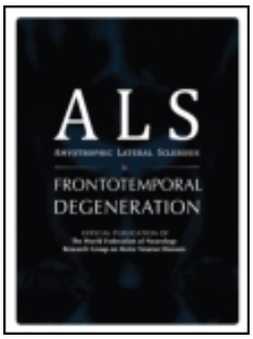

Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration

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THEME 3 COGNITIVE AND PSYCHOLOGICAL ASSESSMENT AND SUPPORT

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THEME 3 COGNITIVE AND PSYCHOLOGICAL ASSESSMENT AND SUPPORT

P47 COGNITIVE AND BEHAVIOURAL SYMPTOMS ALONG THE ALS SPECTRUM: DETECTION, DIFFERENTIATION AND PROGRESSION

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Keywords: cognitive screening, frontotemporal dementia (FTD), behavioural symptomatology

Background: A brief clinical screening tool which can detect cognitive and behavioural symptoms in ALS is pertinent, particularly for patients with an adjunct frontotemporal dementia diagnosis (ALS FTD). The differentiation of ALS FTD from patients with subtle but definite cognitive or behavioural symptoms (ALS plus) and motor symptoms only (ALS pure) is also relevant in light of recent diagnostic criteria. Finally, the emergence of cognitive and behavioural symptoms with disease progression in ALS has important implications for disease management.

Objectives: To investigate the effectiveness of two validated screening tools in detecting, differentiating and elucidating the progression of cognitive and behavioural symptoms along the ALS spectrum: The Mini-Addenbrooke's Cognitive Examination (M-ACE), a brief cognitive screening tool, and the Motor Neuron Disease Behavioural Scale (MiND-B), a behavioural questionnaire.

Methods: ALS patients (n = 54) and controls (n = 45) were recruited from the Motor Neuron Disease Multidisciplinary Clinic at the Prince of Wales Hospital (MiND) and the Frontier Frontotemporal Dementia Research Group, Sydney, Australia. Comprehensive examination enabled classification of the participants into those with an adjunct diagnosis of FTD (ALS FTD; n = 25) and those without (n = 29) were further subdivided into ALS pure (n = 17) and ALS plus (n = 12) according to the Strong criteria (1).

Results: The M-ACE and MiND-B combined correctly classified 88% of ALS FTD patients. Moreover, almost all ALS FTD (> 90%) patients scored below the cut-off of 25/30 on the M-ACE. Neither one of the two screening tools alone differentiated between the three ALS cohorts: the MiND-B differentiated between ALS pure and ALS plus only whereas the M-ACE differentiated between the ALS FTD and those without the adjunct FTD diagnosis. Finally, Rasch modelling of M-ACE and MiND-B items revealed that cognitive and behavioural symptoms emerged synchronously with fluency, memory and apathy being more prominent early in ALS.

Discussion and conclusion: The M-ACE was useful for the detection of ALS FTD patients but did not differentiate between ALS pure and ALS plus groups. This was perhaps not surprising as ALS plus is currently defined according to behavioural symptomatology and executive dysfunction, both of which are not measured by the M-ACE. Interestingly, the severity of behavioural symptomatology did not differ between ALS plus and ALS FTD groups. This likely reflects the heterogeneity of the ALS FTD sample whereby some individuals had prominent behavioural symptoms whereas language impairment predominates in others.

The M-ACE and MiND-B combined detects ALS FTD patients, differentiates along the ALS continuum and offers insight into the progression of non-motor symptomatology in ALS.

Acknowledgements: Motor Neurone Disease Research Institute of Australia, Forefront Research Program, National Health and Medical Research Council of Australia and the ARC Center of Excellence in Cognition and its Disorders.

Reference:

1. Strong MJ. *et al.* ALS 2009; 10: 131–146.

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P48 THE COGNITIVE PROFILES OF ALS AND FTD SHOW CONSIDERABLE OVERLAP

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Keywords: cognitive profile, frontotemporal dementia (FTD), meta-analysis

Background: Cognitive impairment is present in 30% of ALS patients and consists of deficits in executive functions, language, social cognition and memory. Based on the overlap of ALS and frontotemporal dementia (FTD), we aimed to test the hypothesis that the cognitive profiles are similar.

Objectives: To compare the results of a meta-analysis of the cognitive profile of FTD with an update of our previous meta-analysis of the cognitive profile of ALS.

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Methods: Embase, PsycInfo and Medline were searched for neuropsychological studies of behavioural variant FTD patients (bv-FTD) and age and education matched healthy volunteers. At least one validated neuropsychological test had to be used and means and standard deviations had to be presented. All tests were categorized in cognitive domains and effect sizes (Hedges' g) were calculated. The comparison of the cognitive profiles of FTD and ALS was performed using two different approaches. First, the overlap of the confidence intervals of the effect sizes of FTD and ALS was investigated. Second, the effect sizes and the confidence intervals were standardized and expressed as standard deviations from the mean effect size of FTD and ALS ((effect size single domain - mean effect size FTD or ALS) / SD of mean effect size FTD or ALS).

Results: We screened 7384 abstracts and evaluated 449 full text papers. 93 articles were included in the FTD meta-analysis (1855 FTD patients and 2005 controls). All cognitive domains (n = 12) showed significant effect sizes. The largest effect sizes in FTD were seen for social cognition, MMSE and fluency (1.61, 1.51 and 1.44, respectively). There was a considerable difference in effect sizes between FTD and ALS patients, ie, the largest effect size of ALS patients was 0.63, and there was no overlap of the confidence intervals of any of the cognitive domains. The comparison of the standardized effect sizes showed a considerable overlap for social cognition, fluency, executive functions, delayed and immediate verbal memory, visuoperception and the MMSE.

Discussion and conclusion: The cognitive profiles of bv-FTD (deficits in social cognition, fluency, executive functions and memory) and ALS show great overlap. This finding further supports the existence of a disease continuum, with ALS and FTD on both extremes. Deficits of social cognition and memory occur frequently in both diseases, which indicate that cognitive impairment extends beyond executive dysfunction.

