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RESEARCH ARTICLE

The Edinburgh Cognitive and Behavioral ALS Screen (ECAS) in frontotemporal dementia

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Abstract

Objectives: To examine the usefulness of the Edinburgh Cognitive and Behavioral Amyotrophic Lateral Sclerosis (ALS) Screen (ECAS) as a cognitive screening tool for the detection of behavioral variant frontotemporal dementia (bvFTD). A secondary aim was to determine whether people with FTD combined with ALS (ALS-FTD) exhibit a similar ECAS profile to that of people with bvFTD alone. *Methods:* Patients with ALS-FTD and bvFTD and healthy controls were recruited. Participants were administered the ECAS, which comprises tests of language, verbal fluency, executive functions, memory, and visual-spatial functions. They also carried out analogous, full-length cognitive tests that examine naming, spelling, sentence completion, and social cognition skills. *Results:* The study cohort comprised 20 ALS-FTD patients, 23 with bvFTD, and 30 controls. Highly significant group differences were elicited for all cognitive domains, reflecting poorer performance in patients compared to controls. No significant differences in overall test scores were found between ALS-FTD and bvFTD patients, although ALS-FTD patients showed a higher frequency of impairment on verbal fluency. Correlative analyses revealed inter-relationships in patients (but not controls) between scores in different domains, most marked in bvFTD. There were strong correlations between performance on ECAS subtests and analogous cognitive tasks. *Conclusion:* The ECAS is a sensitive and valuable tool for the assessment of FTD. Executive, language and behavioral breakdown may, however, compromise performance in other cognitive domains, reducing the specificity of the 'frontotemporal' cognitive profile. Subtle differences observed between ALS-FTD and bvFTD raise questions regarding the precise relationship between bvFTD with and without ALS.

Keywords: *Motor neurone disease, frontotemporal dementia, cognition*

Introduction

The Edinburgh Cognitive and Behavioral ALS Screen (ECAS) (1) is a well-established screening instrument for detecting cognitive impairments in amyotrophic lateral sclerosis (ALS). It has been translated into multiple languages (2–6) and validated in different populations (2,7–10), showing impressive levels of sensitivity and specificity. Its relative brevity means that the ECAS is appropriate for use with patients who are unable to tolerate lengthy cognitive assessments.

The ECAS is founded on the recognized link between ALS and frontotemporal dementia (FTD). ALS patients show deficits in the same

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domains of cognitive function most affected in FTD (11): language, verbal fluency, executive functions, and social cognition. A key strength of the ECAS is its distinction between those "ALS-specific" domains, mediated by frontotemporal lobe functioning and most likely to be affected in ALS, and other cognitive domains not thought to be specific to ALS: memory and visuospatial functions. Performance profiles offer the potential to distinguish ALS cognitive impairment from that of other neurodegenerative disorders such as Alzheimer's disease (AD) (6) and to have more widespread implications for differential diagnosis of neurodegenerative disease, according to the likelihood of frontotemporal symptomatology (12).

ECAS studies have hitherto focused primarily on ALS patients who exhibit cognitive impairment (ci), behavioral impairment (bi) or both (cbi), according to current definitions of frontotemporal spectrum disorder (11), but who do not meet full criteria for behavioral variant FTD (bvFTD) (13). There are relatively few data pertaining specifically to people with established bvFTD, who by definition, have more severe cognitive/behavioral impairment than the majority of people with ALS.

The primary purpose of the present study was to examine the usefulness of the ECAS as a cognitive screening tool for the detection of bvFTD. It explores the relationship between ALS-specific and ALS non-specific cognitive domains because of the potential for interactions between performance in different cognitive domains, particularly executive functions and memory. A subsidiary aim was to determine whether people with FTD combined with ALS (ALS-FTD) exhibit an ECAS profile identical to or different from that of people with bvFTD alone.

