

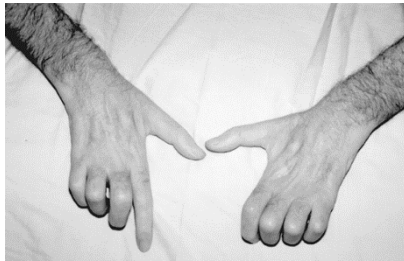
A vertical image on the left side of the slide showing the dome and upper part of St. Martin-in-the-Fields church in London.

Frontotemporal Dementia and ALS in daily Practice: **Red Flags**

Professor Sharon Abrahams

s.abrahams@ed.ac.uk

ALS to FTD



ALS-motor
50%



ALSci- cognitive
impairment and/or
ALSbi – behavioural
impairment
35%



Frontotemporal
dementia (FTD) <15%

***Behavioural variant:
bvFTD
Characterised by
Personality Change***

GGGGCC repeat expansion in the *C9orf72* gene in
ALS and in FTD

TAR DNA binding protein, TDP-43 most common
pathology in ALS and also in FTD

Table 3 International consensus criteria for behavioural variant FTD (FTDC)

II. Possible bvFTD

Three of the following behavioural/cognitive symptoms (A–F) must be present to meet criteria. Ascertainment requires that symptoms be persistent or recurrent, rather than single or rare events.

- A. Early* behavioural disinhibition [one of the following symptoms (A.1–A.3) must be present]:
 - A.1. Socially inappropriate behaviour
 - A.2. Loss of manners or decorum
 - A.3. Impulsive, rash or careless actions
- B. Early apathy or inertia [one of the following symptoms (B.1–B.2) must be present]:
 - B.1. Apathy
 - B.2. Inertia
- C. Early loss of sympathy or empathy [one of the following symptoms (C.1–C.2) must be present]:
 - C.1. Diminished response to other people's needs and feelings
 - C.2. Diminished social interest, interrelatedness or personal warmth
- D. Early perseverative, stereotyped or compulsive/ritualistic behaviour [one of the following symptoms (D.1–D.3) must be present]:
 - D.1. Simple repetitive movements
 - D.2. Complex, compulsive or ritualistic behaviours
 - D.3. Stereotypy of speech
- E. Hyperorality and dietary changes [one of the following symptoms (E.1–E.3) must be present]:
 - E.1. Altered food preferences
 - E.2. Binge eating, increased consumption of alcohol or cigarettes
 - E.3. Oral exploration or consumption of inedible objects
- F. Neuropsychological profile: executive/generation deficits with relative sparing of memory and visuospatial functions [all of the following symptoms (F.1–F.3) must be present]:
 - F.1. Deficits in executive tasks
 - F.2. Relative sparing of episodic memory
 - F.3. Relative sparing of visuospatial skills

III. Probable bvFTD

All of the following symptoms (A–C) must be present to meet criteria.

- A. Meets criteria for possible bvFTD
- B. Exhibits significant functional decline (by caregiver report or as evidenced by Clinical Dementia Rating Scale or Functional Activities Questionnaire scores)
- C. Imaging results consistent with bvFTD [one of the following (C.1–C.2) must be present]:
 - C.1. Frontal and/or anterior temporal atrophy on MRI or CT
 - C.2. Frontal and/or anterior temporal hypoperfusion or hypometabolism on PET or SPECT

IV. Behavioural variant FTD with definite FTLD Pathology

Criterion A and either criterion B or C must be present to meet criteria.

- A. Meets criteria for possible or probable bvFTD
- B. Histopathological evidence of FTLD on biopsy or at post-mortem
- C. Presence of a known pathogenic mutation

Partner's perspective



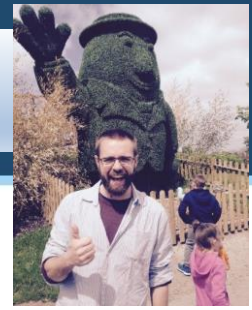
“My own GP has told me that ALS just affects the limbs and NOT the brain functions. I have to disagree as in one year of my late husband's life I saw the change of a caring conscientious family man into an uncaring/unsympathetic, confused, aggressive person. That none of his family or friends recognised. A professional man that did the most bizarre things that were totally out of character.

Even colleagues had noticed that he had gone from a very efficient team leader to a 'couldn't care less attitude' even before he was diagnosed. ... He became unrecognisable like a stranger even though we had been married 22 years.”

“The professionals he saw did not in their appointments see
the man he had become”



Apathy: Dimensional Apathy Scale



Radakovic & Abrahams
<https://das.psy.ed.ac.uk/>

DAS **Dimensional Apathy Scale (Self)** **PN:**

Age..... Sex..... Marital Status.....

Years of Education.....

Choose the answer on how you have **felt, behaved** at the rate of occurrence in the last month: (Circle the statement)

1. I need a bit of encouragement to get things started ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever	7. Before about about ☐ ☐ Occasionally ☐ Hardly Ever
2. I contact my friends ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever	8. I plan my days activities in advance ☐ Almost always
3. I express my emotions ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever	I am not bothered, I don't care
4. I think of new things to during the day ☐ Almost always ☐ Often	
5. I am fat	9. I always ☐ Occasionally ☐ Ever 10. I activation ☐ always
6. I find myself staring in to space ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever	11. I occasionally ☐ Hardly Ever
	12. I struggle to empathise with other people ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever

Emotional Apathy

Executive Apathy

I can't seem to finish things

I am not
bothered, I
don't care

Emotional Apathy

I can't think
of any new
ideas

Initiation Apathy

finish
things

Dimensional Apathy Scale (Self)

PN:

13. I do things for myself almost always ☐ Often ☐ Occasionally ☐ Hardly Ever 14. I try new things ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever 15. I am unconcerned about how others feel about me ☐ Almost always ☐ Often ☐ Occasionally 17. When doing a demanding task, I have difficulty working out what I have to do ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever 18. I keep myself busy ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever	19. I get easily confused when doing several things at once ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever 20. I get bored easily ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever 21. I am spontaneous ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever 23. I am easily distracted ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever 24. I feel indifferent to what is going on around me ☐ Almost always ☐ Often ☐ Occasionally ☐ Hardly Ever
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Initiation Apathy

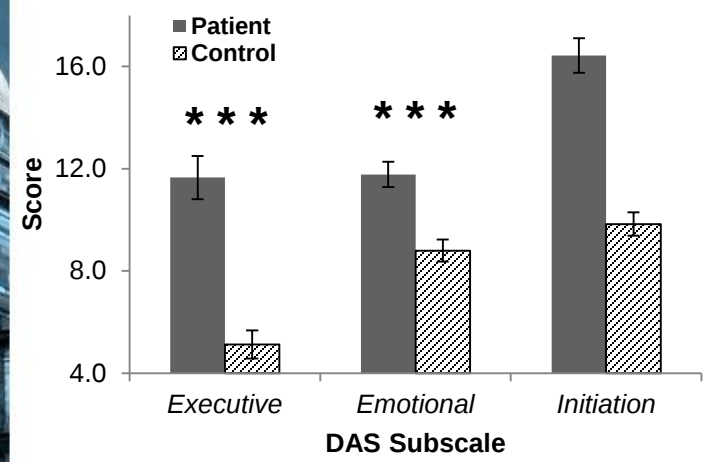
I can't think
of any new
ideas

🚩 Is it Apathy or Depression?:

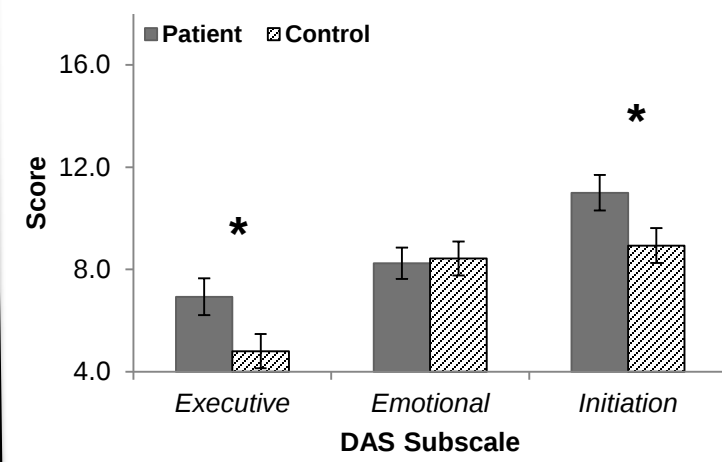
Apathy neutral emotions/thoughts, Depression negative emotions/thoughts



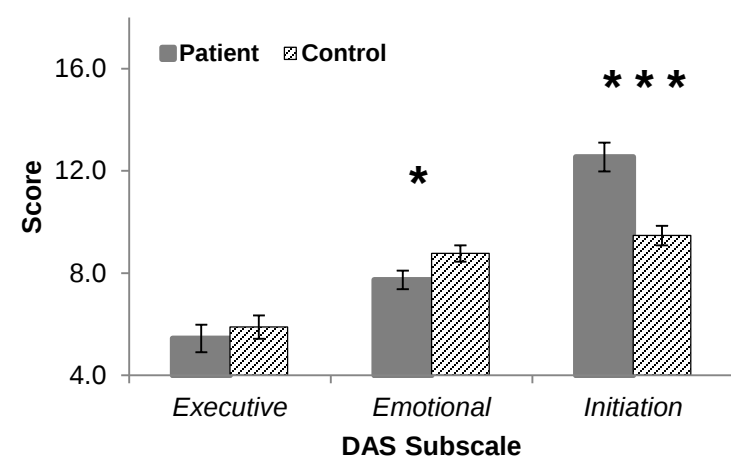
AD



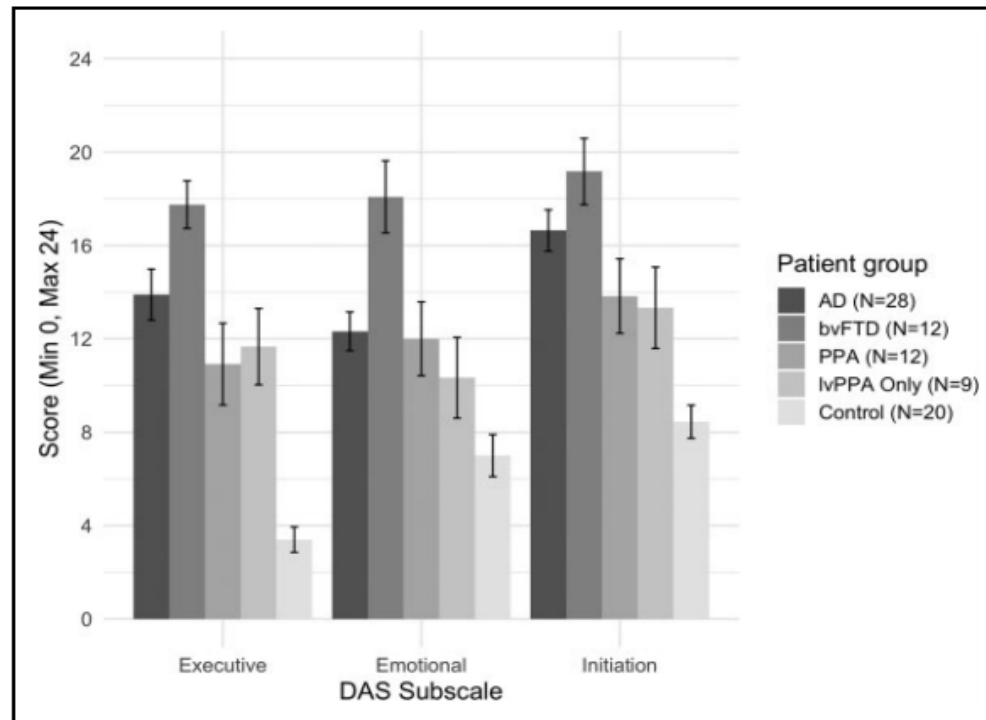
PD



ALS



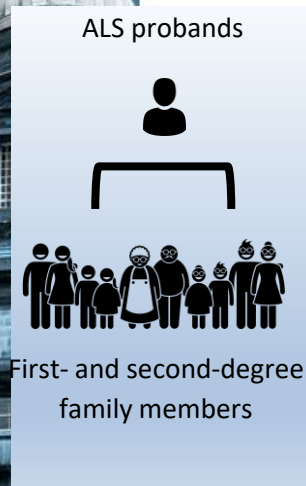
Apathy in bvFTD without ALS



Psychiatric symptoms in ALS

- Depression and anxiety are common in people with ALS and their family members.
- ALS family members compared with healthy controls report higher levels of current anxiety and OCD, and are more likely to report a lifetime episode of:

Psychosis Alcohol use Mania Self-harming behaviour



Current and/or **history** of psychiatric symptoms are associated with poorer cognition and the presence of behaviour changes

ALS Proband

- Depression
- Anxiety
- ADHD
- **Psychosis**
- **Mania**

Family members

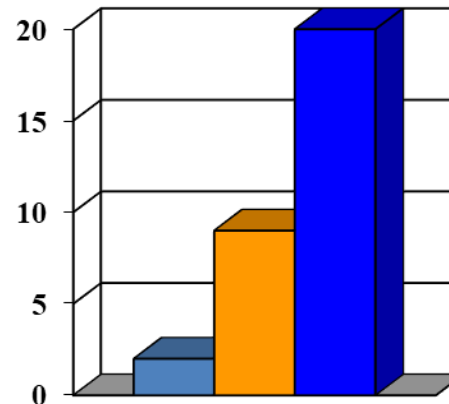
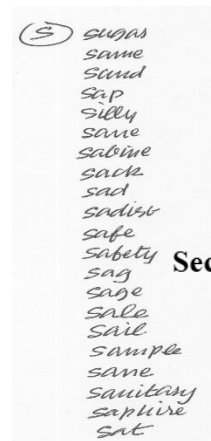
- Anxiety
- ADHD
- **Impulsivity**
- **Autism**



Do not dismiss psychiatric symptoms

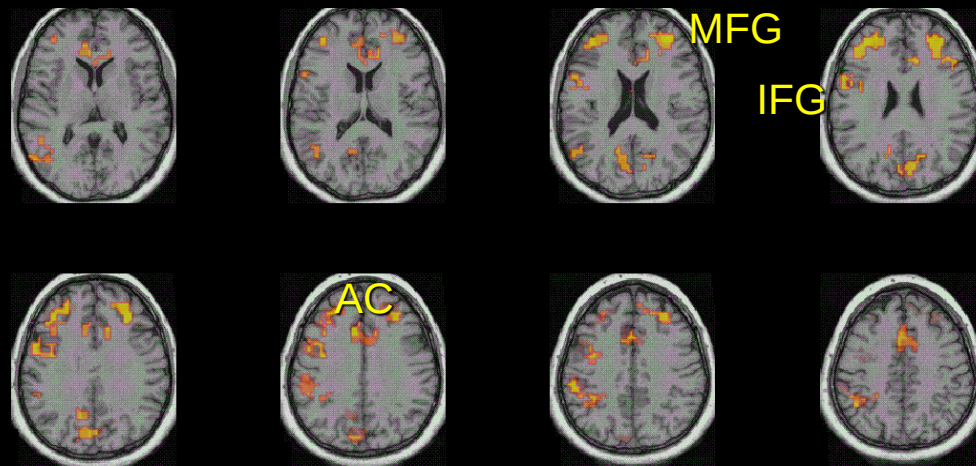
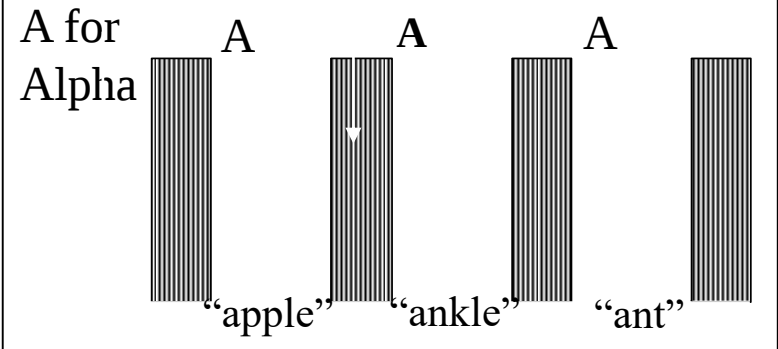
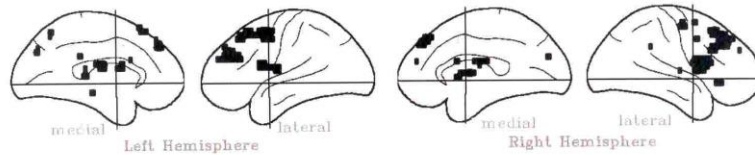
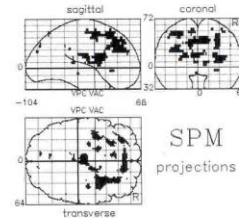