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P49 QUANTIFYING COGNITIVE AND BEHAVIORAL CHANGES IN ALS OVER THE COURSE OF THE DISEASE

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Keywords: frontotemporal dementia (FTD), cognitive, behavioural

Background: Up to 40% of patients with ALS develop cognitive symptoms consistent with frontotemporal dementia (FTD) or dysfunction. The ALS-CBS is a validated screening tool that assesses cognitive and behavioural performance. Prior studies have not examined scores over time to assess stability.

Objective: To explore the relationship of performance on the Amyotrophic Lateral Sclerosis Cognitive Behavioural Screen (ALS-CBS) over time.

Methods: There are two portions to the ALS-CBS: for the cognitive portion (completed by the ALS patient) a cut off score of $>16/20$ is considered cognitively normal, scores $<11/20$ are correlated with FTD, and scores between the two are suggestive of cognitive impairment; for the behavioural ratings (completed by the ALS caregiver) a cut off score of $>35/45$ is considered behaviourally normal, scores $<33/45$ are correlated with FTD, and scores between the two cut offs are highly suggestive of behavioural impairment.

71 patients were screened using the ALS-CBS (45 male, 63.4%; 26 female, 36.6%; average age 63.8 years). 19 of the 71 patients were screened twice, 6 + months apart (13 male, 68.4%; 6 female, 31.6%; average age 62.0 years).

Results: For the cognitive portion: 25.6% had scores in the normal range; 65.6% had scores suggestive of cognitive impairment; and 8.8% had scores indicating the likely presence of FTD. Among the 19 patients who completed the ALS-CBS twice, at the initial screen: 3/19 were in the normal range; 14/19 were in the cognitive impairment range; and 2/19 in the FTD range. At follow up: 7/19 were in the normal range; 10/19 in the cognitive impairment range; and 2/19 in the FTD range. Change in cognitive scores between timepoints ranged from -3 to $+4$ at follow up.

For behavioural ratings: 56% were in the normal range; 17.9% were in the range suggestive of behavioural impairment; and 26.2% were in the range associated with FTD behaviours. Among the 18 patients who had caregiver ratings twice: initial scores were 9/18 in normal range; 3/18 in the behavioural impairment range; and 6/18 in the FTD behavioural range. At follow up: 8/18 were in normal range; 5/18 in the behavioural impairment range; and 5/18 in the FTD behavioural range. Change in behavioural scores at follow up ranged from -27 to $+7$. There were no correlative factors between changes in cognitive scores and behavioural ratings between the two timepoints.

Discussion and conclusion: The ALS-CBS in our patient group suggests that ~75% have cognitive and ~44% have behavioural difficulties. There were discrepancies between patient performance and caregiver ratings. The levels of impairment are relatively stable over 6 months, however changes in both cognitive and behavioural scores in both directions, towards improvement or towards further disability, were noted. Overall, the ALS-CBS appears to be useful in measuring cognitive and behavioural changes over time.

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P50 A PILOT STUDY TO ESTABLISH RELIABLE TELEPHONE-BASED COGNITIVE TESTING FOR THE ALS PATIENT POPULATION

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Keywords: frontotemporal dementia (FTD), cognitive testing, reliability

Background: A national, epidemiologic study performed entirely over the phone that examines the relationship between oxidative stress and sporadic amyotrophic lateral sclerosis (ALS) is currently underway. Since frontotemporal dementia (FTD) is a well-recognized co-morbid disorder with ALS, the necessity to evaluate FTD over the phone is integral.

Objectives: To establish a reliable battery of measures to assess cognitive functioning over the phone.

Methods: In order to assess the validity of an ALS-appropriate, telephone-based screening exam, subjects were administered both in-person and telephone-based testing and randomly assigned to reverse-order groups. The battery included a telephone-modified version of the UCSF Cognitive Test Battery for FTD: (1) The ALS Cognitive Behavioural Screen (ALS-CBS); 2) Written Verbal Fluency and Controlled Oral Word Association Test (COWAT); 3) the Frontal Behavioural Inventory (FBI-ALS); 4) Center for Neurologic Study-Lability Scale (CNS-LS), and 5) the Mini-Mental State Examination (MMSE) and Telephone Interview for Cognitive Status (TICS). ALS assessment consisted of the ALS Functional Rating Scale-Revised (ALS-FRS-R) and % Forced Vital Capacity (% FVC). Patients were assigned to two groups and first received in-person or telephone evaluations followed by the opposite mode of testing approximately two weeks later. Two interviewers, trained to administer the battery, performed the testing, with one interviewer carrying out both modes of testing for each patient.

Results: To date, 20 subjects have completed both over-the-phone and in-person testing. The mean age was 62 (+ 7.4), 40% of the samples were women, the mean ALSFRS-R score was 37.3 (+ 6.2) and the mean %FVC was 83.5 (+ 18.3). Reliability between telephone-based and in-person gold standard testing was assessed using intraclass correlations (ICC). Intraclass correlations ranged from 0.60 to greater than 0.74, indicating that telephone administration was comparable to in-person administration, as reliability was in the good-to-excellent range. Further, the mean scores of the tests did not differ when analyzed using paired t-tests.

Discussion and conclusion: The results of these analyses show that tests within this battery can be successfully used over the phone to assess cognitive functioning. We are continuing the pilot study to reach a total sample size of 30. The development of telephone-based cognitive testing has never been undertaken and could become an integral resource to population-based, research studies. This testing could be especially valuable for subjects with severe, progressive physical disability and high caregiver demands, such as ALS.

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P51 BEHAVIOURAL CHANGE IN PRIMARY LATERAL SCLEROSIS: A CASE SERIES

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Keywords: primary lateral sclerosis (PLS), behaviour, screening

Background: It is now well recognized that ALS patients can have co-morbid behavioural changes that range from subtle findings to severe impairment fulfilling the criteria for behavioural variant fronto-temporal lobar degeneration. However, the literature examining behaviour in other types of motor neuron disease such as primary lateral sclerosis (PLS) is very sparse.

Objective: We aimed to document the frequency and pattern of behavioural changes in PLS patients.

Methods: All patients attending a Tertiary Multidisciplinary Motor Neuron Disease Clinic underwent screening for behavioural changes using a specifically designed and recently validated behavioural scale. We identified and interrogated data pertaining to patients with primary lateral sclerosis. The rate of different behavioural changes in PLS patients was compared to that of 50 age, sex and education matched healthy control that were recruited during tool validation.

