

**EDINBURGH COGNITIVE AND BEHAVIORAL ALS SCREEN – UNIVERSITY OF PENNSYLVANIA
(ECAS-PENN)**

North American English Version Form B, Version 1.1 Revised (2020)

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Adapted for use in North America by Katya Rascovsky, Corey McMillan, and Murray Grossman, University of Pennsylvania, in collaboration with Michael Benatar on behalf of the Clinical Research in ALS and Related Disorders for Therapeutic Development (CReATe) Consortium.

Date of testing:
Occupation:
.....
Handedness:
Years of education:

Name/ID:
Date of Birth:
Highest Completed Degree:.....
Premorbid language difficulties

LANGUAGE - Naming

➡ Ask: Say or write down the names of these pictures:



.....



.....



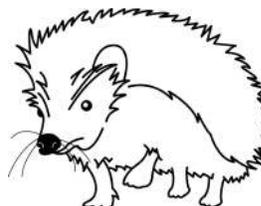
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.....



.....



.....



.....



.....

Score
0-8

LANGUAGE - Comprehension

➡ Ask: Point or say the one which is:

1. Something you wear.....
2. An animal with colorful feathers.....
3. Something you play.....
4. A means of transportation.....
5. Something a plumber might use.....
6. Something that rolls into a ball.....
7. Something used on a farm.....
8. Something that lives in the African savannah.....

Score
0-8

MEMORY – Immediate recall

- ☞ 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 according to the Administration and Guidance Notes.

Three fishing boats helped rescue a whale that swam too close to the shore. The whale was spotted swimming in circles. Alan Williams from the Marine Conservation Society said thirty-two young whales got lost last winter when looking for food.

Score
0-10

Also use this score to calculate % retained (bottom p.7)

LANGUAGE - Spelling

- ☞ Say: 'Spell, either by speaking or writing, the following words.' If the person is using assistive technology, ask them to turn off any predictive text facility. Give one point on "watermelon", "earthquake", "toothpick", and "sunflower" if the person inserts a "space" when spelling the word.

- | | |
|----------------------|----------------------|
| 1. Lecture | 2. Toothpick |
| 3. Gathering | 4. Pollution |
| 5. Receipt | 6. Sunflower |
| 7. Inspire | 8. Argued |
| 9. Watermelon | 10. Dictionary |
| 11. Earthquake | 12. Thought |

Score
0-12

FLUENCY - Letter F

Oral

Written

- ☞ Say: 'I am going to give you a letter of the alphabet and I would like you to say or write as many different words as you can beginning with that letter, but not names of people or places, or numbers.'

- If speaking, say: 'You will have **one** minute. The letter is F.'
- If writing, say 'You will have **two** minutes. The letter is F.'

No. of correct words
=

Time to read/
copy
aloud
=

Vfi =

- ☞ Next the person copies/reads these words aloud.

- 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.'
- If writing, say: 'copy these words as fast as possible. I will time you. Ready? Begin.'

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 < 13.00	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

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:

$$Vfi = \frac{120\text{seconds} - \text{no. of seconds to copy words}}{\text{No. of correct words generated}}$$

Score
0-12

EXECUTIVE – Reverse Digit Span

- ☞ Say: 'I am going to say some numbers and I would like you to say them back to me in reverse order. For example, if I say '2 3 4', you should say '4 3 2'. Let's practice. If I say '7 1 9', what would you say?' Stop when person gets both trials of a line wrong. Score total number of trials correct. After each pair of trials remind the participant 'Now the number of items will increase'.

Score
0-12

Trial		Check	Trial		Check
1	2 8		2	4 9	
3	5 1 3		4	8 2 4	
5	1 5 2 7		6	6 3 8 1	
7	9 4 3 8 6		8	2 4 1 9 7	
9	1 8 3 5 4 2		10	7 3 9 2 6 1	
11	4 8 2 6 8 3 1		12	5 1 6 4 2 9 3	

EXECUTIVE – Alternation

- ☞ Say: 'I want you to alternate between numbers and letters, starting with 1-A, then 2-B, 3-C, and so on. Please continue from there, alternating between numbers and letters, in order, without skipping any until I tell you to stop. Let's begin together: 1-A, 2-B, 3-C...'

