	Memory	Fluency	Language	Visuo spatial	Total Score
EMAS						
Extra pyramidal	0.446**	0.069	0.392**	0.195	0.473**	0.322*
Cerebellar	0.383*	0.050	0.256	0.110	0.433**	0.249
Complex	0.687***	0.366*	0.622***	0.364*	0.744***	0.664***
Total Score	0.566***	0.176	0.488***	0.252	0.615***	0.463***

Interested?

Please contact me for:

- The motor assessment in dementia clinic survey:
<https://www.surveymonkey.com/r/?sm=wPov0zYXVYAve5abWn7Uag%3d%3d>
- Any information regarding EMAS
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