The study forms part of a broader investigation of cognitive and behavioral changes in ALS-FTD and bvFTD, supported by the Motor Neurone Disease Association.

Method

Participants

The study cohort comprised patients clinically diagnosed with bvFTD or ALS-FTD, recruited from specialist cognitive and motor neurone disease clinics within the NorthWest of England. Patients were included if they fulfilled contemporary diagnostic criteria for bvFTD (13) and were in the mild-to-moderate stage of the disease, as measured by the Clinical Dementia Rating scale modified for use with FTD patients (14). ALS-FTD patients also fulfilled the El Escorial criteria for ALS (15). ALS-FTD patients were excluded if they fell into the 'very severe' range of disability (score < 12), as measured by the ALS Functional Rating Scale revised (16), or if they required mechanical respiratory support. Healthy controls were recruited via a local research register of volunteers and through the national Join Dementia Research initiative. Participants were excluded if they had a history of head injury, alcohol or substance abuse, symptoms, and signs of cerebrovascular disease or clinically significant anxiety or depression. All participants were native English speakers. The study was approved by the North West Ethics committee (REC reference 14/NW/1185). Participants (together with their carer in the case of patients) provided informed written consent to participate and for the future publication of fully anonymized material pertaining to them.

Cognitive assessment

The ECAS addresses five domains of function: language, verbal fluency, executive skills, memory, and visuospatial skills. It encompasses the following subtasks: picture naming, comprehension, spelling, generation of words beginning with S, generation of four-letter words beginning with T, digit reversal, the alternation between numbers and letters, unconnected sentence completion, social cognition (judgment of preference), immediate recall of the story, retention over the delay, recognition memory, dot counting, cube counting, and number location. The test was administered according to published guidelines (https://ecas.psy.ed.ac.uk), with patients being offered the opportunity to choose between oral and written modes of response to optimize accessibility for ALS patients with reduced bulbar/limb function. For verbal fluency, a verbal fluency index was calculated to adjust for motor deficits.

To enable direct comparison of performance on ECAS language, executive and social cognition tasks with that of standard full-length neuropsychological tests that assess the same cognitive domains, the study also included the following tests: Graded Naming test (17), a 30-item picture naming test of graded difficulty; PALPA spelling (18); the accuracy score from part B of the Hayling sentence completion test (19) that taps generation and response inhibition and a Judgment of Preference from eye gaze test (20,21) that is a marker of social cognition.

All patient assessments were conducted by the same administrator (JAS) and control tests by JAS, JCT, or JMH. Testers were trained in ECAS administration and had years of experience in the clinical assessment of patients with FTD.

Behavioral screen

The ECAS includes a behavioral screen, which covers the core domains of behavioral change specified in diagnostic criteria for bvFTD (13). Since all patients, by definition, fulfilled behavioral

criteria for bvFTD, the behavioral screen was not evaluated as part of the current study.

Statistics

Statistical analyses were carried out using IBM SPSS Statistics version 25. Group comparisons involved chi-squared tests for categorical data, with Fisher's Exact test applied when cells had expected frequencies below 5. Analysis of variance was used for interval data and Kruskal–Wallis and Mann–Whitney U tests for ECAS measures, for which data were not normally distributed. Significance values shown in tables are uncorrected. They survive Bonferroni correction for multiple comparisons except where stated in the text. The effect size was calculated using the formula $r = Z_{\sqrt{}}n$ as described by Rosenthal (22). Control percentile scores were calculated to determine impaired performance in individual patients, defined as lower than the 5th percentile of control performance. Correlations between tests were carried out using Spearman's rho test. Significance levels are uncorrected.

Results

Demographics

Forty-three patients fulfilled the criteria for the study within the recruitment period, 23 with bvFTD, and 20 with ALS-FTD. Thirteen ALS-FTD patients had some degree of bulbar involvement at the time of testing. Thirty healthy controls were recruited via a local research register of volunteers and through the national Join Dementia Research initiative. There was a preponderance of male patients in the patient groups (Table 1). ALS-FTD patients were older than controls, whereas bvFTD patients showed no significant difference in age. bvFTD patients had fewer years of education than controls, whereas years of education did not differ significantly between the ALS-FTD group and either the bvFTD or control group.

