Published in final edited form as:

Amyotroph Lateral Scler. 2012 October ; 13(6): 592–598. doi:10.3109/17482968.2012.708936.

Processing and memory for emotional and neutral material in amyotrophic lateral sclerosis

Marion Cuddy¹, Benjamin J. Papps², Madhav Thambisetty³, P. Nigel Leigh⁴, and Laura H. Goldstein¹

¹King's College London, KHP Centre for Neurodegeneration Research, Department of Psychology, Institute of Psychiatry, London

²Regional Rehabilitation Unit, Northwick Park Hospital, Harrow, UK

³National Institute on Aging (NIA), Biomedical Research Center, Baltimore, MD, USA

⁴Brighton and Sussex Medical School, Trafford Centre for Biomedical Research, University of Sussex, Sussex, UK

Abstract

Several studies have reported changes in emotional memory and processing in people with ALS (pwALS). In this study, we sought to analyse differences in emotional processing and memory between pwALS and healthy controls and to investigate the relationship between emotional memory and self-reported depression. Nineteen pwALS and 19 healthy controls were assessed on measures of emotional processing, emotional memory, verbal memory and depression. Although pwALS and controls did not differ significantly on measures of emotional memory, a subgroup of patients performed poorly on an emotional recognition task. With regard to emotional processing, pwALS gave significantly stronger ratings of emotional valence to positive words than to negative words. Higher ratings of emotional words were associated with better recall in controls but not pwALS. Self-reported depression and emotional processing or memory variables were not associated in either group. In conclusion, the results from this small study suggest that a subgroup of pwALS may show weakened 'emotional enhancement', although in the current sample this may reflect general memory impairment rather than specific changes in emotional memory. Nonetheless, different patterns of processing of emotionally-salient material by pwALS may have care and management-related implications.

Keywords

Amyotrophic lateral sclerosis; emotional processing; memory; depression

Correspondence: L. H. Goldstein, Department of Psychology PO77, Institute of Psychiatry, De Crespigny Park, London SE5 8AF, UK. Fax: 0 20 7848 5006. laura.goldstein@kcl.ac.uk.

Declaration of interest: M. Cuddy, B. J. Papps and M. Thambisetty have no interests to declare.

Introduction

As many as ~15% of people with amyotrophic lateral sclerosis (pwALS) develop dementia, characterized by a range of cognitive and behavioural problems reflecting frontotemporal dysfunction (1,2). An additional ~35% of pwALS show some degree of cognitive impairment (1–3), most frequently in the area of executive functioning. Memory, language and visuospatial deficits have also been reported (4). Structural and functional brain abnormalities, particularly in prefrontal regions but also in temporal and parietal regions, have been linked to changes in cognitive functioning (e.g. (5,6)).

Papps et al. found that the normative pattern of ‘emotional enhancement’ (i.e. superior memory for emotional material relative to neutral material) was absent in a group of individuals with ALS (7). This altered pattern of responding has previously been reported in patients with amygdala damage (8), and neuropathological changes in the amygdala have been found in post mortem studies of pwALS (9). Additionally, in vivo positron emission tomography imaging has shown a trend for reduced amygdala volume in non-demented pwALS (10). Further evidence for an absence of ‘emotional enhancement’, and for reduced activation more generally across the right hemisphere was reported in an fMRI study, which suggested the involvement of a wider network of brain structures in ALS rather than the amygdala alone (11).

The processing of emotional material by pwALS has been investigated in several studies. Difficulty in recognizing facial expressions of emotion (12,13) and a tendency towards impaired interpretation of mental state from eye expressions (13) were identified. Another study indicated that when presented with emotional pictures, pwALS gave more positive ratings of emotional valence and more neutral ratings of arousal than controls (14).

Changes in emotional processing might be related to affective changes in pwALS. Moore et al. (15) found clinically significant levels of depression in only 23% of a sample of pwALS. However, a recent study reported a prevalence of 26–56%, depending on the depression measure used and level of depression considered (16); another study concluded that the prevalence of depression in pwALS may not differ from that in patients with other neuromuscular disorders (17). The factors increasing the likelihood of pwALS becoming depressed are unclear, although certain studies report a link between depression and symptom severity (18).