Score
0-12

Trial		Check									
1	4-D		2	5-E		3	6-F		4	7-G	
5	8-H		6	9-I		7	10-J		8	11-K	
9	12-L		10	13-M		11	14-N		12	15-O	

FLUENCY - Letter D

Oral

Written

- ☞ Say: 'I am going to give you a letter of the alphabet and I would like you to say or write as many different words as you can beginning with that letter, but not names of people or places, or numbers. This time the word must only be **four letters** long. No more or less than four letters'
- If speaking, say 'You will have **one** minute. The letter is D.'
 - If writing, say: 'You will have **two** minutes. The letter is D.'

No. of
correct
words
=

Time to
read/
copy
aloud
=

Vfi =

- ☞ Next the person copies/reads these words aloud.
- 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.'
 - If writing, say: 'copy these words as fast as possible. I will time you. Ready? Begin.'

VFI conversion to score table

SPOKEN VFI	WRITTEN VFI	Score
≥ 20.00	≥ 35.00	0
16.75 to < 20.00	28.50 to < 35.00	2
13.50 to < 16.75	22.00 to < 28.50	4
10.25 to < 13.50	15.50 to < 22.00	6
7.00 to < 10.25	9.00 to < 15.50	8
3.75 to < 7.00	2.50 to < 9.00	10
< 3.75	< 2.50	12

Verbal Fluency Index (Vfi) calculation:

If spoken:

$$Vfi = \frac{60 \text{seconds} - \text{no. of seconds to read aloud words}}{\text{No. of correct words generated}}$$

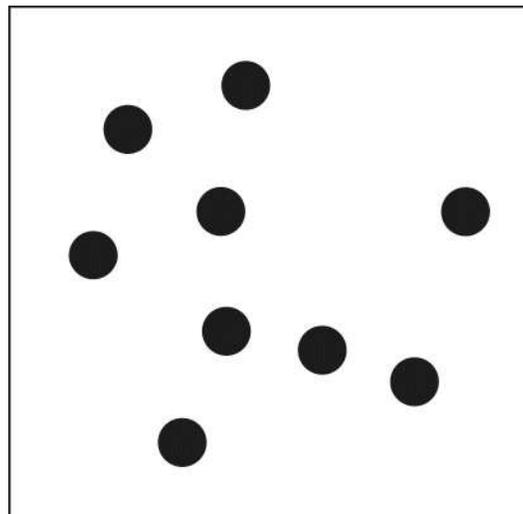
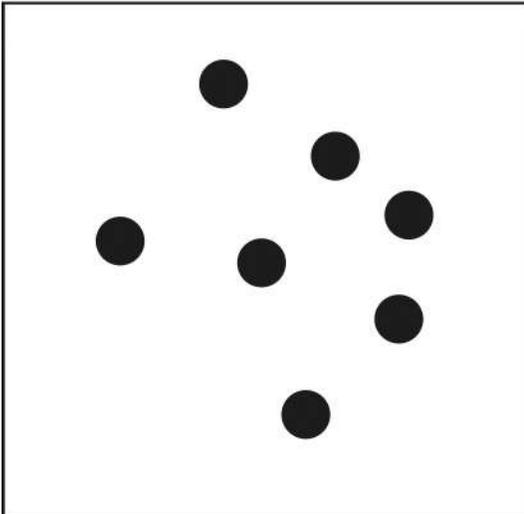
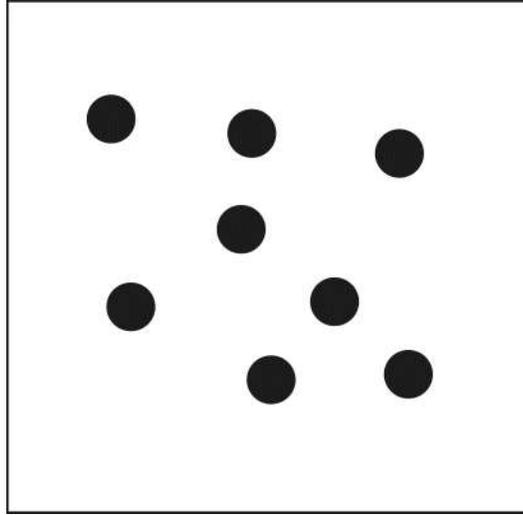
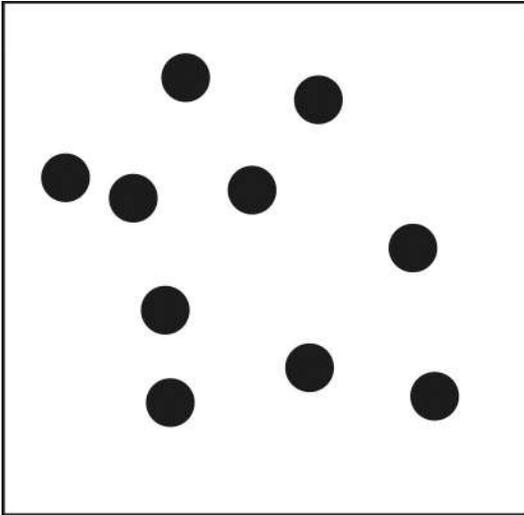
If written:

$$Vfi = \frac{120 \text{seconds} - \text{no. of seconds to copy words}}{\text{No. of correct words generated}}$$

Score
0-12

VISUOSPATIAL – Dot Counting

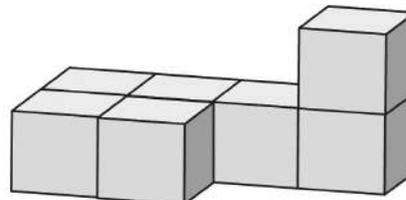
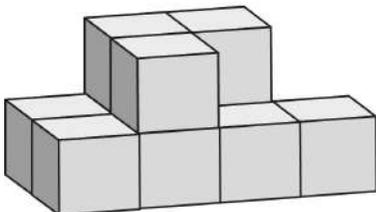
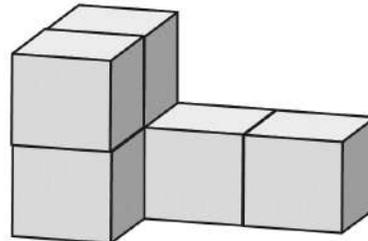
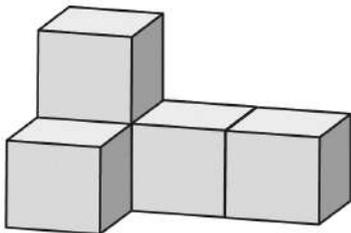
➤ Say: 'I would like you to count how many dots are in each box, but without pointing to them.'



Score
0-4

VISUOSPATIAL – Cube Counting

➤ Say: 'How many cubes are in each structure, including the ones you may not be able to see?'