Letter Fluency Ship, Shore, Snake, Silly, Send, Silver, Sonnet, Sun...


$$V_{fi} = \frac{\text{time allowed for test} - \text{time to copy word}}{\text{number of words}}$$


Letter fluency & Functional Brain Imaging

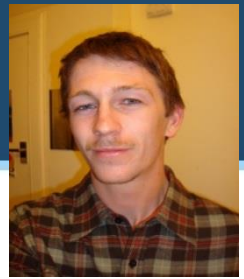
Word Generation
Impaired Activation in
MNDi group
compared with Controls
($p < 0.001$)



PET ($H_2^{15}O$) and fMRI
dorsolateral prefrontal
cortex and Anterior
Cingulate dysfunction
during letter fluency

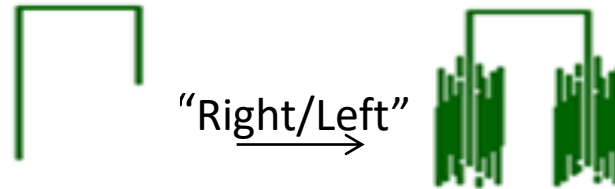
Executive deficits, not processing speed relates to abnormalities in distinct prefrontal tracts in amyotrophic lateral sclerosis

Lewis D. Pettit,^{1,2,3} Mark E. Bastin,^{1,2,4,5} Colin Smith,⁶ Thomas H. Bak,^{1,2,3,4,7} Thomas H. Gillingwater^{1,8} and Sharon Abrahams^{1,2,3,4,7}



Is the cognitive deficit in ALS due to slowed processing speed or executive dysfunction (multi-tasking)?

Processing Speed Task - Visual Inspection Time (VIT):



Stimulus

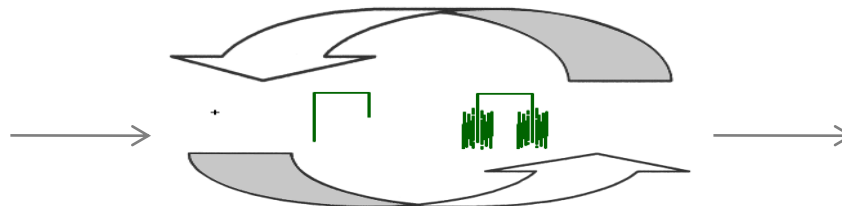
Mask

Response

Dual Task Procedure

Present Numbers

“2 6 4 7 1 9”



Visual Inspection Task

Recall Numbers

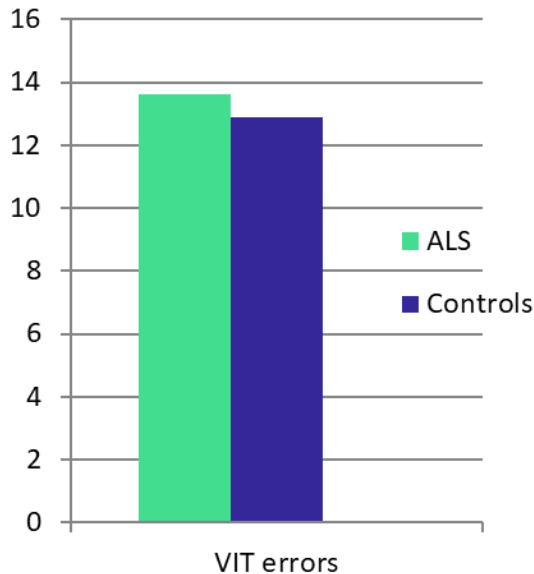
“2 6 4 7 1 9”



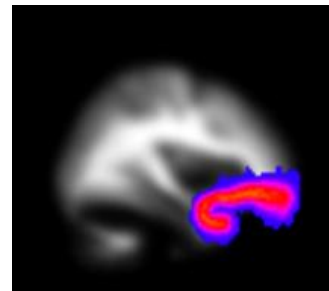
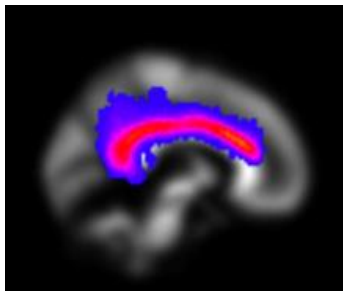
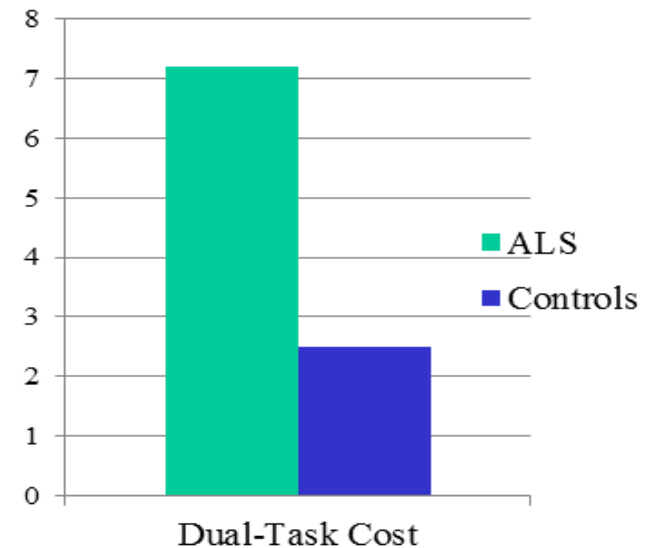


ALS patients shows dual task (executive) impairment and normal information processing speed

Processing Speed



Dual Task



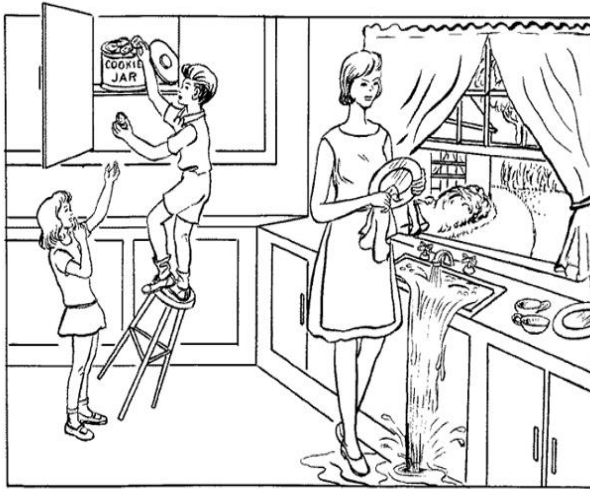
Uncinate and Cingulate integrity affected in ALS

Dual task and verbal fluency correlates with DTI indices for middle and superior frontal lobe white matter