The present study aimed to extend previous findings relating to emotional memory (7), by assessing both recall and recognition in order to establish whether the deficit occurred at the encoding or retrieval stage. We predicted that pwALS would not show the normative pattern of ‘emotional enhancement’ on assessments of emotional recall and recognition. In view of the mixed findings of memory impairment in pwALS (see (2,4)), we attempted to differentiate specific changes in emotional memory from more general memory impairment by including a standardized measure of verbal memory. We also predicted that the affective ratings of emotional words by pwALS would differ from those of healthy controls. Finally, we wished to explore the relationship between self-reported depression, emotional memory

and emotional processing to provide preliminary data concerning inter-relationships between these processes.

Methods

Participants

Ethics approval was obtained from the Joint South London and Maudsley & Institute of Psychiatry NHS Research Ethics Committee and King's College Hospital Research Ethics Committee; all participants provided written informed consent. The sample consisted of 19 patients with ALS and 19 healthy controls. A power calculation, based on Papps et al.'s study, indicated that this sample size would have 87% power to detect a 2.5-point difference (when $\mu = 12$, $\sigma = 2.5$) at $p = 0.05$ in two-tailed tests. Tests and questionnaires were administered in the same order for all participants. Data were analysed using SPSS V15.0 for Windows.

Control participants were recruited by poster advertisements at local community centres and churches, and by approaching partners and friends of the patients who took part in the study. Patients and controls were included if aged 35–75 years and English was their first language. Exclusion criteria for both groups were a history of another neurological disease, head injury, cerebrovascular disease, hypertension, diabetes, currently being prescribed psychoactive medication or reporting excessive daytime sleepiness (19). Patients were excluded if diagnosed more than two years previously, had a diagnosis of 'ALS-dementia', any atypical form of ALS, if they were anarthric, were too ill to take part or if they had evidence of respiratory muscle weakness (e.g. FVC < 70%).

Potential participants with ALS were patients attending the King's MND Care and Research Centre, London. Diagnosis of probable or definite ALS according to El Escorial criteria (20) was the main inclusion criterion ($n = 114$). Twenty patients were excluded due to age, and 10 had been diagnosed more than two years previously. Twenty-eight patients were not approached due to severity of illness or dysarthria. Five individuals were excluded due to respiratory problems/daytime sleepiness and a further 20 due to additional physical, neurological or psychiatric diagnoses. Six did not have English as their first language. Finally, six individuals declined to participate.

Measures

General intellectual ability and memory—We assessed background differences between groups in terms of premorbid and current general intellectual ability, as well as verbal memory, using the following measures:

- Wechsler Test of Adult Reading (WTAR) (21). This measure of premorbid intellectual ability requires the person to read aloud 50 words, from which a Full Scale IQ score can be predicted.
- Two-subtest Wechsler Abbreviated Scale of Intelligence (WASI) (22). The Vocabulary and Matrix Reasoning subtests provided an estimate of current intellectual functioning without being unduly affected by motor difficulties experienced by pwALS.

- California Verbal Learning Test–Second UK Edition (CVLT-II) (23). This list-learning test assesses verbal learning and memory. It yields information about immediate and delayed free recall, cued recall and recognition, as well as recall and recognition errors and learning strategies.

Executive functioning was not formally assessed, as it was deemed important to keep the neuropsychological battery brief to minimize patient fatigue.

Emotional memory

- Brierley-Medford Sentences (8). Previously used in ALS research (7), this test measures recognition for emotionally salient and neutral material. Participants read aloud 42 sentences in which emotional tone is manipulated by the inclusion of an emotional or neutral target word (e.g. He would abuse/amuse the children at every party). Recognition memory for the target words is tested after a 30-min delay. We employed a parallel version of the test used previously (7) to avoid interference between words used in the previously-employed version and the Phelps Words (24).
- Phelps Words (24). This test measures emotional processing and emotional recall memory. Participants are presented with 27 words (nine positive, e.g. ‘smile’, nine negative, e.g. ‘cancer’, and nine neutral, e.g. ‘chair’), which they rate for emotional valence on a 5-point scale, ranging from ‘very negative’ to ‘very positive’. An unanticipated free recall test is given after a filled delay of 5 min. Healthy adults have demonstrated superior recall for positive or negative words over neutral words on this task.