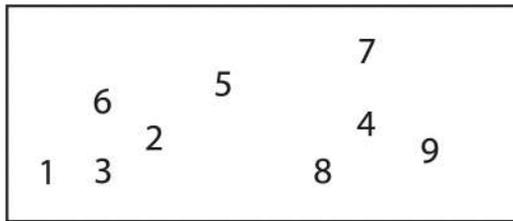
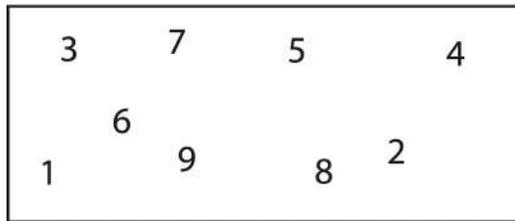
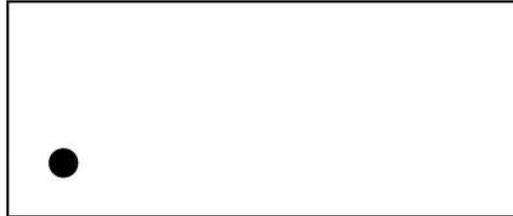
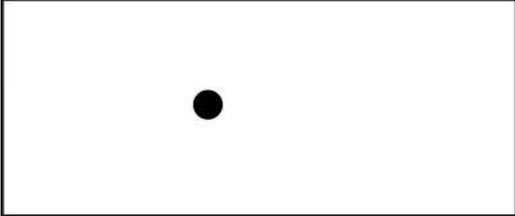
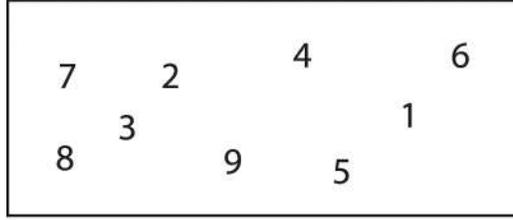
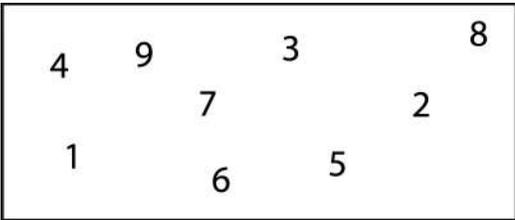


Score
0-4

VISUOSPATIAL – Number Location

➤ Say: 'Which number corresponds to the position of the dot?'

Score
0-4



EXECUTIVE – Sentence Completion

➤ Say: 'Listen carefully to these sentences and as soon as I have finished reading them, please tell me, or write, a word that finishes the sentence as quickly as possible. For example, '*She was so tired that she went straight to...bed*'. Do not score.

1. He called up the restaurant to reserve a
2. When she got up in the morning, the sun was.....

➤ Say: 'Now I'd like you to do that again, but this time the word you give should not make sense whatsoever in the context of the sentence. It must not be related to the word that actually completes the sentence. For example, '*John cut his hand with the sharp...orange*'. If the person does not respond within 20 seconds, move onto the next question.

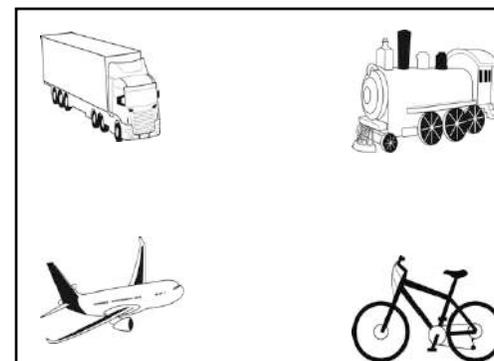
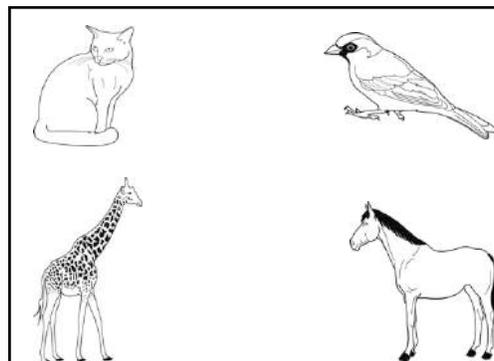
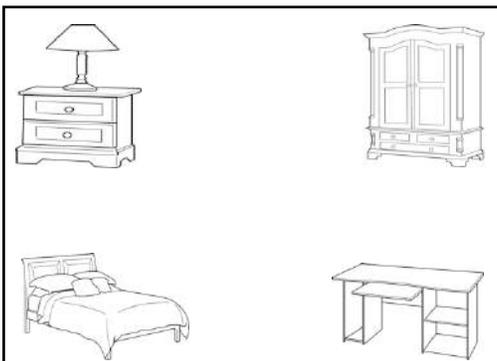
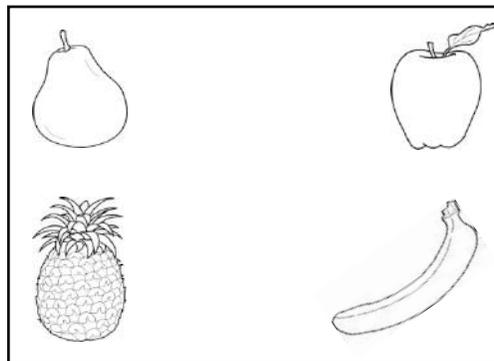
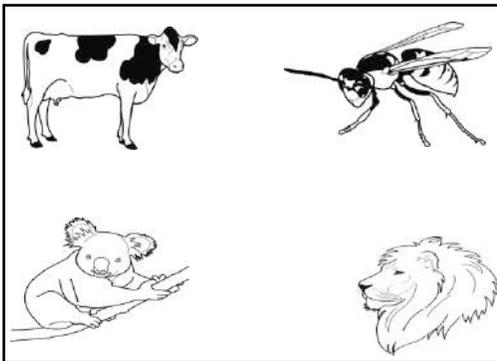
1. She answered the phone because it was
2. The joke was so funny, he started to
3. Daniel unlocked the door with a
4. The child cut paper with a pair of
5. After months of practice, Lisa passed her driving
6. Simon ate his dinner with a knife and