Group comparison of ECAS domain and total scores

An examination of ECAS domain, ALS-specific, ALS nonspecific, and total scores revealed highly significant group differences for all cognitive domains, based on Kruskal–Wallis tests (Table 2). Post hoc Mann–Whitney U tests showed that these differences lay exclusively between patients and controls. The bvFTD and ALS-FTD groups each performed more poorly than controls, with significance levels at $p < 0.001$ for all domains. No differences in scores between bvFTD and ALS-FTD groups reached conventional levels of significance of $p < 0.05$. The highly significant differences between patient groups and controls ($p < 0.001$) remained when younger and more educated

Table 1. Demographics Characteristics.

	Patients				
	ALS-FTD	bvFTD	Controls		*p*-Value
Number	20	23	30		
Gender, M:F	12:8	14:9	9:21	$\chi^2 = 6.58$	0.04
Age, mean (SD)	65 (8)	60 (7)	59 (8)	$F = 3.89$	0.03^
Years of education, mean (SD)	13 (3)	12 (3)	14 (3)	$F = 3.86$	0.03*
Duration, mean years (SD)	3 (1)	5 (4)	n/a	$t = 2.6$	0.02
Dementia severity[1]	9.5 (2.8)	9.3 (2.8)	n/a	$t = 0.26$	0.80

^Differences lie between ALS-FTD and control (post-hoc Bonferroni test $p = 0.03$). *Differences lie between bvFTD and control (post-hoc Bonferroni test $p = 0.02$). Other comparisons non-significant.
[1]Modified Clinical Dementia Rating, mean number of boxes (14).

Table 2. Group comparisons of ECAS domains.

	Patients				
	ALS-FTD	bvFTD	Controls	Statistic	Group analysis
	Median (range)	Median (range)	Median (range)	Kruskall–Wallis	*p*-Value
Language/28	21 (5–28)	20 (1–28)	28 (25–28)	35.3	<0.001[a,b]
Verbal fluency/24	3 (0–14)	8 (0–18)	20 (6–24)	42.8	<0.001[a,b]
Executive/48	19.5 (4–43)	23.5 (4–42)	40.5 (30–45)	32.9	<0.001[a,b]
Memory/24	7 (0–16)	10 (0–21)	18 (12–22)	34.5	<0.001[a,b]
Visuospatial/12	11 (3–12)	11 (0–12)	12 (11–12)	21.2	<0.001[a,b]
ALS total/100	45 (12–81)	55.5 (14–87)	89 (72–95)	40.0	<0.001[a,b]
Non-ALS total/36	19 (6–27)	16 (0–32)	30 (24–34)	36.7	<0.001[a,b]
ECAS total/136	65 (18–98)	78 (15–119)	118 (100–127)	40.3	<0.001[a,b]

Post-hoc Mann–Whitney test significant at $p < 0.001$ between [a]ALS-FTD and controls and [b]bvFTD and controls. No significant differences between ALS-FTD and bvFTD were observed.

Table 3. ECAS subtests: FTD patients (ALS-FTD + bvFTD) compared to controls.