Other measures—Socio-economic status was determined using the Standard Occupational Classification System; here Social Classes I–II denote primarily professional and technical occupations, whilst Social Classes III–V refer to clerical and manual skilled and unskilled occupations (25). The depression subscale of the Hospital Anxiety and Depression Scale (HADS-D) (26) measured self-reported depression. As in other studies of patients with ALS (e.g. (5)) and validated through Rasch analysis (27), the item concerning ‘feeling slowed down’ was removed, yielding a maximum possible score of 18. The ALS Functional Rating Scale-Revised (ALSFRS-R) (28), Emotional Lability Questionnaire (ELQ) (29) and Epworth Sleepiness Scale (ESS) (19) were administered to measure severity of additional ALS symptoms in the patient group and to screen for potentially confounding variables.

Results

Sample characteristics

The pwALS had been diagnosed on average 13 months previously (range 1–24 months) and had relatively mild physical impairments (mean ALS-FRS-R score, 37.7 (SD = 6.4)). The two groups did not differ in age ($t = 0.8$, $df(36)$; $p = 0.46$), gender ($\chi^2 = 1.0$, $df(1)$; $p = 0.32$) or social class distribution ($\chi^2 = 0.6$, $df(1)$; $p = 0.43$) (Table I). No significant differences were found on the ESS ($t = 0.6$, $df(36)$, $p = 0.52$) or the ELQ ($Z = -0.5$, $p = 0.62$).

Neuropsychological test and questionnaire scores are presented in Table II. The groups were matched in terms of estimated current intellectual functioning. Control participants showed a non-significant tendency to obtain higher scores than pwALS on the WTAR, possibly as a result of the mild dysarthria experienced by some pwALS. As the distribution of current IQ was similar in the two groups, and results did not suggest current decline in IQ, WTAR scores were not entered as a covariate in subsequent analyses.

The pwALS obtained higher HADS-D scores than controls; this difference was not significant and both groups' mean scores were in the normal range. One participant from each group scored above the suggested cut-off for depression. In subsequent analyses where HADS-D was related to performance, this variable was entered as a covariate. CVLT scores for pwALS and control participants were compared using ANOVAs, with HADS-D score as a covariate. No significant between-groups differences were found in verbal memory performance (Table II).

Emotional memory

Mean scores on the two measures of emotional memory are presented in Table III. Inspection of the raw data suggested a differential pattern of responding for the two groups, particularly on the Brierley-Medford Sentence task, with an apparently greater degree of 'emotional enhancement' in control participants. The data were analysed using repeated measures two-way (Emotional Valence $\times$ Group) ANOVA. HADS-D scores were entered as a covariate in the Phelps Recall Task analysis.

On the Brierley-Medford Sentence Task, there was an effect of Emotional Valence ($F = 55.3$, $df = 1, 36$; $p < 0.001$); more emotional words were recognized than neutral words. However, there was no effect of Group ($F = 2.8$, $df = 1, 36$; $p = 0.10$), and no Emotional Valence $\times$ Group interaction ($F = 1.3$, $df = 1, 36$; $p = 0.27$).

Scores on the Phelps Recall Task were low across both groups. Again, there was an effect of Emotional Valence ($F = 5.14$, $df(1, 35)$; $p < 0.01$), but no effect of Group ($F = 1.41$, $df(1, 35)$; $p = 0.24$) and no Emotional Valence $\times$ Group interaction ($F = 0.38$, $df(1, 35)$; $p = 0.69$). Post-hoc paired-sample t -tests revealed superior recall of positive words compared to neutral words in the sample as a whole ($t = 2.8$, $df = 37$; $p < 0.01$).