Univ

Language Dysfunction in ALS – Spelling Errors



Copyright © 1983 by Lee & Feltner

BOY IS SLAVE COOKS

MOTHER IS WASHING UP

SINK, IS OVERFLOWER

CS LOOL FALLING

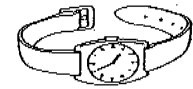
Ask the subject to name the following pictures:

PENCH



PENGIL

WATCH



ANKSD

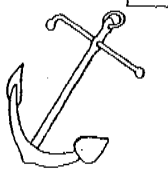
KANGROOS



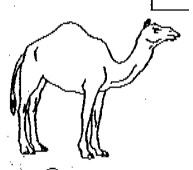
CAMELS



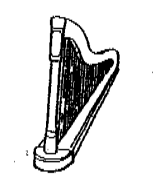
HARSKOR



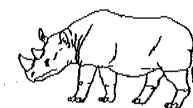
WITHALF



BARRAL



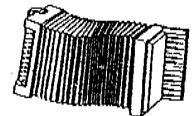
CROND



ALICTOR



HARDISST



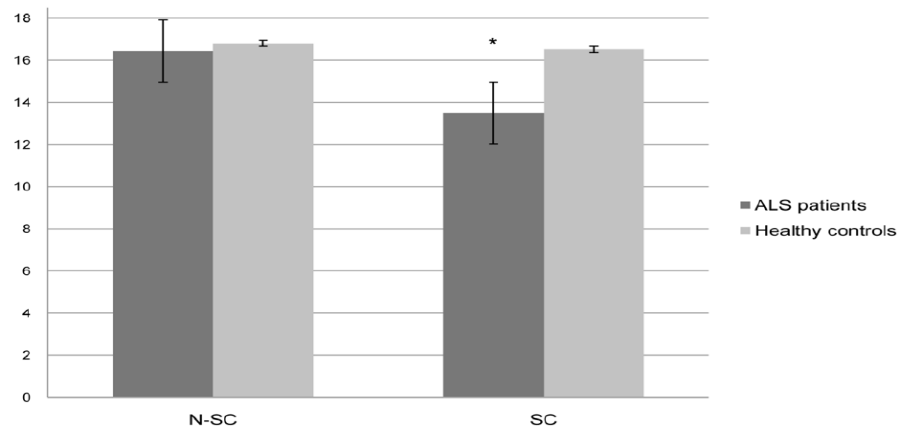
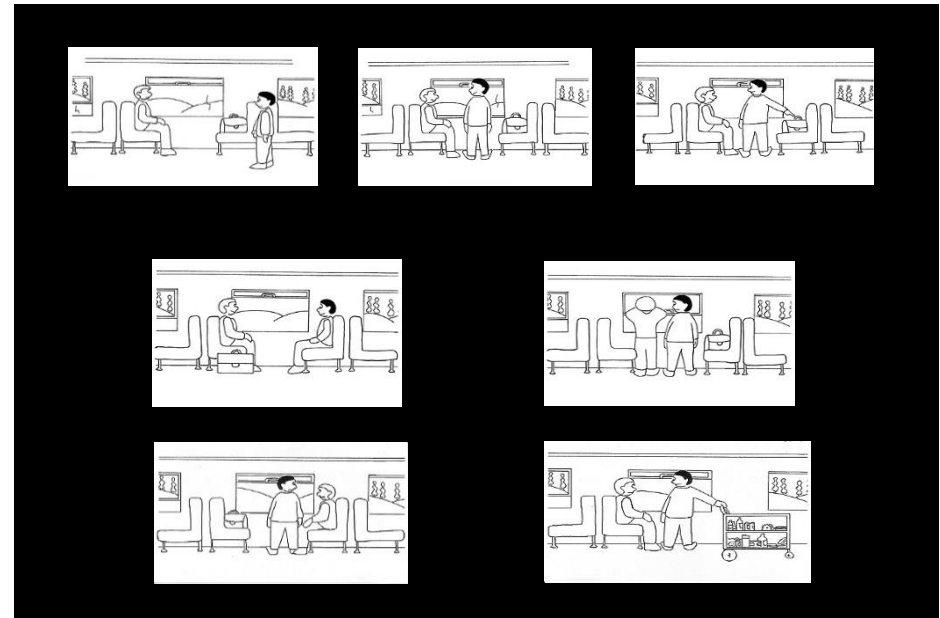
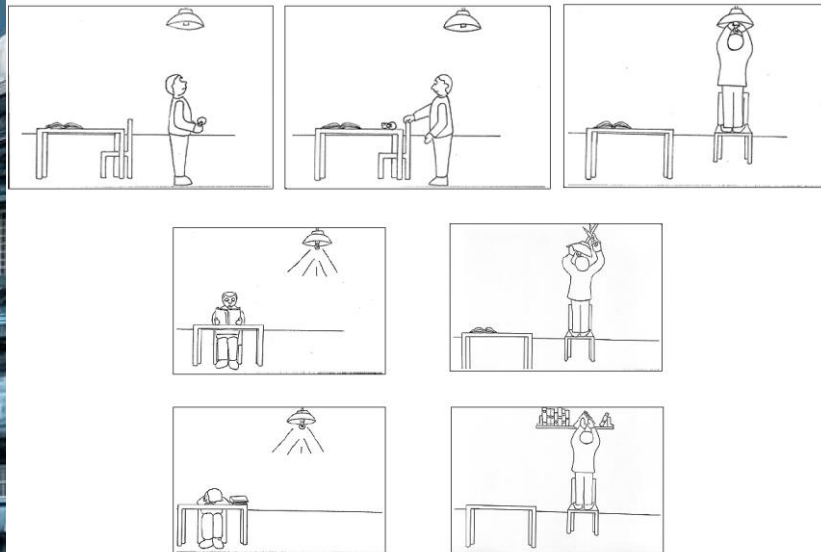
11/25 (44%) of ALS patients tested who displayed dysarthria showed language difficulties with in particular spelling problems.
Philipa Rewaj and Thomas Bak



Look for communication problems which are not due to physical dysarthria

Evidence of Social Understanding Impairment in Patients with Amyotrophic Lateral Sclerosis

Marco Cavallo^{1,2}, Mauro Adenzato^{3,4*}, Sarah E. MacPherson^{5,6}, Gillian Karwig⁵, Ivan Enrici³, Sharon Abrahams^{5,6,7}

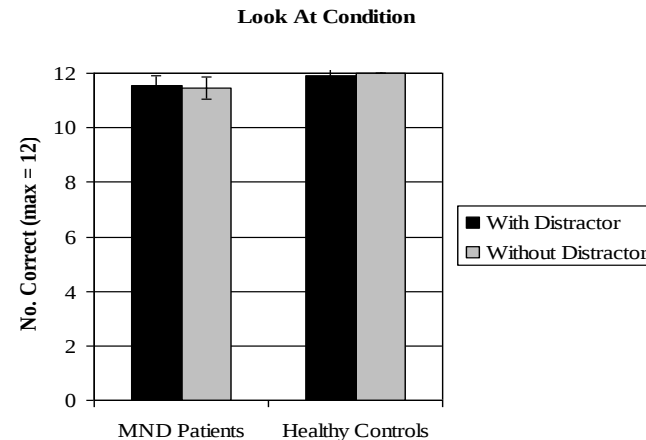
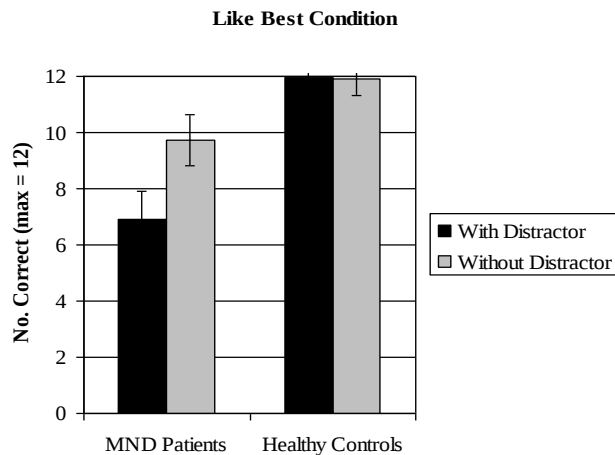
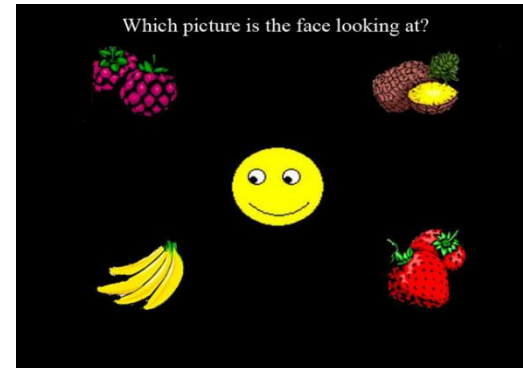
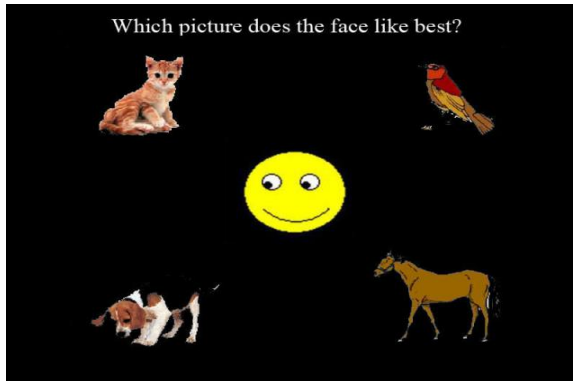