	FTD		Controls		Mann–Whitney			
Test	Median	Range	Median	Range	U	z	p-Value	Effect size
ALS-specific								
Naming/8	6	1–8	8	7–8	177.0	−5.5	<0.001	−0.65
Comprehension/8	8	0–8	8	8–8	375.0	−4.0	<0.001	−0.47
Spelling/12	8	0–12	12	10–12	199.5	−5.0	<0.001	−0.60
Fluency letter S/12	1	0–10	10	4–12	60.0	−6.4	<0.001	−0.78
Fluency letter T/12	0	0–10	10	4–12	73.0	−6.3	<0.001	−0.77
Reversed digits/12	4	0–8	8	4–10	151.5	−5.5	<0.001	−0.65
Alternation/12	6	0–12	12	5–12	222.0	−4.7	<0.001	−0.56
Sentence completion/12	4.5	0–11	11	4–12	127.0	−5.7	<0.001	−0.68
Social cognition/12	6	0–12	12	6–12	277.5	−3.9	<0.001	−0.47
ALS nonspecific								
Immediate recall/10	2	0–7	7	4–10	105.0	−6.1	<0.001	−0.71
Retention over delay/10	3	0–10	9	4–10	316.0	−3.6	<0.001	−0.42
Delayed recognition/4	1	0–4	2	0–4	270.5	−3.9	<0.001	−0.47
Dot counting/4	4	0–4	4	3–4	437.5	−2.9	0.004	−0.34
Cube counting/4	3.5	0–4	4	3–4	315.0	−4.2	<0.001	−0.50
Number location/4	4	0–4	4	3–4	450.0	−2.7	0.007	−0.32

controls were excluded so that the groups were matched for age and education.

Not all patients were able to complete all tasks. In the bvFTD group, patients' behavioral/cognitive disorder precluded assessment of verbal fluency in one case and of executive function in three cases. In the ALS-FTD group, six patients could not be assessed on at least one domain because of a combination of behavioral/cognitive and physical difficulty: two for language, six for verbal fluency, four for executive functions, three for memory, and three for visuospatial function. In those patients, the absence of data for one or more tasks precluded the calculation of a meaningful overall domain score (although 'impairment' could be inferred).

Patients' total scores for ALS-specific and ALS-nonspecific tasks were expressed as a proportion of the maximum possible score (100 and 36, respectively). A Wilcoxon test showed no difference in relative performance for these two broad aspects of cognitive function in either patient group: ALS-FTD $z = -0.63$, $p = 0.53$; bvFTD $z = -0.52$, $p = 0.60$.

Subdomain performance in FTD

Patients' impaired performance in non-ALS specific, as well as ALS-specific domains, is notable. It raises the question of whether there are particular subtests within each domain that are particularly vulnerable to impairment. Performance on individual subtests is shown in Table 3. In view of the absence of significant domain differences between ALS-FTD and bvFTD groups these patient groups are considered together as a composite FTD group for subdomain analyses and compared to controls. Each subtest within the domains of language, verbal fluency, and executive functions elicited significant group differences, most at $p < 0.001$ (Table 3). Following Bonferroni correction for multiple comparisons ($p < 0.003$) differences on dot counting and number location tests no longer reached statistical significance. The largest effect size was elicited for verbal fluency, immediate recall, and inhibitory sentence completion.

Frequency of impairment

Table 4 shows the percentage of ALS-FTD and bvFTD patients whose performance fell below the 5th percentile of control group scores, and on that basis can be considered impaired. For verbal fluency, one outlier control score was excluded in order to normalize the distribution upon which percentiles were calculated. Both ALS-FTD and bvFTD groups showed a high frequency of impairment in all cognitive domains. There was a higher frequency of impairment in ALS-FTD than bvFTD for verbal fluency (Fisher's Exact ($n = 43$), $p = 0.04$). Frequency of impairment in the two patient groups ($n = 43$) did not differ significantly for other domains: Language Fisher's Exact, $p = 0.73$, Executive Fisher's Exact $p = 0.25$, Memory $\chi^2(1, n = 43) = 0.97$, $p = 0.32$, visuospatial skills $\chi^2 = 0.05$, $p = 0.82$ ALS-specific total Fisher's Exact, $p = 0.42$, ALS nonspecific $\chi^2 = 1.16$, $p = 0.28$, Total ECAS Fisher's Exact $p = 0.11$.