Analysis of percentile scores

The number of patients obtaining 'impaired' scores (i.e. scores at or below the fifth percentile of controls' scores) on measures of emotional memory was examined, to determine whether a subgroup of pwALS might be showing a different pattern of responding. Subgroup analyses of this type have been carried out in other studies of cognitive functioning in ALS, in response to growing evidence that the cognitive profile of pwALS is heterogeneous (2,3).

The distribution of scores obtained by pwALS and controls on the Brierley-Medford Sentence Task is shown in Figure 1. Eight patients (42%) scored $\leq$ 5th percentile score of control participants on the measure of emotional word recognition; this difference in proportions was significant (Fisher's exact test, $p < 0.001$). There were no differences in the

proportion of pwALS and controls obtaining ‘impaired’ scores on the measure of neutral word recognition ($p = 1.0$).

Patients with ‘impaired’ recognition of emotional words also performed worse than the other patients the short-delay free recall ($t = -2.6$, $df(17)$; $p = 0.02$; 95% CI -5.84 to -0.59), short-delay cued recall ($t = -2.8$, $df(17)$; $p = 0.01$; 95% CI -5.70 to -0.75), and long-delay free recall trials ($t = -3.6$, $df(17)$; $p < 0.01$; 95% CI -7.03 to -1.84) of the CVLT. The two patient subgroups did not differ in their estimated current or premorbid IQ scores, or on any of the questionnaire measures.

Six patients (32%) scored 5th percentile of controls’ scores on the Phelps positive word recall task; this difference in proportions was not significant (Fisher’s exact test, $p = 0.09$). As these participants’ scores were at floor level it was not deemed appropriate to carry out further analyses. There were no significant differences in the proportion of pwALS and controls obtaining ‘impaired’ scores on the neutral or negative word recall tasks.

Four patients obtained ‘impaired’ scores on both emotional memory tasks. A subgroup of this size was too small to perform further statistical analyses.

Emotional processing

Mean affective ratings of Phelps words are presented in Table IV. High scores indicate higher ratings of emotional valence, either positive or negative. The pwALS and control participants did not differ in their affective ratings of positive, negative or neutral words. Wilcoxon signed-ranks tests, conducted separately for the two groups, indicated that pwALS gave higher affective ratings to positive compared to negative words ($Z = -3.3$; $p < 0.01$). This difference was not present in control participants.

Preliminary exploration of the relationship between depression and emotional memory and processing

We explored the association between HADS-D depression score and emotional memory profile. ‘Emotional memory profile’ was defined as the difference between the number of emotional and neutral words recalled/recognized on an emotional memory task, divided by the total number of words recalled/recognized. Correlations were performed separately for each group. No association was found between HADS-D scores and emotional memory profile or emotional processing in either group.

Correlations between affective ratings and recall of Phelps words in the same emotional category indicated that higher ratings of negative words were associated with better recall in control participants ($r_s = 0.48$; $p = 0.04$) but not in the ALS group ($r_s < 0.1$; $p = 0.73$).

Discussion

The present study sought to consider the processing of and memory for emotional material in pwALS. In contrast to our previous findings (7), no differences were found in the overall emotional memory performance of the two groups; both groups demonstrated the expected ‘emotional enhancement’ effect. However, examination of percentile scores suggested that a

subgroup of pwALS might demonstrate weakened memory for emotional material, although the difference in proportions of the ALS and control groups showing reduced memory for emotional words was only statistically significant for the Brierley-Medford Sentence task. Findings therefore support the view that the cognitive profile of pwALS is heterogeneous (3), but add only tentative support to previous findings regarding changes in emotional memory in this patient group (7,11). Our failure to replicate a between-group difference in terms of emotional enhancement on the Brierley-Medford Sentence task may reflect heterogeneity in the ALS population (which may become more apparent when comparing small samples as here with those in our previous study) (7), the greater similarity between groups in HADS-D scores in our previous study (7) than currently, or potentially the difference in versions of the Brierley-Medford Sentences test used. Between-group differences on the Phelps recall task may have been masked by the low scores obtained by participants in both groups, which meant that it was not feasible to compare encoding and retrieval processes in emotional memory performance. It is possible that methodological factors may have contributed to the poor recall performance in this study, as the original study used a computerized version of the task, which may have had an impact on the extent to which words were encoded.