Social Cognition Deficit in ALS

Deficit in understanding social but not non-social situations

Deficits in Emotional and Social Cognition in Amyotrophic Lateral Sclerosis

Alessandra Girardi, Sarah E. MacPherson, and Sharon Abrahams
University of Edinburgh



36% of ALS impaired in simple theory of mind test
Increased egocentric responses in ALS.



Look for difficulties in social behaviour

How can we assess quickly and efficiently?

The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) in ALS





Cognitive assessment part of routine clinical care in the UK



Edinburgh Cognitive and Behavioural ALS Screen (ECAS)

Brief: 20 min assessment for ALS health professional

Designed for physical disability (written or spoken responses)

For health care professionals supervised by a neuropsychologist.

All people with ALS in Scotland are routinely offered to undertake this brief assessment. All nurses have been trained to undertake this assessment.

The results are incorporated into routine clinical care to help with management.

EDINBURGH COGNITIVE AND BEHAVIOURAL ALS SCREEN - ECAS
English Version (2013)

Date of testing: 03-08-2018
Age at leaving full-time education: 18
Occupation: BUILDING CONTRACTOR
Name: JOHN WHITE
Date of birth: 18-08-1956
Hospital No. or Address: _____
Handedness: RIGHT

LANGUAGE - Naming
Ask: Say or write down the names of these pictures.

Picture	Name	Score (1-4)
Scorpion	SCORPION	
Bow	BOW	
Helicopter	HELICOPTER	
Fox	FOX	
Axe	AXE	
Squirrel	SQUIRREL	
Swan	SWAN	
Book	BOOK	

LANGUAGE - Comprehension
Ask: point to the one which is:

Question	Options
1. Something you can fly in	1. Something with webbed feet
2. An animal that climbs trees	2. Something used for chopping
3. A means of transport	3. Something with a sharp edge
4. Something with a sting	4. Something with a diet of nuts and seeds



<https://ecas.psy.ac.uk>

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Edinburgh Cognitive and Behavioural ALS Screen (ECAS)

The ECAS is a brief multidomain assessment originally designed for people with Amyotrophic Lateral Sclerosis (ALS/Motor Neurone Disease) but is also useful in other neurodegenerative disorders.

It makes cognitive assessment fast and accessible and can be undertaken by a health care professional in the clinic or in a patient's home.

The ECAS was designed by Sharon Abrahams, Thomas Bak and Judy Newton.

ECAS News & Events

New Publication – September 2018 – ALS-specific cognitive and behavior changes associated with advancing disease stage in ALS

Check out ECAS research at the 29th International Symposium on ALS/MND, Glasgow

New Publication – May 2018 – Sensitivity and Specificity of the ECAS in Parkinson's Disease and Progressive Supranuclear Palsy

New Publication- July 2018- Factors influencing implementation of cognitive

15:52
30/09/2018

ECAS

Anterior functions/ALS-Specific:
language, executive and fluency)

Posterior functions/
ALS Non-Specific:
memory and visuospatial

Language

Naming

Comprehension

Spelling

EDINBURGH COGNITIVE AND BEHAVIOURAL ALS SCREEN – ECAS English Version (2013)	
Date of testing: Name: Age at leaving full-time education: Date of Birth: Occupation: Hospital No. or Address: Handedness:	
LANGUAGE - Naming	
Ask: Say or write down the names of these pictures:	
	
	
	
	
LANGUAGE - Comprehension Ask: point to the one which is:	
1. Something you can fly in 3. An animal that climbs trees 5. A means of transport 7. Something with a sting	2. Something with webbed feet 4. Something used for chopping 6. Something with a sharp edge 8. Something with a diet of nuts and seeds

S. Abrahams & T. H. Bak 1

LANGUAGE - Spelling	
Say: 'Spell, either by writing or speaking, the following words.' If the person is using assistive technology, ask them to turn off any predictive text facility.	
1. Envelope 3. Constructing 5. Biscuit 7. Deliver 9. Coathanger 11. Screwdriver	2. Skateboard 4. Partner 6. Lawnmower 8. Recorded 10. Orchestra 12. Brought

ECAS

- Letter Fluency

Free Fluency: S Words

Sail, Send, Sorry

Fixed Fluency: T 4L Word

That, Talk, Tank

Verbal Fluency Index

EXECUTIVE – Fluency Letter S																														
➤ Say: 'I am going to give you a letter of the alphabet and I would like you to say or write as many words as you can beginning with that letter, but not names of people or places, or numbers.' • If writing, say: 'You will have two minutes. The letter is S.' • If speaking, say 'You will have one minute. The letter is S.'			No. of correct words =																											
➤ Next the person copies/reads these words aloud. • If writing, say: 'copy these words as fast as possible. I will time you. Ready? Begin.' • If speaking, say: 'read aloud these words as fast as possible. Before you do this, check that you can read them. I will time you. Ready? Begin.'			Time to copy/ read aloud =																											
Verbal Fluency Index (VFI) calculation: If spoken: $VFI = \frac{60 \text{ seconds} - \text{no. of seconds to read aloud words}}{\text{No. of correct words generated}}$ If written: $VFI = \frac{120 \text{ seconds} - \text{no. of seconds to copy words}}{\text{No. of correct words generated}}$			<table border="1"> <thead> <tr> <th colspan="3">VFI conversion to score table</th> </tr> <tr> <th>SPOKEN VFI</th> <th>WRITTEN VFI</th> <th>Score</th> </tr> </thead> <tbody> <tr> <td>≥ 12.00</td> <td>≥ 20.00</td> <td>0</td> </tr> <tr> <td>10.00 to <12.00</td> <td>16.50 to < 20.00</td> <td>2</td> </tr> <tr> <td>8.00 to < 10.00</td> <td>13.00 to < 16.50</td> <td>4</td> </tr> <tr> <td>6.00 to < 8.00</td> <td>9.50 to < 13.50</td> <td>6</td> </tr> <tr> <td>4.00 to < 6.00</td> <td>6.00 to < 9.50</td> <td>8</td> </tr> <tr> <td>2.00 to < 4.00</td> <td>2.50 to < 6.00</td> <td>10</td> </tr> <tr> <td>< 2.00</td> <td>< 2.50</td> <td>12</td> </tr> </tbody> </table>	VFI conversion to score table			SPOKEN VFI	WRITTEN VFI	Score	≥ 12.00	≥ 20.00	0	10.00 to <12.00	16.50 to < 20.00	2	8.00 to < 10.00	13.00 to < 16.50	4	6.00 to < 8.00	9.50 to < 13.50	6	4.00 to < 6.00	6.00 to < 9.50	8	2.00 to < 4.00	2.50 to < 6.00	10	< 2.00	< 2.50	12
VFI conversion to score table																														
SPOKEN VFI	WRITTEN VFI	Score																												
≥ 12.00	≥ 20.00	0																												
10.00 to <12.00	16.50 to < 20.00	2																												
8.00 to < 10.00	13.00 to < 16.50	4																												
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4.00 to < 6.00	6.00 to < 9.50	8																												
2.00 to < 4.00	2.50 to < 6.00	10																												
< 2.00	< 2.50	12																												
Score 0-12			Score 0-12																											