Score
0-12

Give 2 points for different word, 1 point for related word (e.g. associated or opposite meaning) or 0 points for exact word.

SOCIAL COGNITION – Part A

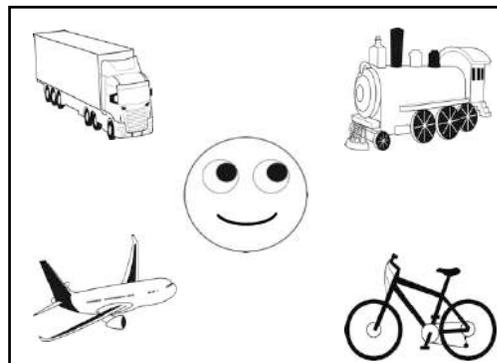
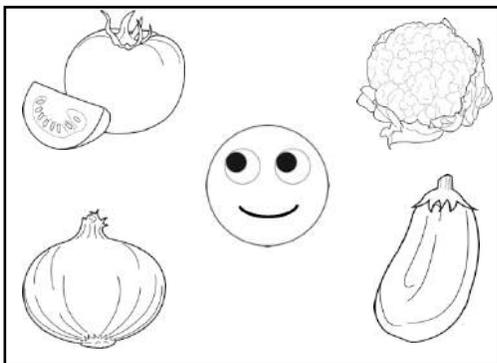
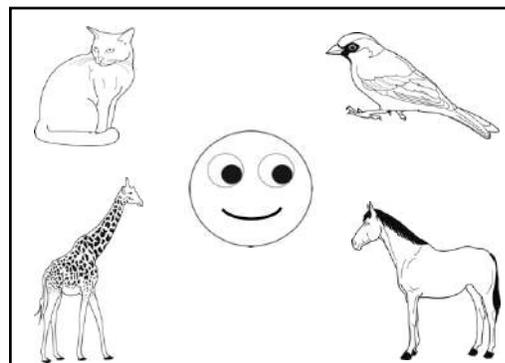
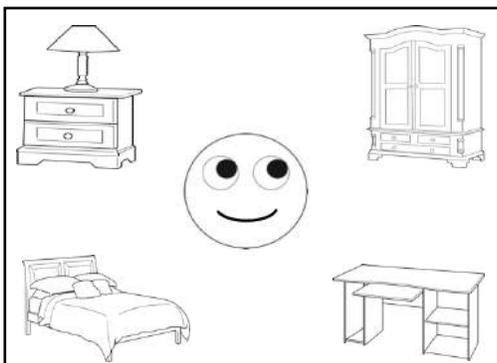
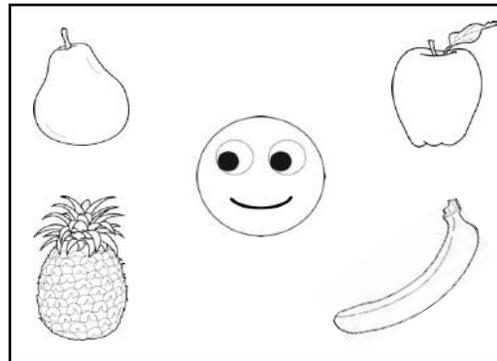
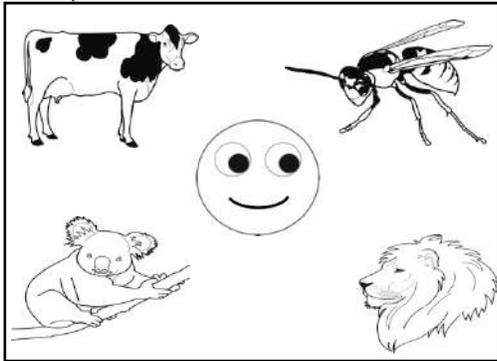
➤ Say: 'You are going to see some pictures, one in each corner of a box. You have to choose **which picture you like best**. Either point to or say which picture you like best. Please respond as quickly as possible.' Circle participant's choice.