Comparisons were made using non-parametric tests for continuous variables and Chi square with Monte-Carlo correction for proportions.

Results: Seven PLS patients were identified**.** Mean age was 62.9 years and 5 were males. Median R-ALSFRS score was 39.0 and median disease duration at time of testing was 100 months.

Two out the seven patients had scores above the cut off for abnormal behaviour, in both cases in within the range for severe behavioural change (as opposed to mild). These two patients were the oldest patients in the series and had the lowest ALSFRS-R scores in the cohort.

The most frequently reported behavioural change in PLS patients was over-sensitivity to external stimuli such as touch, smell etc reported in 6/7 PLS patients compared to 1/50 controls ($p < 0.0001$). Other behavioural changes observed more frequently in PLS patient compared to controls were inability to plan or foresee/solve problems ($p < 0.0001$); aggressiveness ($p < 0.0001$); repetitive behaviour ($p = 0.005$); self-centredness ($p = 0.005$); increased grammar mistakes ($p = 0.033$); and reduced display of emotion ($p = 0.013$).

Discussion and conclusion: Behavioural changes are not uncommon in PLS patients. Age and severe disease may be a risk factor. Larger studies are needed to confirm these findings.

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P52 COGNITIVE PHENOTYPES IN EUROPE

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Keywords: cognition, behaviour, phenotype

Objectives: The aim of this study was to develop and apply a single method for measuring cognitive change across European centres in an attempt to harmonize cognitive screening and compare cognitive phenotypes.

Methods: The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) has been previously shown to be sensitive to cognitive impairment in ALS (1). Here the ECAS was translated and applied to a total of 466 ALS patients (Scotland, n = 78, Ireland, n = 73, Italy-Milan n = 59, Italy-Turin n = 33, Holland, n = 40, Spain n = 40, Germany n = 73 and Switzerland, n = 35). Furthermore local normative data was collected within each country and cut-offs for abnormality determined.

Results: The ECAS was found to be sensitive to the types of impairment typically present in ALS patients across countries. The frequency of impairment within each country significantly differed ranging from 10 to 53% with a higher proportion of impaired patients from the samples in Ireland, Italy and Scotland and lower rates in Germany and the Netherlands. Furthermore the profile of impairment across cognitive domains also differed between countries with a greater proportion of patients with non-specific cognitive dysfunction (Memory, Visuospatial) in Ireland, while those in Scotland, Italy and Germany showed a more typical disproportionate impairment in ALS Specific functions (Executive, Language and Fluency).

There was a significant difference between samples in diagnostic delay and this significantly correlated with the degree of impairment in executive functions. Patients with a shorter delay indicating a more aggressive disease having more cognitive impairment. Furthermore duration of illness significantly correlated with ECAS scores, those patients with a longer duration of illness less likely to experience cognitive change. Age of onset also significantly correlated with cognitive impairment, with younger patients less likely to experience cognitive change. Frequency of impairment did not significantly differ according to symptom site of onset, which contradicts previously described associations with bulbar dysfunction, once tests which accommodate for speech and limb disability as the ECAS are used.

Genetic analyses were available from 81 patients. Those positive for the *C9orf72* mutation displayed ALS-specific cognitive dysfunction but were also more likely to have visuospatial dysfunction than those found to be negative for this mutation. The findings are related to clinical profiles including severity of disease and population differences.

Acknowledgements: This study was funded by the Motor Neurone Disease Association UK.

Reference:

1. Abrahams S. *et al.* Amyotroph Lateral Scler Frontotemporal Degener. 2014; 15: 9–14.

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P53 PROGRESSION OVER TIME OF GENDER DIFFERENCES IN EMERGING ALS COGNITIVE DECLINE FROM THE TEMPORAL TO FRONTOTEMPORAL REGIONS

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Keywords: gender, cognition, disease progression

Background: ALS is associated with frontotemporal lobar degeneration in ~50% of patients, in the absence of dementia, characterized by primary progressive aphasia and/or behavioural decline.

Objectives: We sought to investigate gender differences in pattern of emergence of cognitive decline in ALS and progression over 30 months, with repeat cognitive assessment every 6 months.

Methods: Beginning with a sample size of 356, we evaluated 152, 77, 48, 21 and 15 patients with the Penn State Brief Exam of Frontal and Temporal Dysfunction Syndromes (PSFTS) over the 30 month time period.

Results: Significant relative difficulties at time 1 were evident for females for 2-D constructions ($p = 0.044$), similarities ($p = 0.005$), judgment ($p = 0.018$), and calculations ($p = 0.045$). Medication records review of female oestrogen status for patients aged 31–74, showed a strong positive relationship between oestrogen levels and executive functioning capacities (similarities ($p = 0.005$), judgment ($p = 0.018$), letter fluency ($p = 0.004$)). At time 2, relative difficulties for females continued to be evident for 2-D constructions and calculations ($p = 0.15$), with a trend emerging for fluency advantage ($p = 0.110$). Time 1–2 comparisons evidenced progression among ALS cognitively normal (n = 80) to ALS cognitive impairment (ALSci) of 7.5%, with 25% now evidencing 1 deficiency and 67.5% remaining free of deficiencies. Gender differences were no longer evident by time 3, with 90% of patients found to be within the normal range on all measures administered. All patients remaining for times 4–6 were cognitively normal.

Discussion and conclusion: Gender differences are present in emerging ALSci, and relate to oestrogen status. Initial relative declines for females associate with the bilateral temporal regions subserving visual perception and verbal classification abilities, with a trend for relative difficulty for frontal cortical mediated working memory. With disease progression, female working memory relative disadvantage remains evident, while a female advantage begins to emerge for frontal cortical mediated verbal fluency. Time 1 findings evidence the potential of oestrogen as a therapeutic agent in emerging ALS-FTLD. Given the overlap in genes associated with ALS, FTLD and breast cancer, gonadal steroidal hormones likely serve as immuno-modulatory agents. Oestrogen analogs are needed to attenuate neurodegeneration while inhibiting over-activation in the breast and uterus, akin to the selective oestrogen receptor modulators currently applied as therapeutics in the treatment of breast cancer. Likely reasons for the lack of gender differences with disease progression include an increasing inability for individuals to pursue formal assessment due to ALS associated impairments, as well as the relative viability of individuals who remain alive and testable over 30 months, most of whom showed no progression of cognitive decline over time.