In the bvFTD group, ALS-specific domain impairment was slightly more common than for ALS nonspecific domains (Fisher's Exact test ($n = 23$), $p = 0.03$). Differences in these domains did not reach significance in the ALS-FTD group (Fisher's Exact test ($n = 20$), $p = 0.63$). Overall, the ALS-specific domain showed high sensitivity in detecting impairment: ALS-FTD 90%, bvFTD

Table 4. Percentage of patients showing impairment.

Criterion	Below 5th percentile of control scores		Using 2014 cutoffs (1)		Using age and education adjusted cutoffs (9)	
Domain	ALS-FTD	bvFTD	ALS-FTD	bvFTD	ALS-FTD	bvFTD
Language	80	74	85	78	65	61
Verbal fluency	95	70	100	70	95	78
Executive	90	74	90	74	80	70
Memory	75	61	80	78	60	61
Visuospatial	40	44	40	44	35	43
ALS-specific	90	78	95	87	85	70
ALS nonspecific	80	65	80	74	60	57
Total/136	100	83	100	91	85	74

78%, but low specificity in relation to ALS-nonspecific impairment: ALS-FTD 20%, bvFTD 35%.

To enable direct comparison with studies of ALS, the percentage of impaired patients, based on originally published cutoffs (1) and revised age and education-adjusted cutoffs (9) are also shown in Table 4. The latter seemed particularly relevant in view of differences in age and education between patients and controls. Frequencies of impairment according to these different criteria yield a coherent pattern, with slightly more conservative estimates of impairment typically elicited when age and education adjusted norms are applied.

Correlation between domains

Domain scores showed no significant inter-correlations in the control group, possibly reflecting ceiling level scores on some tasks. In the patient groups, significant inter-correlations were apparent, particularly marked in bvFTD (Table 5). Inter-correlations in ALS-FTD were generally non-significant, the notable exception being the strong relationship between verbal fluency and executive scores ($r_s(11) = 0.75$, $p = 0.003$). In bvFTD, strong correlations between language and memory performance were present for immediate recall ($r_s(21) = 0.74$, $p < 0.001$), delayed recall ($r_s(21) = 0.69$, $p < 0.001$) and delayed recognition ($r_s(21) = 0.57$, $p = 0.005$), whereas correlations between executive performance and memory were mainly driven by the immediate recall ($r_s(18) = 0.57$, $p = 0.008$). Executive performance and delayed recall showed a more modest association ($r_s(18) = 0.52$, $p = 0.02$) and delayed recognition no significant relationship ($r_s(18) = 0.29$, $p = 0.22$).

ECAS scores in relation to standard neuropsychological tests

There were strong correlations between patients' performance on ECAS language and executive tests and their corresponding full version neuropsychological test: ECAS naming and Graded Naming ($r_s(39) = 0.65$, $p < 0.001$); ECAS spelling and PALPA spelling ($r_s(34) = 0.86$, $p < 0.001$; ECAS sentence completion and Hayling test ($r_s(29) = 0.68$, $p < 0.001$); ECAS social cognition and Judgment of Preference ($r_s(30) = 0.64$, $p < 0.001$). The sensitivity of the task in detecting impairment, based on control cutoff scores, was broadly similar, albeit slightly higher for the full test version: ECAS naming 56%, Graded naming 68%; ECAS spelling 61%, PALPA spelling 64%; ECAS sentence completion 55%, Hayling test 76%; ECAS social cognition 47%, Judgment of Preference 55%. Specificity was 100%, with the exception of PALPA spelling (93%), ECAS sentence completion (97%), and Judgment of Preference (97%).

Table 5. Inter-correlations between domains in ALS-FTD and bvFTD (Spearman's rho).