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 the face likes 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



MEMORY – Delayed Recall

Scoring procedure for retention: obtain delayed recall performance (page 8) and, together with immediate recall score (page 2), determine percentage retained. Convert percentage retained to Score using table below. If delayed recall = 0, score = 0.

Delayed recall to percentage retained calculation

$$\frac{(\text{Delayed recall score [page 8]})}{(\text{Immediate recall score [page 2]})} \times 100 = \% \text{ retained}$$

(.....) x 100 =% retained

(.....)

Percentage retained to converted score table

Percentage retained	Score	Percentage retained	Score
1-10%	1	51-60%	6
11-20%	2	61-70%	7
21-30%	3	71-80%	8
31-40%	4	81-90%	9
41-50%	5	91-100+ %	10

☞ Say: 'At the beginning of this interview, I read you a short story. Tell me as much as you can remember from that story'. Score according to the Administration and Guidance Notes.

Delayed recall raw score (0-10)

Three fishing boats helped rescue a whale that swam too close to the shore. The whale was spotted swimming in circles. Alan Williams from the Marine Conservation Society said thirty-two young whales got lost last winter when looking for food.

Converted retention score (0-10)

MEMORY – Delayed Recognition

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 recognized in this section. Use table below to calculate score.

Score 0-4

Were there four fishing boats?	T	<u>F</u>	1
Was the story about rescuing dolphins?	T	<u>F</u>	1
Did this take place close to the shore?	<u>I</u>	F	1
Was the animal swimming in circles?	<u>I</u>	F	1
Was the man in the story called Mr. Williams?	<u>I</u>	F	1
Was his first name Stephen?	T	<u>F</u>	1
Were thirty-two whales lost last Summer?	T	<u>F</u>	1
Were the lost whales looking for food?	<u>I</u>	F	1

Yes = True (T)
No = False (F)

Recognition to recognition score table	
Number of correct answers	Score
0-4	0
5	1
6	2
7	3
8	4

SCORES

Language	Naming, Comprehension, Spelling	/28
Verbal Fluency	Fluency Letter F, Fluency Letter D	/24
Executive	Reverse Digit Span, Alternation, Sentence Completion, Social Cognition	/48
ALS-SPECIFIC:		/100
Memory	Immediate recall, Delayed recall score, Delayed recognition	/24
Visuospatial	Dot Counting, Cube Counting, Number Location	/12
ALS NON-SPECIFIC:		/36
ECAS TOTAL SCORE:		/136