ECAS

- Executive Functions

Reverse Digits

Alternation

Inhibitory Sentence

Completion

Social Cognition

1. The postman knocked on the 2. He brought his umbrella with him in case of 3. Sally spread her toast with butter and 4. John went to the barbers to get his hair 5. She dived into the swimming 6. They all went to the local café for something to	Score 0-12
Score 2 for different word, 1 for related word (e.g. associated or opposite meaning) or 0 for exact word.	
SOCIAL COGNITION – Part A	
Say: 'You are going to see some pictures, one in each corner of a box. You have to choose which picture you like best. Either point to or say which picture you like best. Please respond as quickly as possible.' Circle participant's choice.	
	
	
	

ECAS

- Executive Functions

Reverse Digits

Alternation

Inhibitory Sentence

Completion

Social Cognition

SOCIAL COGNITION – Part B

☞ Say: "You are going to see some pictures, one in each corner of a box. You have to choose which picture does the face like best. Either point to or say which picture the face likes best. Please respond as quickly as possible." Circle participant's choice. Correct items = 2 points, error = 1 point, egocentric error = 0 points.

Score
0-12



ECAS

- Memory

Story Recall

Immediate

Delayed Retention (%)

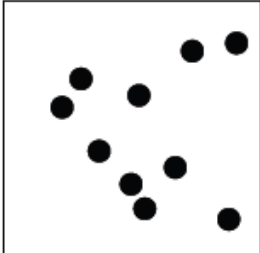
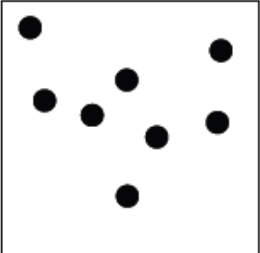
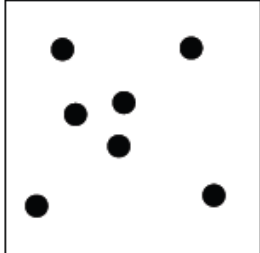
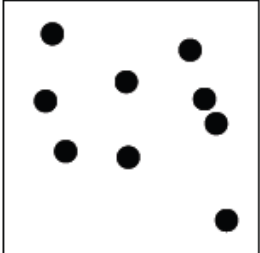
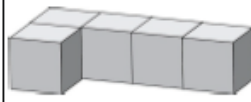
Recognition

MEMORY – Immediate recall		Score 0-10																																
☞ Say: 'I am going to read you a short story. Please listen carefully. When I am finished, say or write as much as you can remember'. Score 1 point for each (either entire or part of) underlined section recalled. <i>Last Sunday, the <u>annual litter collection</u> took place in <u>Primrose Woods</u>. <u>Forty two</u> people joined in to remove <u>old bicycles and shopping trolleys</u>. <u>Mr Douglas Watt</u> from the <u>woodland project</u> told local reporters that he was very <u>impressed and especially proud</u> of the <u>17 children</u> who came along.</i>		 Also use this score to calculate % retention later																																
MEMORY – Delayed Recognition		Score 0-4																																
If all items recalled, skip and score 4. Otherwise ask questions below. Say: 'Let's see if you can remember anything more about that story. I will ask you some questions, please tell me if they are true or false'. Circle responses (true or false) and score 1 point for each item recognised in this section. Use table below to calculate score. <table border="0"> <tr> <td>Was the story about an event that occurred last Saturday?</td> <td>T</td> <td>F</td> <td>1</td> </tr> <tr> <td>Was the event the annual litter collection?</td> <td>T</td> <td>F</td> <td>1</td> </tr> <tr> <td>Did this take place in Primrose Woods?</td> <td>T</td> <td>F</td> <td>1</td> </tr> <tr> <td>Did they remove old drink cans and sweet wrappers?</td> <td>T</td> <td>F</td> <td>1</td> </tr> <tr> <td>Was the man in the story called Mr Watt?</td> <td>T</td> <td>F</td> <td>1</td> </tr> <tr> <td>Was his first name 'Thomas'?</td> <td>T</td> <td>F</td> <td>1</td> </tr> <tr> <td>Was he from the local council?</td> <td>T</td> <td>F</td> <td>1</td> </tr> <tr> <td>Was he especially proud of the children for coming along?</td> <td>T</td> <td>F</td> <td>1</td> </tr> </table>		Was the story about an event that occurred last Saturday?	T	F	1	Was the event the annual litter collection?	T	F	1	Did this take place in Primrose Woods?	T	F	1	Did they remove old drink cans and sweet wrappers?	T	F	1	Was the man in the story called Mr Watt?	T	F	1	Was his first name 'Thomas'?	T	F	1	Was he from the local council?	T	F	1	Was he especially proud of the children for coming along?	T	F	1	
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8	4																																	

ECAS

- Visuospatial
- Dot Counting
- Cube Counting
- Number Location

VISUOSPATIAL – Number Location	
→ Say: 'Which number corresponds to the position of the dot?' 3 7 1 9 4 5 2 8 6 6 7 9 3 2 5 8 1	Score 0-4
5 8 2 1 3 4 6 9 7 6 2 9 4 3 8 1 5 7	

VISUOSPATIAL – Dot Counting		
→ Say: 'I would like you to count how many dots are in each box, but without pointing to them.'    	Score 0-4	
VISUOSPATIAL – Cube Counting → Say: 'How many cubes are in each structure, including the ones you may not be able to see?'    		Score 0-4

Screening for cognition and behaviour changes in ALS

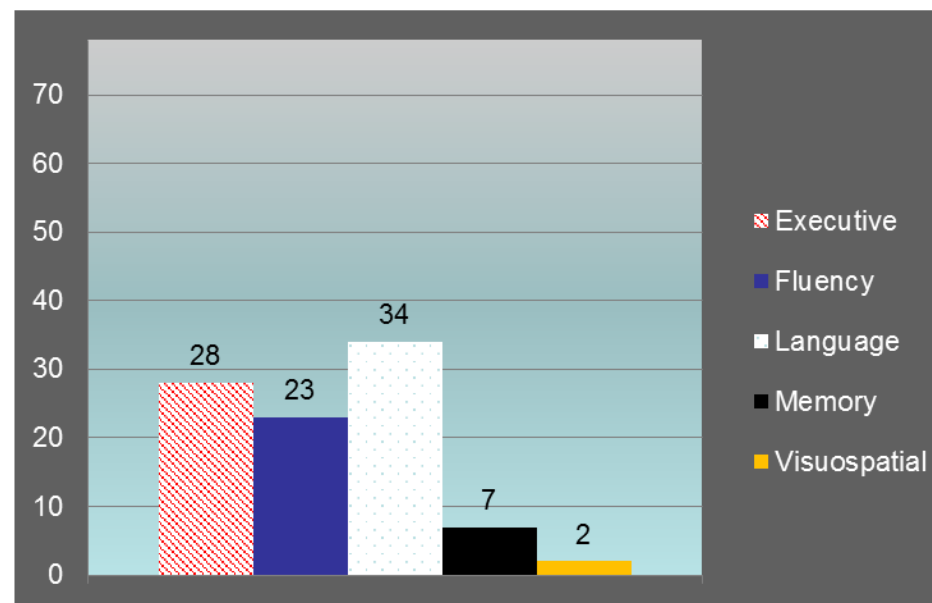
SHARON ABRAHAMS^{1,4}, JUDITH NEWTON^{1,3,4}, ELAINE NIVEN³, JENNIFER FOLEY³ & THOMAS H. BAK^{1,4}