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P54 THE COGNITIVE RESERVE IN AMYOTROPHIC LATERAL SCLEROSIS WITH COGNITIVE IMPAIRMENT

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Keywords: cognition, cognitive reserve, genetics

Background: 50% of ALS patients show variable degrees of cognitive impairment, ranging from frank frontotemporal dementia (ALS-FTD), to executive cognitive impairment (ALS-ECI), non-executive cognitive impairment (ALS-NECI) and pure behavioural impairment (ALS-Bi). A protective role of cognitive reserve (CR) has been reported in Alzheimer's disease (AD) and pure FTD. No data about ALS with comorbid FTD are available.

Objectives: To verify the CR hypothesis in ALS patients with different degrees of cognitive impairment.

Methods: 183 incident cases resident in Piemonte and diagnosed from January 1st 2009 to December 31st 2011 constituted our discovery cohort. Our validation cohort (n = 113) included incident cases from Piemonte between January 1 2012 and June 30 2013 and patients coming from other regions diagnosed in the same period. All patients underwent neuropsychological testing and genetic analysis. In the discovery cohort: 49.7% were cognitively normal; 12.6% ALS-FTD; 19.7% ALS-ECI; 5.5% ALS-NECI; 6.0% ALS-Bi; 6.0% non-classifiable cognitive impairment (ALS-NCCI); one AD. In the validation cohort: 50.5% were cognitively normal; 15.9% ALS-FTD; 18.6% ALS-ECI; 3.5% ALS-NECI; 6.2% ALS-Bi; 5.3% ALS-NCCI. A Reserve Index (RI) (2–12) was calculated from years of education and occupational attainment.

Results: In the discovery cohort ALS-FTD patients had lower education level (4.7 years, SD 1.9) than all other groups and lower RI (4.9, SD 1.3) than all other groups ($p = 0.0001$). In the validation cohort ALS-FTD patients (7.0 years, SD 2.6) had the same education level of ALS-NECI (7.0, SD 1.4), that was lower than all other cognitive categories ($p = 0.003$). ALS-FTD patients had lower RI (5.7, SD 1.6) than all other groups but ALS-NECI ($p = 0.003$). Results were independent from sex, age and site of onset and were confirmed among *c9orf72* expansion carriers ($p = 0.012$). Neuropsychological tests related to RI were TMT B, TMT B-A, Stroop Colour - Word Interference Test, WAIS-R Block Design, WMS-R-Form 2, FAB ($p = 0.0001$) and CPM total score ($p = 0.001$).

Discussion and conclusion: Reserve mechanisms may play a role in cognitive impairment related to ALS, primarily in full-blown FTD, including *c9orf72* mutations carriers. CR in ALS-FTD is mainly related to frontal functioning.

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P55 MIRROR NEURON FUNCTIONING IN ALS – A LINK BETWEEN MOTOR AND THEORY OF MIND IMPAIRMENT?

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Keywords: mirror neuron, fMRI, theory of mind

Background: Amytrophic lateral sclerosis (ALS) is well known for its motor impairment and associated motor cortical changes. There is also increasing evidence that ALS patients show theory of mind (ToM) impairments. ToM is defined as the ability to attribute mental states, such as intentions, to others in order to understand and predict their actions. Interestingly, ToM has been associated with motor action observation in monkeys and healthy humans, via the so-called mirror neuron network. The mirror neuron network comprises dorsal and ventral premotor cortices, as well as inferior frontal gyrus (IFG) and inferior parietal regions. ALS cortical impairment in premotor regions could therefore potentially explain their ToM deficits. However, to date no study has investigated mirror neuron functioning in ALS, which could explain the relationship between premotor cortical integrity and ToM in these patients.

Objective: To establish the mirror neuron neural correlates in ALS.

Methods: Forty participants (ALS n = 22; age-matched controls n = 18) underwent a functional MRI experiment, which consisted of different action observations. Action observations involved hand-object interactions by an actor as well as a control condition (no interaction), based on previous monkey and fMRI work. Participants were shown all the videos before scanning commenced and watched blocks of each clip while lying in the scanner. The task was passive, not requiring motor responses from participants. Debriefing assured that participants watched and paid attention to the clips.

Results: Controls showed the expected mirror neuron network, including bilateral premotor and parietal cortices as well as IFG, when contrasting action observation and control conditions. By contrast, ALS patients showed markedly reduced activations for the same mirror neuron regions for the same contrast. The most marked differences were observed in the right dorsal and ventral premotor cortex as well as IFG for the action observation condition.

Discussion and conclusion: We demonstrate for the first time mirror neuron functioning in ALS. Our findings clearly indicate reduced activation in the regions implicated in mirror neuron function in ALS compared to age-matched controls. This was particularly pronounced over the right premotor cortices and IFG. These results give the first indication that ToM deficits in ALS might be inherently linked to the motor cortical deficits seen in this disease. Future studies need to investigate whether these mirror neuron changes are predictive of ToM functioning in ALS and might also explain ToM disturbances in related diseases, such as frontotemporal dementia.

In conclusion, mirror neuron network activations are reduced in ALS compared to controls, which establishes for the first time a link between motor and ToM impairments in this disease.

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P56 PREVALENCE AND IMPACT OF ALS BEHAVIOURAL IMPAIRMENT ON PATIENT AND CAREGIVER OUTCOMES OVER TIME

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Keywords: behavioural impairment, depression, caregiver burden

Background: Frontotemporal syndromes more common than frank FTD, particularly mild behavioural and personality changes are associated with significant caregiver burden in ALS. Few studies have examined frontotemporal behavioural syndromes longitudinally and their implications for both caregiver and patient psychological wellbeing.

Objectives: To determine whether the prevalence of ALS behavioural impairment increases as physical functioning deteriorates and to determine the impact of ALS behavioural impairment on patient and caregiver mood and burden of care over time.

Methods: Sixty-one patients with ALS (53% male; mean age = 62.6 years, SD = 12.4 years) and their caregivers (38% male; mean age = 60.0 years, SD = 12.2 years) were included. Standardised measures were used to evaluate changes in behaviour, depression, caregiver burden and physical functioning over time. Patients were sub-classified into ALS pure or ALS with behavioural impairment (ALSbi) according to current consensus criteria for the diagnosis of frontotemporal behavioural syndromes in ALS.