**EDINBURGH COGNITIVE AND BEHAVIORAL ALS SCREEN – UNIVERSITY OF PENNSYLVANIA
(ECAS-PENN)
American English Form A, Version 1 (2018)**

BEHAVIOR SCREEN – Caregiver Interview

➤ Please ask the caregiver about the following possible behaviors. Symptoms should have occurred repeatedly and not just on one instance, and may have occurred prior to the development of any motor signs. Tick 'Yes', 'No' or 'Don't Know'. If 'Yes', please provide a brief written description. Give one point for every 'Yes' response (maximum = 10).

A Behavioral disinhibition				
1	Socially inappropriate behavior, e.g. <i>inappropriate behavior with strangers criminal behavior</i>	Y	N	DK
2	Loss of manners or decorum, e.g. <i>crude or sexually explicit remarks, jokes or opinions that may be offensive to others lack of response to social cues</i>	Y	N	DK
3	Impulsive, rash or careless actions, e.g. <i>new onset gambling, or buying or selling property without regard for consequences giving out personal information inappropriately, e.g. credit card numbers</i>	Y	N	DK
B Apathy or inertia				
4	Loss of interest, drive or motivation, e.g. <i>passivity and lack of spontaneity needs prompting to initiate or continue routine activities</i>	Y	N	DK
C Loss of sympathy or empathy				
5	Diminished response to other people's needs and feelings <i>Positive rating on this feature should be based on specific examples that reflect a lack of understanding or indifference to other people's feelings, e.g. hurtful comments disregard for others' pain or distress</i>	Y	N	DK
6	Diminished social interest, interrelatedness, personal warmth or general closeness in social engagement, e.g. <i>coldness lack of eye contact</i>	Y	N	DK
D Perseverative, stereotyped, compulsive or ritualistic behavior				
7	Simple repetitive movements, e.g. <i>tapping, clapping scratching, picking skin or clothing repeating words</i>	Y	N	DK
8	Complex, compulsive or ritualistic behaviors, e.g. <i>counting, cleaning rituals, checking collecting, hoarding</i>	Y	N	DK

BEHAVIOR

E Hyperorality and altered food preferences					
9	Altered food preferences, e.g. <i>food fads</i> <i>carbohydrate craving (particularly sweets)</i>	Y	N	DK	
10	Binge eating or hyperorality, e.g., <i>cramming or continuing to eat despite satiety</i> <i>oral exploration or consumption of inedible objects</i>	Y	N	DK	
SCORE					
TOTAL				/10	
SYMPTOMS					
➡ Please check box if at least one of the symptoms was present in each of the following categories.					
A. Behavioral disinhibition					
B. Apathy or inertia					
C. Loss of sympathy or empathy					
D. Perseverative, stereotyped, compulsive or ritualistic behaviour					
E. Hyperorality and altered food preferences					
ALS Psychosis Screen					
➡ Please ask the caregiver about the following possible symptoms. Check 'Yes', 'No' or 'Don't Know'. If 'Yes', please provide a brief written description. Give one point for every 'Yes' response (maximum = 3).					
1	Has strange and/or bizarre beliefs and behaviors	Y	N	DK	
2	Hears or sees things that are not there, and/or feels the presence of someone who is not there	Y	N	DK	
3	Is overly suspicious, and/or feels persecuted	Y	N	DK	
SCORE					
TOTAL				/3	
ONSET AND DURATION OF SYMPTOMS					
➡ Please check or complete box to indicate response.					
1. Do these symptoms represent a CHANGE from the patient's previous behavior?				Y	N
If yes, did the changes occur:					
a. BEFORE the onset of the disease?				Y	N
b. at the same time as other symptoms?				Y	N
c. AFTER the onset of the disease?				Y	N
2. Do they still persist?				Y	N
3. If not, how long did they last?					