	Verbal fluency	Executive	Memory	Visuo-spatial
ALS-FTD	–	–	–	–
Language	0.51	0.35	0.13	0.25
Verbal Fluency	–	0.75**	0.19	0.48
Executive	–	–	−0.06	0.39
Memory	–	–	–	−0.11
bvFTD	–	–	–	–
Language	0.74 ***	0.77***	0.78***	0.66***
Fluency	–	0.71***	0.70***	0.40
Executive	–	–	0.62**	0.67***
Memory	–	–	–	0.43*

***$p < 0.001$; **$p < 0.01$; *$p < 0.05$.

Discussion

The findings indicate that the ECAS is highly sensitive to the cognitive impairment of people with the behavioral form of FTD, both when this occurs in the context of ALS and in isolation. Unsurprisingly, the level and frequency of impairment are substantially greater than typically reported in studies of ALS.

The impairments demonstrated in language, verbal fluency, and executive functions accord with expectation and are in keeping with findings of excellent sensitivity of the ECAS to these

ALS-specific impairments (7). It is notable, however, that impairments were demonstrated with high frequency also for ALS nonspecific cognitive domains, driven particularly by poor memory test performance. Indeed, immediate recall elicited the highest effect size in the data set after verbal fluency, substantially higher than that for social cognition. Such findings suggest that whereas the ECAS is sensitive to the type of cognitive dysfunction associated with frontotemporal spectrum disorder some diagnostic specificity may be lost when the test is applied to patients whose disorder is sufficiently severe to meet criteria for bvFTD since patients perform poorly across all or most tasks.

Early consensus criteria for frontotemporal dementia (23) identified "severe amnesia" and "visuospatial disorder" as exclusion criteria for FTD. Similarly, revised criteria (13) refer to "relative sparing of episodic memory" and "relative sparing of visuospatial skills" in bvFTD. Why then should the boundaries between ALS-specific and ALS nonspecific domains be so blurred? The clue lies in the qualifying terms used in published consensus statements: "no *severe* amnesia" and "*relative* sparing of memory". It is to be expected that the performance will not be entirely normal. bvFTD patients with severe frontal executive disorder inevitably perform poorly on open-ended memory tests by virtue of executive demands on attention and use of strategy. It is instructive that executive scores correlated specifically with immediate recall, suggesting a failure of registration of information secondary to executive factors. It is notable too that memory performance in bvFTD was strongly related to language performance. The ECAS memory task takes the form of a verbal narrative in which the discrete elements to be recalled, which include people's names and numbers, are essentially unrelated. The specific characteristics of the task might render performance particularly vulnerable to both executive and language impairments.

The findings reinforce earlier reports that test scores alone may mask different reasons for test failure: bvFTD patients make more frequent confabulatory and misconstruction responses in story recall compared to AD patients (24), suggesting a greater contribution of executive breakdown. Distinct neural substrates have also been demonstrated. One imaging study (25) found that the frontal and anterior temporal lobes underpinned episodic memory in bvFTD, but a more widespread network in AD. Another study (26) showed a correlation between memory performance and frontal lobe atrophy in bvFTD but both medial temporal and frontal lobe atrophy in AD. Memory performance in bvFTD may, moreover, be task-dependent: bvFTD patients show impaired immediate recall (24,27) but less rapid loss than AD patients over a delay. Explicit memory in bvFTD is reported to be poorer than implicit memory but may benefit from retrieval cues (28). Arguably, the open-ended ECAS story recall task may not be the optimal memory measure in the differential diagnosis of FTD.

A smaller proportion of ALS-FTD and bvFTD patients showed impairment in visuospatial compared to memory tests. Nevertheless, here too there was a correlation in bvFTD between visuospatial and both language and executive test performance, complementing previous findings that executive impairments may impact secondarily performance on visuospatial tasks (24): bvFTD patients made organizational but not spatial errors in the drawing.