Results: The average time interval for the follow-up assessment was 11.7 months (SD = 6.3). There was no significant change in the proportion of patients with behavioural impairment at baseline (29%) and follow-up assessment (36%), $p = 0.6$. ALSbi patients had significantly more depressive symptoms than patients without behavioural impairment at baseline only ($p < 0.05$). Burden of care in caregivers of patients with behavioural impairment was significantly higher compared to caregivers of ALS pure patients at baseline only ($p < 0.05$). There was no significant difference in depression levels of ALS pure and ALSbi caregivers at baseline ($p = 0.4$) and follow-up ($p = 0.8$). Patient ($p = 0.9$) and caregiver ($p = 0.6$) depression as well as caregiver burden ($p = 0.2$) did not change over time despite a significant decline in patients' physical functioning ($p < 0.05$).

Discussion and conclusion: The prevalence of ALS behavioural impairment did not increase as the disease progressed. Although patients with behavioural impairment had more depressive symptoms and their caregivers a greater sense of burden initially, patient and caregiver depression levels as well as caregiver burden remained stable over time. These findings suggest the importance of considering individual differences related to psychological and coping responses of ALS patients. This will provide much needed guidance on how patients are most likely to benefit from psychological support appropriate to their psychological status and needs.

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P57 THE RELATIVE IMPACT OF PATIENTS' DISEASE SYMPTOMS, COGNITION AND BEHAVIOUR ON THE PSYCHOSOCIAL WELLBEING OF ALS CAREGIVERS

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Keywords: caregiver outcomes, disease severity, cognitive-behavioural impairment

Background: In addition to the debilitating physical impairment of ALS, up to 50% of non-demented patients may experience mild-to-moderate cognitive and behavioural symptoms qualitatively similar to that shown by patients with Frontotemporal dementia (1). ALS patients have demonstrated deficits on tasks assessing executive function and social cognitive abilities, such as the ability to identify emotions from faces or attribute mental states to others (2). The relative impact of the physical and behavioural consequences of ALS on caregivers has been considered (3, 4), but no studies have examined if objective indices of patients' neuropsychological performance on tasks of executive function and social cognition contribute to caregivers' experiences of the disease.

Objectives: This study sought to examine the relative impact of patients' disease symptoms, behavioural involvement, executive dysfunction and impairment in social cognition on caregivers' psychosocial functioning.

Methods: Thirty-five spouse caregivers rated their mood, perceived level of burden and marital satisfaction. Spouses also rated their partners' behaviour in terms of everyday executive dysfunction, apathy, disinhibition and emotional lability. Composite scores of patients' performance on a battery of neuropsychological tests were created to measure their executive function and social cognition abilities. Patients' disease severity was also recorded.

Results: Bivariate correlational analysis was used to identify potential predictors of the caregiver outcomes. Regression analyses found that, together, the severity of patients' limb involvement ($\beta = 0.69$, $t(28) = 9.3$, $p < 0.001$) and behavioural problems ($\beta = -0.51$, $t(28) = -0.68$, $p < 0.001$) predicted caregiver burden ($R^2 = 0.84$, $F(2, 28) = 80.6$, $p < 0.001$). In addition, while limb symptom severity ($\beta = -0.45$, $t(31) = -2.8$, $p = 0.008$) predicted caregiver depression ($R^2 = 0.21$, $F(1,31) = 8.1$, $p = 0.008$), behavioural involvement ($\beta = 0.40$, $t(30) = 2.4$, $p = 0.0$) predicted caregiver anxiety ($R^2 = 0.16$, $F(1,30) = 5.65$, $p = 0.02$). Together, behavioural problems ($\beta = -0.42$, $t(27) = -4.65$, $p < 0.001$) and caregivers' self-rated marital satisfaction prior to their spouses' ALS ($\beta = 0.68$, $t(27) = 7.59$, $p < 0.001$) predicted caregivers' current marital satisfaction ($R^2 = 0.796$, $F(2,27) = 52.68$, $p < 0.001$). The cognitive composite scores were not associated with caregiver outcomes.

Discussion and conclusion: The study highlights the influence of ALS functional disability and perceived everyday behavioural dysfunction in ALS patients on caregivers' psychosocial health. The results suggest a possible specificity with which different ALS symptoms may impact on spouse caregivers. Caregivers' perceptions of cognitive-behavioural impairment in ALS patients may be more important for caregivers' outcomes than patients' objective neuropsychological performance. Clinical communication with ALS families should emphasize both physical and psychological challenges presented by the disease.

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P58 DEPRESSION IN PATIENTS WITH ALS/MND AND ITS ASSOCIATION WITH FUNCTIONAL STATUS AND COPING STRATEGIES

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Keywords: depression, coping strategies, functional status

Background: Data on depression in patients with ALS/MND show mixed results (1–2) and further research is needed to estimate the prevalence of mood disorders in such a population and better understand the relationship between depressive symptoms and other relevant variables.

Objectives: The aims of the present study are those of evaluating depressive symptoms in a population of patients with ALS and examining the associations between depression and functional status or coping strategies.

Methods: Scales for the evaluation of depression (BDI-II) (3), coping strategies (MND Coping Scale) (4) and functional status (ALSFRS) (5) were administered to 43 patients with ALS/MND. Bivariate correlations between the scales were calculated.

Results: The average BDI-II score is 16.17 ± 9.63. Most of the patients (approximately 70%) show minimal or mild depressive symptoms. Depression negatively correlates ($p < 0.05$) with functional status and with the following coping strategies: Positive Action ($p < 0.01$); Independence ($p < 0.01$); Positive Thought ($p < 0.01$).

Discussion and conclusion: Even if possibly biased by the items referring to physical symptoms of depression, the BDI-II scores of the present sample confirm that clinical depression is not widespread in patients with ALS/MND, and that perceived autonomy and active coping strategies could be related with better adjustment.

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P59 COPING STRATEGIES AMONGST NEWLY DIAGNOSED ALS PATIENTS

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Keywords: coping strategies, newly diagnosed, support

Background: Amyotrophic lateral sclerosis is a fatal disease with impact on both physical function and psychological well-being. Patients use different coping strategies to manage symptoms and disease progression, but there is scarce knowledge about coping in an early stage of the disease.