Furthermore, although as a group pwALS performed at a similar level to healthy controls on standardized measures of verbal memory, patients who showed impaired emotional memory performed more poorly than non-impaired patients on a number of verbal memory measures. Thus, the results may be indicative of general memory impairment in a subgroup of patients, rather than a specific alteration in memory for emotional material. Other studies have reported memory impairment in pwALS, although findings in this area are inconsistent (4). This could be interpreted as further evidence that memory is affected in some but not all individuals with ALS, or it could be that the likely involvement of executive functions in strategic encoding and semantic clustering, of relevance to performance on the CVLT, distinguished those who showed altered memory for emotional material. Nevertheless, it is noteworthy that only a subgroup of pwALS showed relatively poor CVLT performance, despite the test's reliance on executive processes. Despite having collected measures of IQ, memory and mood, more extensive neuropsychological assessment would have been informative, especially in relation to executive functions (30), as these might have influenced learning and emotional functioning.

As a group, pwALS gave stronger affective ratings to positive words than to negative words, suggesting a somewhat blunted affective response to negative stimuli. These findings are consistent with the changes in emotional processing suggested elsewhere (14) and may contribute to considerations of why the prevalence of depression in ALS is lower than expected, at least in some studies (15); a subdued emotional response to negative information may potentially reduce its impact on mood.

In the present study, no associations were found between depression scores and emotional memory or processing in pwALS. However, scores were mostly quite low and the restricted range of scores may have influenced analyses. It may be informative to compare emotional processing in clinically-depressed and non-depressed pwALS, as well as in patients with other chronic or degenerative conditions, to clarify whether these changes are neurological

in nature or related to the patients' adjustment to their illness. The use of different means to assess depression (16,17) may yield different patterns of association with measures of emotional processing.

Stronger emotional ratings of words were associated with better recall in control participants, but not in patients. The mechanism behind this difference is unclear. Lulé et al. (14) reported that compared to control participants, pwALS rated emotional pictures as significantly less arousing. Emotional arousal is thought to affect certain hormonal and brain systems involved in the storage of information in memory and it contributes to 'emotional enhancement' (31). The muted response of pwALS to emotional stimuli suggests reduced emotional arousal and might lead to a weaker memory trace for those stimuli. This could account for the absence of 'emotional enhancement' in some patients, as well as the lack of association between affective ratings and memory for emotional words. This suggests that changes in the processing of emotional information may contribute to the reduced 'emotional enhancement' found in some pwALS.

The cerebral mechanism mediating changes in emotional memory and processing in pwALS is unknown. However, it is possible that changes in the amygdala contribute to changes in emotional responding. Selective damage to the amygdala has been linked to impaired emotional memory (32). Furthermore, dissociation between emotional processing and emotional memory has been documented following amygdala damage (33). Thus, it is possible that the dissociation between affective ratings and recall of emotional words in the current pwALS may be related to changes in the amygdala reported in pwALS (9,10).

Summary and conclusions

Our findings support the heterogeneity of cognitive involvement in ALS. Although as a group, pwALS did not demonstrate an altered emotional memory profile, analyses suggested that a subgroup of pwALS showed reduced memory for emotional material compared to control participants, though these findings may reflect general memory impairment rather than changes in emotional memory. The ALS group also showed some changes on measures of emotional processing. The results raise the possibility that emotional memory and processing are mediated differently in pwALS compared to controls. Further research needs to determine and understand changes in cognitive and affective functioning in this population and their clinical management implications.