ORIGINAL ARTICLE

Validation of the Edinburgh Cognitive and Behavioural Amyotrophic Lateral Sclerosis Screen (ECAS): A cognitive tool for motor disorders

ELAINE NIVEN¹, JUDITH NEWTON^{1,3,4}, JENNIFER FOLEY¹, SHUNA COLVILLE^{3,4}, ROBERT SWINGLER^{3,4}, SIDDHARTHAN CHANDRAN^{3,4}, THOMAS H. BAK^{1,2,3,4} & SHARON ABRAHAMS^{1,2,3,4}

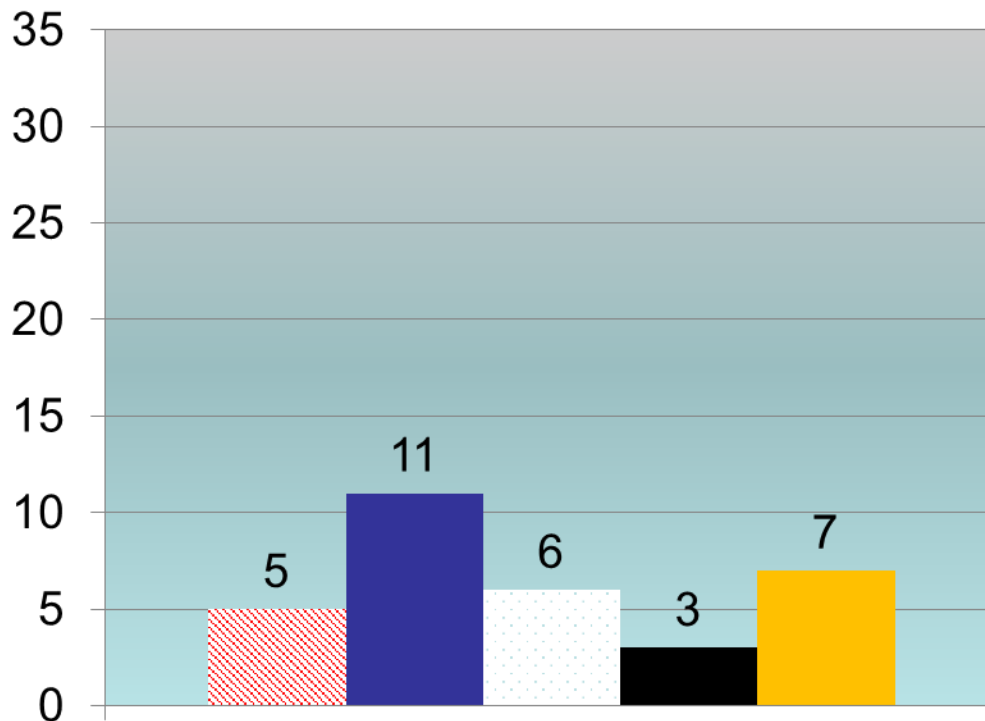
	Frequency of Deficit	% Abnormality
ALS-Specific	28	35.9%
ALS Non-Specific	6	7.7%
ECAS Total	28	35.9%



Scores 85% to 92 % sensitive and 85% specific against gold standard full neuropsychological assessment



ECAS behaviour screen



35 carer interviews, 51% of patients with behaviour change (1 or more domains)

EDINBURGH COGNITIVE AND BEHAVIOURAL ALS SCREEN – ECAS									
English Version (2013)									
BEHAVIOURAL SCREEN – Care Interview									
D Please ask the carer about the following possible behaviours. Symptoms should be present at least 12 months before the first diagnosis of ALS. If you are unsure, ask the carer. Yes, "Yes", "No", "Not sure", "Not known", "Not applicable". If you are unsure, please ask the carer to provide a brief verbal description for every symptom (maximum = 15).									
A Behavioural distribution									
1 Discrete hypochondriac behaviour ☐ Y ☐ N ☐ DK Hypochondriac behaviour with ongoing clinical behaviour									
2 Loss of memory or decision ☐ Y ☐ N ☐ DK State age, memory, decision, insight, insight, plans or options in relation to the future									
3 Irritability, rage or excessive anxiety ☐ Y ☐ N ☐ DK never, partially, or having or feeling properly related to the symptoms, or having or feeling depressed frequently, e.g. small and frequent									
4 Quality or results									
5 Loss of interest, drive or motivation ☐ Y ☐ N ☐ DK possible, active or spontaneous, never, partially or having or feeling properly related to the symptoms									
6 Loss of spontaneity or spontaneity									
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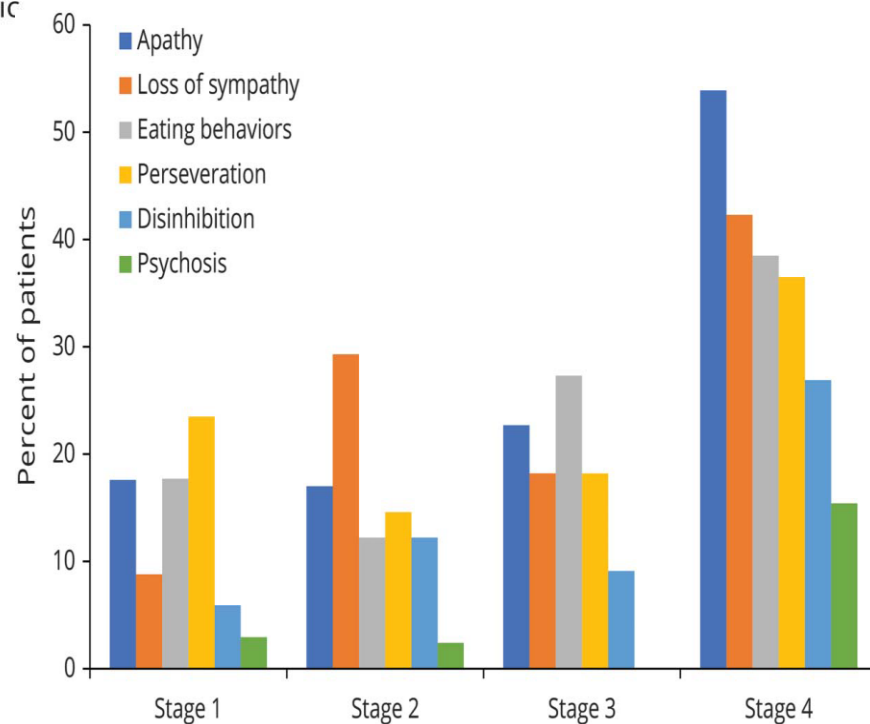
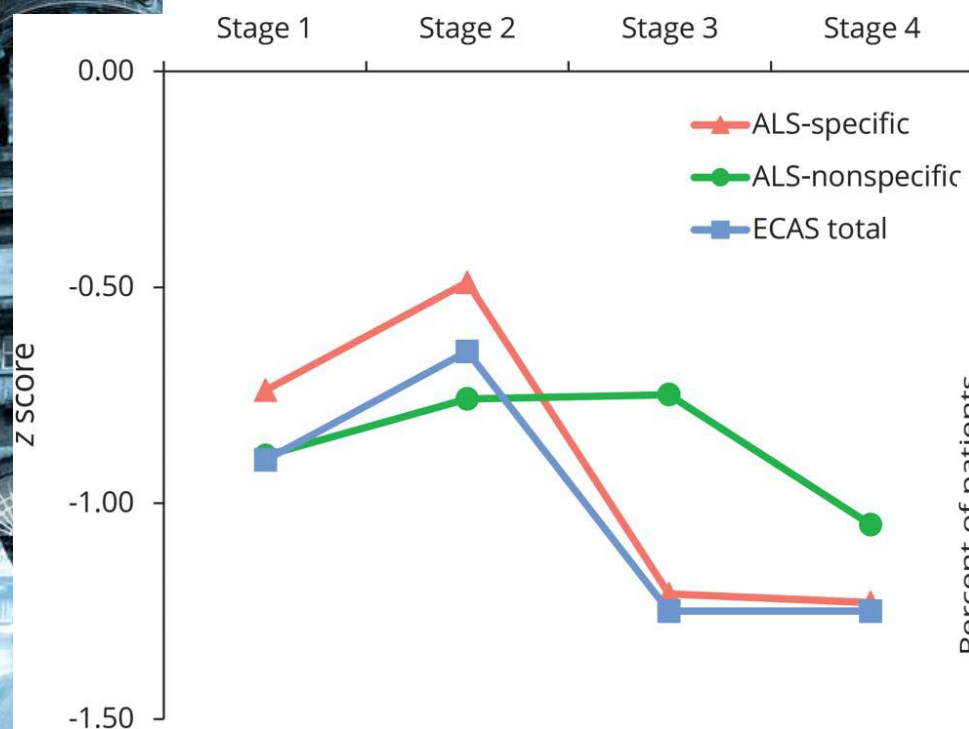
S. Abrahams & T. H. Bak 10