Objectives: The aim of this study was to prospectively identify coping strategies used by newly diagnosed ALS patients and whether they change over time. We also wanted to determine if there were a correlation between physical function, psychological well-being, age and gender and the use of different coping strategies.

Methods: Thirty three patients were included in the study. Coping strategies was measured using the MND coping scale (1) and psychological well-being was measured with the Hospital Anxiety and Depression Scale (2). The patients' physical function was estimated with the revised ALS Functional Rating Scale (3). The evaluation was made one to three months, and six months after diagnosis.

Results: At both time points, support and independence was the most commonly used strategies, whilst the most seldom used were avoidance/venting and information seeking. Patients < 64 years old used positive action more often than older patients. There was a positive correlation between positive action and the patients' physical function at time point 1. Psychological well-being was correlated with the use of different coping strategies.

Discussion and conclusion: Support, which probably involves different kind of aids, seems to help patients to be independent and to cope with disease progression. Patients' psychological well-being correlated with different coping strategies and the use of different strategies changed over time.

The knowledge about the variation in use, and the correlation between coping strategies and psychological well-being in an early stage of ALS is important in developing support for the patient during disease progression.

Acknowledgements: This study was founded by the Ulrica Croné Foundation, Uppsala University and Uppsala University Hospital. A special thanks to the patients who participated in this study.

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P60 INFORMATION NEEDS AND INFORMATION SEEKING PREFERENCES OF ALS PATIENTS AND THEIR CARERS

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Keywords: information seeking, internet search, caregivers

Background: Information seeking behaviour has been investigated in relation to numerous aspects of disease. This is of particular relevance in view of a rapidly advancing health information technology such as the Internet. Many studies refer to patients with cancer or other chronic diseases and are not easily applicable to patients with ALS. Two studies investigated the information seeking behaviour in ALS and PLS, one in Italy and one in the United States (1, 2). These data cannot easily be transferred to ALS patients in other countries as Internet access and health related Internet use differs between countries, even within Europe.

Objectives: To investigate the information seeking behaviour in patients with ALS and their caregivers and their rating of the usefulness of different information sources in Germany.

Methods: Survey in 106 patients and 100 caregivers in two university ALS outpatient clinics.

Results: Before seeing a doctor, 28% of patients and 23% of caregivers had used other sources to find symptom-related information, mostly via the Internet. Although two-thirds were satisfied with the way of diagnosis disclosure, 88% of patients and 85% of caregivers searched for additional information, most often on the internet (patients 72%, caregivers 85%), followed by patient brochures (patients 58%, caregivers 66%). Internet, patient brochures and the German Neuromuscular Disease Society were rated most frequently as useful/very useful. Traditional print media and interpersonal contacts were also frequently used and most respondents relied on more than one source for information. Only few respondents used the Internet for exchange with other patients. Two-thirds wanted to discuss web contents with their physician.

Discussion and conclusion: Patients with ALS and their caregivers clearly have additional information needs. Besides traditional information sources the Internet is frequently used. Therefore, reliable and useful websites should be provided. The patients' and caregivers' need to discuss their findings with a physician should be acknowledged.

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P61 THE SUBJECTIVE FEELING OF AUTONOMY AND FAMILY BONDING – CONFLICTING PRIORITIES IN DECISIONS TO PROLONG OR SHORTEN LIFE IN ALS

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Keywords: decision-making, autonomy, caregivers

Background: The patients with amyotrophic lateral sclerosis (ALS) are faced with different decisions in the course of the disease regarding life prolonging (non-invasive ventilation, mechanical long-term ventilation and percutaneous endoscopic gastrostomy) and life shortening treatments (Euthanasia and physician assisted suicide). Determinants of these decisions are multifactorial and only some have been defined so far. The aim of this study was to identify possible determinants of the decision making process such as feeling of autonomy and family bonding.

Methods: A total of 100 ALS-patients were interviewed with questionnaires concerning their decisions of life prolonging and life shortening treatments. Possible determinants were acquired such as subjective feeling of autonomy (autonomy as core value, independence as coping strategy) and family bonding (demographic data, shared decision making). Furthermore, determinants such as subjective quality of life, depression, strategies in coping, religiousness and various factors of cognition were measured. Logistic and linear regressions were used for statistical analysis of the identification of determinants on decision making. Additionally, qualitative in-depth interviews ($n = 10$) were performed and analysed by qualitative content analysis.

Results: Family bonding was a strong determinant of decisions to prolong life. The majority (93%) of the patients named the wishes of their caregivers as important for them and 79% declared that the opinion of their caregivers influences their decisions. Similarly, increasing number of children of the patients showed a significant impact on the decisions to prolong life ($p = 0.03$, $R^2 = 0.38$). Concurrently, the patients showed a strong need for autonomy which turned out to be a strong determinant of decisions to shorten life ($p = 0.04$, $R^2 = 0.51$). Furthermore, degree of depression ($p < 0.01$,

$R^2 = 0.21$) and religiousness ($p = 0.02$, $R^2 = 0.23$) had a significant influence on fatal decision making. Cognitive impairments however had no impact on decisions (all $p > 0.05$).

Discussion and conclusion: The results demonstrate a distinct discrepancy between the patients need for autonomy and the influence of the patient's family bonding on their decisions. Patients that are more influenced by their need for autonomy decide towards life shortening treatments, whereas the patients that are influenced by their family ties tend to decide towards life prolonging treatments.

Among other determinants, conflicting issues of subjective feeling of autonomy and family bonding have to be considered by the multidisciplinary teams in counselling, treatment and therapy of ALS patients.

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P62 AN EXPLORATION OF BEREAVED FAMILY CARERS' ACCOUNTS OF THE END-OF-LIFE EXPERIENCE OF PEOPLE WITH MND

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Keywords: dignity therapy, interviews, caregivers

Background: Palliative care is traditionally provided mainly to cancer patients. Expansion of palliative care services into non-malignant conditions has led to improvements in symptom control, quality of life and caregiver burden for people with MND. However, there are still gaps for people with MND and their families when it comes to access to specialist palliative care services. The end of life can be especially difficult for people with MND and families as the terminal phase in MND is not predictable and care needs are typically high. When there are gaps or problems in the provision of care at the end of life for people with MND, the burden falls squarely on family carers who are often overburdened and already experiencing high levels of distress. Identifying where the gaps are and what are unmet needs will help provide best practice end of life care for all people with MND and will help to reduce the burden on family carers and MND health professionals.