Acknowledgments

P. N. Leigh has received honoraria or consultancy fees and travel expenses from Sanofi Aventis, GlaxoSmithKline (GSK), Acceleron, Cytokinetics, Neuronova, and is/or has been a member of advisory boards for GSK, Acceleron, Cytokinetics, Neuronova, TauPx. He has received research support from the Motor Neurone Disease Association and the Wellcome Trust UK. L. H. Goldstein receives salary support from the National Institute for Health Research (NIHR), Dementia Biomedical Research Unit at South London and Maudsley NHS Foundation Trust and King's College London. The views expressed are those of the author and not necessarily those of the NHS, the NIHR or the Department of Health. She serves on the Scientific Awards Panel for Epilepsy Action (UK); has received travel expenses and honoraria for speaking and educational activities not funded by industry; receives royalties from the publication of *Clinical Neuropsychology* (Wiley, 2004) and *The Clinical Psychologist's Handbook of Epilepsy* (Routledge, 1997); and receives research support from Department of Health/National Institute for Health Research (NIHR) UK and the Motor Neurone Disease Association UK. She has also received research support from Epilepsy Research UK, the Institute of Social Psychiatry and the Wellcome Trust UK.

The authors alone are responsible for the content and writing of the paper.

References

1. Phukan J, Elamin M, Bede P, Jordan N, Gallagher L, Byrne S, et al. The syndrome of cognitive impairment in amyotrophic lateral sclerosis: a population based study. *J Neurol Neurosurg Psychiatry*. 2012; 83:102–8. [PubMed: 21836033]
2. Phukan J, Pender NP, Hardiman O. Cognitive impairment in amyotrophic lateral sclerosis. *Lancet Neurology*. 2007; 6:994–1003. [PubMed: 17945153]
3. Ringholz GM, Appel SH, Bradshaw M, Cooke N, Mosnik DM, Schulz PE. Prevalence and patterns of cognitive impairment in sporadic ALS. *Neurology*. 2005; 65:586–90. [PubMed: 16116120]
4. Goldstein, LH. Control of symptoms: cognitive dysfunction. In: Oliver, D.Borasio, GD., Walsh, D., editors. *Palliative Care in Amyotrophic Lateral Sclerosis*. Oxford: Oxford University Press; 2006. p. 111-27.
5. Abrahams S, Goldstein LH, Kew JJ, Brooks DJ, Frith C, Leigh PN. Frontal lobe dysfunction in amyotrophic lateral sclerosis. A PET study. *Brain*. 1996; 119:2105–20. [PubMed: 9010014]
6. Tsermentseli S, Leigh PN, Goldstein LH. The anatomy of cognitive impairment in amyotrophic lateral sclerosis: more than frontal lobe dysfunction. *Cortex*. 2012; 48:166–82. [PubMed: 21396632]
7. Papps B, Abrahams S, Wicks P, Goldstein LH, Leigh PN. Changes in memory for emotional material in amyotrophic lateral sclerosis (ALS). *Neuropsychologia*. 2005; 43:1107–14. [PubMed: 15817168]
8. Brierley B, Medford N, Shaw P, David AS. Emotional memory and perception in temporal lobectomy patients with amygdala damage. *J Neurol Neurosurg Psychiatry*. 2004; 75:593–9. [PubMed: 15026504]
9. Anderson VE, Cairns NJ, Leigh PN. Involvement of the amygdala, dentate and hippocampus in motor neuron disease. *J Neurol Sci*. 1995; 129(Suppl 8):75–8. [PubMed: 7595627]
10. Pinkhardt EH, van Elst LT, Ludolph AC, Kassubek J. Amygdala size in amyotrophic lateral sclerosis without dementia: an in vivo study using MRI volumetry. *BMC Neurology*. 2006; 48:6.
11. Palmieri A, Naccarato M, Abrahams S, Bonato M, D'Ascenzo C, Balestreri S, et al. Right hemisphere dysfunction and emotional processing in ALS: an fMRI study. *J Neurol*. 2010; 257:1970–8. [PubMed: 20593194]
12. Zimmerman EK, Eslinger PJ, Simmons Z, Barrett AM. Emotional perception deficits in amyotrophic lateral sclerosis. *Cogn Behav Neurol*. 2007; 20:79–82. [PubMed: 17558250]
13. Girardi A, MacPherson SE, Abrahams S. Deficits in emotional and social cognition in amyotrophic lateral sclerosis. *Neuropsychology*. 2011; 25:53–65. [PubMed: 20919762]
14. Lulé D, Kurt A, Jurgens R, Kassubek J, Diekmann V, Kraft E, et al. Emotional responding in amyotrophic lateral sclerosis. *J Neurol*. 2005; 252:1517–24. [PubMed: 15977000]
15. Moore MJ, Moore PB, Shaw PJ. Mood disturbances in motor neuron disease. *J Neurol Sci*. 1998; 160:S53–6. [PubMed: 9851650]
16. Wicks P, Abrahams S, Masi D, Hejda-Forde S, Leigh PN, Goldstein LH. The prevalence of depression in amyotrophic lateral sclerosis. *Eur J Neurol*. 2007; 14:993–1001. [PubMed: 17718691]
17. Taylor L, Wicks P, Leigh PN, Goldstein LH. Prevalence of depression in amyotrophic lateral sclerosis and other motor disorders. *Eur J Neurol*. 2010; 17:1047–53. [PubMed: 20158515]
18. Goldstein LH, Adamson M, Jeffrey L, Down K, Barby T, Wilson C, et al. The psychological impact of MND on patients and carers. *J Neurol Sci*. 1998; 160(Suppl 1):S114–21. [PubMed: 9851660]
19. Johns M. A new method for measuring daytime sleepiness: The Epworth Sleepiness Scale. *Sleep*. 1991; 14:540–5. [PubMed: 1798888]
20. Brooks BR, Miller RG, Swash M, Munsat TL. World Federation of Neurology Research Group on Motor Neuron Diseases. El Escorial revisited: revised criteria for the diagnosis of amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Other Motor Neuron Disord*. 2000; 1:293–9. [PubMed: 11464847]