Correlations between domain scores were much less apparent in ALS-FTD than bvFTD. The ALS-FTD group was smaller at the outset and further diminished by 'missing' data due to patients' inability to comply with tasks. Thus, the ALS-FTD data has reduced the power to elicit statistical effects. Nevertheless, this is unlikely to provide a sufficient explanation. The correlation between verbal fluency and executive performance in ALS-FTD (0.75) was as strong as those correlations found in bvFTD. By contrast, no association was found with language scores. The disparities raise the possibility that different factors may underpin performance breakdown in bvFTD and ALS-FTD. The disproportionately high frequency of impairment in verbal fluency in ALS-FTD compared to bvFTD is instructive given that verbal fluency deficits are the most frequently reported cognitive deficit in ALS (29). Moreover, the association of verbal fluency performance with deficits in executive, rather than primary language, skills accords with earlier findings in ALS (30).

Behavioral changes, present in all patients, together with physical limitations in ALS-FTD compound the challenges of cognitive assessment in FTD. Notwithstanding the obstacles, there were strong correspondences between performance on naming, spelling, sentence completion, and social cognition subtests of the ECAS and longer, 'standard' versions of those same tasks, providing evidence of convergent validity and complementing previous findings (7,9).

A limitation of the present study was the imprecise matching of groups. Nevertheless, comparisons of patient and control performance yielded similar frequencies of impairment as comparisons with published ECAS norms (1), slightly more conservative estimates of frequency arising when compared to age and education adjusted norms (9). Moreover, comparisons using only a sub-cohort of the control group matched for age and education yielded similar results. The relatively small sample size of the two patient groups

and, in particular, 'missing' data, due to patients' inability to engage with the task, may have reduced the power to detect potential differences between bvFTD and ALS-FTD. It is notable, for example, that ALS-FTD patients achieved a numerically lower median verbal fluency score than bvFTD patients: 3 compared to 8, yet the group difference was non-significant. By contrast, the analysis of the frequency of impairment, involving a larger number of patients because it included those unable to comply for behavioral/cognitive reasons, yielded a significant group difference. The study necessitated the use of nonparametric statistical techniques because of the skewed distribution of data. More powerful parametric methods may have been possible with a larger sample. Studies involving larger patient cohorts are warranted. The study cohort represents a prevalent sample and so caution is needed in assuming that it is fully representative of an incident population. Selection bias, arising from a volunteer cohort of patients, would, however, more likely lead to an underestimate rather than overestimate of cognitive impairment, suggesting that the ECAS findings are likely to be robust.

The findings in bvFTD and ALS-FTD add to the body of knowledge about the ECAS in neurodegenerative disease. The high frequency of impairment is similar to that reported in progressive supranuclear palsy and contrasts with the much lower frequency of impairment (30%) found in Parkinson's disease (12). It is, moreover, higher than typically found in studies of ALS. In a comparative study of ALS and AD (6) 50% of the ALS group showed impaired performance, but only 21% impairment on ALS nonspecific tasks. The latter proved most sensitive in differentiating ALS and AD. The implication is that only when executive and language breakdown becomes severe, as in FTD, does it impact secondarily on visuospatial and memory test performance.

Conclusions

The ECAS is highly sensitive to the cognitive changes in FTD and as such can be considered a valuable screening tool. Nevertheless, changes in behavior, language, and executive function, inherent in FTD, can have a secondary impact on test performance in other cognitive domains, thereby obscuring putative dissociations and complicating diagnostic differentiation. Thus, the ECAS, when performance is impaired, should be considered a prelude to more extensive neuropsychological assessment that includes a qualitative examination of performance, with a focus on the analysis of errors as well as test scores. Normal ECAS performance, in patients with ALS, may obviate the immediate need for full psychometric testing, although clinical monitoring at intervals and rechecking of behavior with relatives is recommended. Subtle differences observed in this study between ALS-FTD and bvFTD underscore the importance of a more extensive evaluation of behavior and cognition in these two patient groups.

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Declaration of interest

The authors report no conflict of interest.

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Data availability

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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