Objectives: This qualitative study explored the accounts of bereaved family carers about their perceptions of the end-of-life experience of people with MND.

Methods: Semi-structured interviews were used to elicit accounts of the experience of the end of life and the death of the person with MND from 12 bereaved family carers who were bereaved between three and 15 months. A social constructionist approach was used to elicit people's own experiences as the study was exploratory and applied. The study focused on three main areas: 1) the health care services used in the last three months of life with a special focus on the last week of life, 2) the information provided about the end of life to families from health care providers, and 3) the family carer's view of the death experience. The semi-structured approach allowed for people to give other information they felt relevant. Thematic analysis of the transcribed interviews was conducted. A former MND family carer and four MND specialist health care providers associated with an MND Clinic served as the Project Advisory Group to provide input into all aspects of the study.

Results: Three key themes emerged: The provision of support; information seeking; and preparation and readiness for death. Sub-themes included who people received support from, unmet needs, what worked well and personal strategies for coping. Recommendations for enhancing support and providing best care at the end of life are discussed.

Acknowledgements: This research is supported by a grant from the Cancer and Palliative Care Research Unit at the University of Western Australia. The research was also supported by the MND Association WA and Silver Chain Hospice Service. Thanks are also given to the advisory group members and all participants who generously gave their time.

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P63 THE EXPERIENCE OF MEDITATION IN ALS: A QUALITATIVE STUDY ABOUT THE EFFICACY OF A MINDFULNESS MEDITATION PROTOCOL WITH ALS SUBJECTS AND CAREGIVERS

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Keywords: quality of life, mindfulness, meditation

Background: Amyotrophic lateral sclerosis (ALS) is a progressive and fatal neurodegenerative disease, clinically characterized by progressive weakness leading to death by respiratory insufficiency, usually within three years. Although the patient's intellect and personality usually remain unimpaired, as the disease progresses the patient develops a worsening disability and impairment of the speech, often resulting in social isolation and a high degree of psychological suffering. The combination of ALS with progressive dependence and immobility might elicit feelings of despair, depression and anxiety.

To date, some of the most promising clinical treatments for the reduction of distress are based on mindfulness meditation practices, in particular the Mindfulness-Based Stress Reduction (MBSR), developed by Jon Kabat-Zinn. This program has proven useful to reduce stress levels and promoting resilience.

Objectives: This study tests the hypothesis that stress reduction methods based on mindfulness meditation can attenuate and prevent anxiety and depression, relieving the distress of living with ALS and providing skills to manage stressful events connected with the illness.

Methods: Sixty-three subjects with ALS were assigned to the ALS-MBI protocol or to a control condition (usual care). The Mindfulness-based Intervention protocol is derived from

MBSR, whilst respecting clinical peculiarities of ALS: physical exercises were removed, with a higher focus on the cognitive and affective issues. Other exercises, taken from different approaches with similarities to the mindfulness techniques (eg, Feldenkrais method), have also been inserted. Each of the eight sessions emphasized accepting the discomfort and physical limitation of ALS, focusing on resources and abilities that still remain. Seven exercises were devoted to mindful eating, breathing awareness, body scan, practice of loving kindness, Hatha yoga, music meditation, visualizations and motor imagery. Participants were asked to do homework on a daily basis. As part of a larger study, subjects were interviewed about their experience with mindfulness, aimed to understand the changes following the intervention. Qualitative data were analyzed with content analysis.

Results: Our preliminary results showed that both ALS patients and caregivers expressed positive evaluations on the mindfulness intervention. Acceptance and non-judgment seem to be powerful tools for the improvement of well-being in the ALS field. According to these preliminary results, the ALS-MBI protocol can improve (or restore, when required) the expression and the awareness of positive emotions.

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P64 DIGNITY THERAPY FOR PEOPLE WITH MOTOR NEURONE DISEASE AND THEIR FAMILY CAREGIVERS: A FEASIBILITY STUDY

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Keywords: dignity therapy, palliative care, caregivers

Background: There are calls to explore psychological interventions to reduce distress in Motor Neurone Disease (MND) patients and their family caregivers (FC). Dignity Therapy (DT) is a short term psychotherapy intervention shown to alleviate distress for people with life limiting illnesses. Patients are invited to discuss issues that matter most or that they would most want remembered about their life. Sessions are transcribed and edited with a final version (generativity document) returned to the patient, for the patient to bequeath to a family member or a friend, thus becoming part of a personal legacy.

Objectives: To assess the acceptability, feasibility and effectiveness of DT to reduce distress in people with MND and their FCs.

Methods: The study used a repeated measures design pre and post-intervention. Acceptability and feasibility were assessed using participants' ratings of the helpfulness of the intervention across several domains and time and resources required. Effectiveness measures for patients included: dignity-related distress (Patient Dignity Inventory); hopefulness (Herth Hope Index); spiritual wellbeing (FACIT-sp 12). Those for FCs included burden (Zarit Burden Interview); hopefulness (Herth Hope Index); anxiety; depression (HADS).

Results: 27 patients and 18 FCs completed the intervention. DT was well accepted including by patients who required assisted communication devices. The high satisfaction and endorsement of DT by patients suggests it has influenced various important aspects of end of life experience such as helped them attend to unfinished business and made them feel like they were still themselves. FCs overwhelmingly agreed that the DT document is and will continue to be a source of comfort to them and they would recommend DT to others in the same situation.

Discussion and conclusion: This is the first DT study to focus on MND and on home-based caregiving. The therapy needs to be offered earlier. Results established the importance of narrative and generatively for patients with MND and may open the door for other neurodegenerative conditions.

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P65 DO PATIENTS WITH MND EXPERIENCE DIFFICULTIES IN PERFORMING FINANCIAL ACTIVITIES AT HOME? EVIDENCE FROM A NOVEL ECOLOGICAL INSTRUMENTAL ADL TASK

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Keywords: functional disability, financial ability, neuropsychiatric symptoms

Background: With the recognition of the pervasive mild cognitive and neuropsychiatric symptoms in MND (1), it is critical to investigate how these symptoms may directly affect everyday living tasks requiring more complex reasoning.