21. Holdnack, J. Wechsler Test of Adult Reading. San Antonio, Texas: Psychological Corporation; 2001.
22. Wechsler, D. Wechsler Abbreviated Scale of Intelligence. San Antonio, Texas: Psychological Corporation; 1999.
23. Delis, DC., Kaplan, E., Kramer, JH., Ober, BA. California Verbal Learning Test, 2nd UK edition. London: Harcourt; 2000.
24. Phelps EA, LaBar KS, Spencer D. Memory for emotional words following unilateral temporal lobectomy. *Brain Cogn.* 1997; 35:85–109. [PubMed: 9339304]
25. Office for National Statistics. Standard Occupational Classification. 2. London: Office for National Statistics; 2000.
26. Zigmond AS, Snaith RP. The Hospital Anxiety and Depression Scale. *Act Psychiatr Scand.* 1983; 67:361–70.
27. Gibbons CJ, Mills RJ, Thornton EW, Ealing J, Mitchell JD, Shaw PJ, et al. Rasch analysis of the Hospital Anxiety and Depression Scale (HADS) for use in motor neuron disease. *Health and Quality of Life Outcomes.* 2011; 9:82. [PubMed: 21955749]
28. Cedarbaum JM, Stambler N, Malta E, Fuller C, Hilt D, Thurmond B, et al. The ALSFRS-R: a revised ALS functional rating scale that incorporates assessments of respiratory function. BDNF ALS Study Group (Phase III). *J Neurol Sci.* 1999; 169:13–21. [PubMed: 10540002]
29. Newsom-Davis IC, Abrahams S, Goldstein LH, Leigh PN. The emotional lability questionnaire: a new measure of emotional lability in amyotrophic lateral sclerosis. *J Neurol Sci.* 1999; 169:22–5. [PubMed: 10540003]
30. Strong MJ, Grace GM, Freedman M, Lomen-Hoerth C, Woolley S, Goldstein LH, et al. Consensus criteria for the diagnosis of frontotemporal cognitive and behavioural syndromes in amyotrophic lateral sclerosis. *Amyotroph Lateral Scler.* 2009; 10:131–46. [PubMed: 19462523]
31. Cahill L, McGaugh JL. Mechanisms of emotional arousal and lasting declarative memory. *Trends Neurosci.* 1998; 21:294–9. [PubMed: 9683321]
32. Cahill L, Babinsky R, Markowitsch HJ, McGaugh JL. The amygdala and emotional memory [2]. *Nature.* 1995; 377:295–6. [PubMed: 7566084]
33. Papps BP, Calder AJ, Young AW, O'Carroll R. Dissociation of affective modulation of recollective and perceptual experience following amygdala damage. *J Neurol Neurosurg Psychiatry.* 2003; 74:253–4. [PubMed: 12531962]