ALS-specific cognitive and behavior changes associated with advancing disease stage in ALS

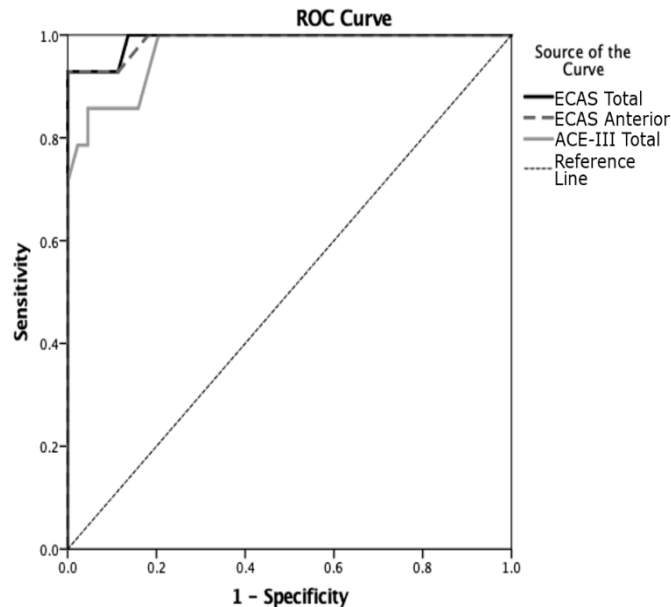
Christopher Crockford, PhD, Judith Newton, MSc, Katie Lonergan, BSc, Theresa Chiwera, MSc, Tom Booth, PhD, Siddharthan Chandran, MD, Shuna Colville, MPH, Mark Heverin, MSc, Iain Mays, BSc, Suvankar Pal, PhD, Niall Pender, PhD, Marta Pinto-Grau, MSc, Ratko Radakovic, PhD, Christopher E. Shaw, MD, Laura Stephenson, MSc, Robert Swingler, MD, Alice Vajda, PhD, Ammar Al-Chalabi, PhD, Orla Hardiman, MD, and Sharon Abrahams, PhD

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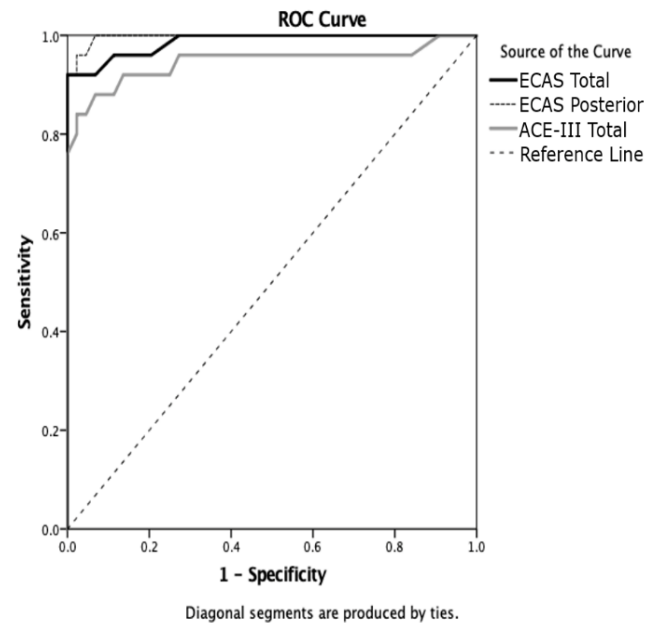


ECAS sensitive to bvFTD without ALS and AD



ECAS vs ACE-III in bvFTD

Anterior Score (executive, language, fluency) 94% sensitive 92% specific



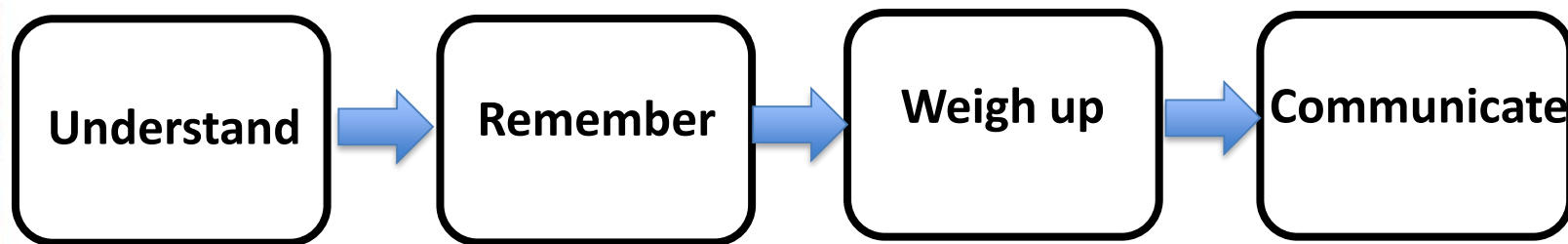
ECAS vs ACE-III in AD

Posterior Score (memory and visuospatial) 97% sensitive 96% specific

Mental capacity in ALS



- The ability to make a *specific* decision at a *specific time*.
- Capacity assumed unless demonstrated otherwise.
- Should I join this clinical trial?
- Should I consent to having a gastrostomy when/if the time comes?
- Should I consent to genetic testing



- Cognitive assessment (ECAS) can help to inform this process, but cannot tell you if someone has capacity to make a decision. You need to refer for a capacity assessment to a Psychiatrist/Psychologist.



Video case

Person with bvFTD



Poll

1. Did the woman in the video show...

Cognitive and Behaviour change

Cognitive change only

Behavioural change only

None of the above

We can't tell

2. Did the daughter's description of her mothers behaviour include

Disinhibition

Loss of sympathy/empathy

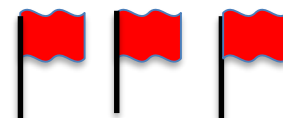
Apathy

Perseverative behaviour

All of the above



Summary of red flags



- ALS is not only a disease of the motor system
- 'Mother knows best', do a separate interview with someone who knows them well to check for behaviour change
- Be careful to consider other causes of behaviour abnormalities, e.g. marital discord...
- Do not dismiss psychiatric symptoms
- Take into account physical disability in the assessment
- Look for problems with attention, decision making and also language and social behaviour
- Use an appropriate assessment supervised by a neuropsychologist
- Remember some people will show behaviour change but unimpaired cognition, check both
- If someone is cognitively impaired does not mean that do not have mental capacity... Capacity assessments must be undertaken in full.



Thank you to

Research Team: Debbie Gray, Caroline McHutchison, Ratko Radakovic, Lewis Pettitt.

Thomas Bak

CARE-ALS Team: Judy Netwon, Suvankar Pal, Siddharthan Chandran,
Neuropsychologists Sharon Mulhern, Luke Williams

and all the people with ALS and their families who have participated
in our research