Objectives: To investigate patients' ability to perform a well-validated ecological financial task.

Methods: 18 patients in the ALSFTD continuum (15 MND; 1 FTDMND; 2 FTD) were evaluated in an ecological finance task from the IADL Profile (2), well validated in patients with traumatic brain injury. The 'making a budget' task requires planning and management of expenses to achieve a fictitious goal. It assesses three operations: the ability to 1) plan; 2) carry out the task, including perceiving and correcting one's own errors during the task; and 3) verify if the task goal was successfully achieved or whether the task needs to be redone. Scores of independence are generated, one for each operation, based on type and amount of assistance required. Other assessments included global cognition (ACE-R: maximum score 100; cut off 88) and neuropsychiatric symptoms (MiND-B: maximum score 36; cut off 32).

Results: Only 22% of patients were completely independent in performing the IADL task. In fact, 61.1% of patients

required verbal assistance to complete the task and 16.7% could not complete the task at all. More specifically, patients had marked difficulty in 'carrying out the task' (83.3% were dependent), and difficulty in 'verifying attainment of goal' (77.8% required help). There was no clear difference in scores between the FTD, FTDMND and MND patients.

There was a strong association ($p < 0.05$; $R = 0.604$) between scores on the IADL task and the MiND-B, reflecting the relationship between ALS plus symptoms and difficulties in carrying out the budget task.

Discussion and conclusion: The great majority of patients in this study could not perform a complex ecological validated assessment of financial abilities independently or efficiently. These novel preliminary findings suggest that it may be advisable to oversee and support patients with neuropsychiatric symptoms as these can be affecting important daily functions.

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P66 AN ISOLATED DEFICIT: SOCIAL AFFECTIVE DEFICITS IN BULBAR ONSET ALS PATIENTS

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Keywords: neuropsychology, social-affective, impairment

Background: There is considerable debate about as to whether social cognitive processes are subsumed by metacognitive and executive functions, or whether they represent auxiliary frontotemporal processes. Cortical and subcortical atrophy has been noted in Amyotrophic Lateral Sclerosis (ALS), congruent with the neuroanatomical basis for social cognitive processes. The present study investigated Theory of Mind (ToM) deficits in patients with ALS.

Objectives: The aims of this research were to investigate whether social cognitive processes declined at a congruent rate to executive dysfunction in ALS.

Methods: Participants were recruited as part of an ongoing Irish population based study investigating cognitive decline in ALS. After removing patients whom were *C9orf72* positive, participants were grouped based on whether patients had bulbar ($n = 20$) or spinal onset ($n = 39$) ALS. Gender, age, IQ and education matched healthy controls were used to generate culturally specific comparative data for within-patient analyses ($n = 60$). Measures of social cognition included the Reading the Mind in the Eyes Test, measuring affective ToM and the Judgement of Preference Task, measuring cognitive ToM. Executive function was assessed using The Brixton Spatial Anticipation Test, Digit-Span and Lexical Fluency.

Results: On affective ToM, there was a significant difference between bulbar and spinal onset patients on this task ($p = 0.001$). Comparing bulbar and spinal patients standardized scores of executive function yielded no significant differences.

Discussion and conclusion: Results indicate the presence of social-affective deficits within ALS, prior to characteristic executive and language dysfunction, for bulbar onset patients without comorbid deficits. These scores may illustrate a decline in social cognition prior to other higher order functional deficits. Bulbar patients scored significantly lower than both spinal onset patients and controls, and this could be due to a number of contributory factors. These findings shall be discussed in relation to current neuroimaging research and neuropsychological theory.

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P67 EATING BEHAVIOR AND METABOLIC CHANGES ACROSS THE SPECTRUM OF MOTOR NEURON DISEASE AND FRONTOTEMPORAL DEMENTIA

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Background: Increasingly it is recognised that motor neuron disease and frontotemporal dementia are spectrums of the one disease. Abnormal eating behaviours represent a core diagnostic feature for behavioural frontotemporal dementia (bvFTD), yet little is known about their effect on metabolic health and how this contrasts to the metabolic profile in motor neuron disease (MND).

Objective: We aimed to define the patterns of eating behaviour and intake in bvFTD and examine the association between eating behaviours and metabolic health of FTD compared with MND, where the metabolic profile has been hypothesised to affect disease progression. We hypothesised that as cognitive impairment increased across the spectrum of FTD MND so too would BMI and that changes in BMI, rather than being related to swallowing difficulties and loss of muscle mass may be centrally mediated.

Methods: Carers of 21 bvFTD, 26 Alzheimer disease control and 18 healthy control subjects completed validated questionnaires on appetite and eating behaviour. Body mass index (BMI), and blood samples measuring cholesterol and insulin levels were prospectively collected. BMI measurements were compared to a cohort of 100 MND (bulbar and limb) and FTD MND patients.

Results: BvFTD patients displayed significant abnormalities in all domains of eating compared to AD patients. BvFTD patients had significantly increased carbohydrate intake and a trend towards increased sugar intake compared to controls, yet they had similar hunger and satiety levels.

BMI measurements were significantly higher ($p < 0.01$) in the bvFTD, FTD MND and MND plus groups compared to the pure MND (bulbar and limb) and control groups. BvFTD patients had significantly ($p < .001$) increased insulin levels, triglyceride levels and an increased total cholesterol to HDL ratio, and a lower HDL cholesterol level compared to controls, suggesting they are insulin resistant and hyperlipidemic.

Discussion and conclusion: Abnormal eating behaviour is prominent in bvFTD, and is associated with increased sugar and carbohydrate intake and not explained by changes in satiety and hunger. Similar findings have been found in obese individuals and related to changes in the hypothalamus. Abnormal eating behaviour in FTD is associated with changes in BMI, and a blood metabolic profile similar to that seen in MND. BMI tends to increase as cognitive impairment develops in MND suggesting a FTD MND continuum, and that similar cortical structures to those involved in FTD may be involved in MND. The present findings have prognostic implications in terms of disease progression and may help to better delineate the metabolic profile of MND and FTD patients.

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