Figure 1.
Distribution of scores on the Brierley-Medford Sentence Task.

Table I

Sample characteristics.

	ALS Group Mean (SD) or <i>n</i>	Control Group Mean (SD) or <i>n</i>
Age (years)	56.1 (8.3)	53.8 (10.3)
Male:Female	13:6	10:9
ESS	6.1 (3.7)	5.4 (3.3)
ELQ (median (range))	0 (0 – 29)	0 (0 – 10)
Social Class I–II	15	16
Social Class III–IV	4	3

ESS: Epworth Sleepiness Scale; ELQ: Emotional Lability Questionnaire.

Table II

Neuropsychological and depression scores.

Measure	ALS Group Mean (SD)	Control Group Mean (SD)	<i>t</i>	Df	<i>p</i>	95% CI
Current IQ (WASI)	115.9 (12.1)	116.7 (12.1)	-0.2	36	0.83	-8.82, 7.13
Prenorbid IQ (WTAR)	103.8 (2.5)	108.7 (2.9)	-2.0	36	0.06	-10.10, 0.21
CVLT Measures			F	Df	<i>p</i>	95% CI
Immediate Free Recall	45.6 (8.7)	52.6 (11.4)	2.8	1,35	0.10	-13.7, -0.4
Short Delay Free Recall	9.7 (3.1)	11.3 (3.8)	0.97	1,35	0.33	-3.9, 0.7
Short Delay Cued Recall	10.4 (2.9)	11.8 (2.8)	1.5	1,35	0.22	-3.3, 0.5
Long Delay Free Recall	9.3 (3.4)	11.3 (3.5)	2.1	1,35	0.16	-4.2, 0.3
Long Delay Cued Recall	9.9 (3.0)	11.7 (3.5)	2.3	1,35	0.14	-3.9, 0.4
Recognition	14.4 (1.4)	15.1 (1.7)	1.8	1,35	0.19	-1.8, 0.3
Questionnaires			<i>t</i>	Df	<i>p</i>	95% CI
HADS depression	3.4 (2.9)	1.6 (2.7)	1.9	36	0.06	-0.11, 3.58

CI: Confidence interval; WASI: Wechsler Abbreviated Scale of Intelligence; WTAR: Wechsler Test of Adult Reading; CVLT: California Verbal Learning Test; HADS: Hospital Anxiety and Depression Scale.

Table III

Emotional memory performance.

Measure	ALS Group Mean (SD)	Control Group Mean (SD)
Brierley-Medford Sentences		
Emotional words (maximum possible score: 21)	15.8 (2.9)	17.5 (1.6)
Neutral words (maximum possible score: 21)	13.2 (2.5)	13.9 (3.2)
Phelps Words Task		
Positive Words (maximum possible score: 9)	1.5 (1.4)	2.1 (1.3)
Negative words (maximum possible score: 9)	1.3 (1.0)	1.6 (1.2)
Neutral words (maximum possible score: 9)	1.0 (0.8)	1.3 (0.9)

Table IV

Affective ratings of Phelps words.

Measure	ALS Group Mean (SD)	Control Group Mean (SD)	Z	p
Positive words	14.5 (3.2)	14.6 (2.5)	-0.3	0.82
Neutral words	1.6 (1.7)	1.4 (1.0)	-0.1	0.91
Negative words	12.7 (2.9)	13.9 (2.4)	-0.3	0.70